Retina Atlas

Series Editors: Sandeep Saxena · Richard F. Spaide · Eric H. Souied · Timothy Y.Y. Lai

Trauma and Miscellaneous Disorders in Retina

Eduardo Büchele Rodrigues
Carsten Helmut Meyer
Eduardo Tomazoni
Editors

Springer

Retina Atlas

Series Editors

Sandeep Saxena, MS, FRCSEd, FRCS, FRCOphth
Department of Ophthalmology,
King George's Medical University,
Lucknow, Uttar Pradesh, India

Richard F. Spaide, MD
Vitreous Retina Macula Consultants of New York,
New York, NY, USA

Eric H. Souied, MD
Department of Ophthalmology,
University Paris-Est Créteil,
Créteil Cedex, France

Timothy Y.Y. Lai, MD, FRCS, FRCOphth
Department of Ophthalmology and Visual Sciences,
Chinese University of Hong Kong,
Hong Kong, Hong Kong

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Eduardo Büchele Rodrigues
Carsten Helmut Meyer • Eduardo Tomazoni
Editors

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Editors
Eduardo Büchele Rodrigues
Department of Ophthalmology and
Visual Sciences
Federal University of São Paulo
São Paulo
Brazil

Department of Ophthalmology
Saint Louis University School of Medicine
Saint Louis, MO
USA

Eduardo Tomazoni
Ophthalmology, Retina Fellowship
Visum Retina
Florianópolis
Santa Catarina
Brazil

Carsten Helmut Meyer
Department of Ophthalmology
Philipp University of Marburg
Marburg
Germany

ISSN 2662-5741 ISSN 2662-575X (electronic)
Retina Atlas
ISBN 978-981-13-8549-0 ISBN 978-981-13-8550-6 (eBook)
https://doi.org/10.1007/978-981-13-8550-6

This Springer imprint is published by the registered company Springer Nature Singapore Pte Ltd.
The registered company address is: 152 Beach Road, #21-01/04 Gateway East, Singapore 189721, Singapore

Contents

About the Editors

Eduardo Büchele Rodrigues is an Assistant Professor of the Saint Louis University and Affiliated Professor of Ophthalmology of the Federal University of São Paulo. He has published around 160 scientific articles in leading international journals, and he is one of the editors of the major international book on the use of medications in the treatment of retinal diseases, *Retinal Pharmacotherapy*, published in 2010, and is Co-Editor-in-Chief of the *International Journal of Retina and Vitreous*. He has presented more than 250 papers at national and international conferences and received 19 awards at congresses.

Carsten Helmut Meyer holds an M.D. degree and is a specialist in vitreoretinal diseases. He attended medical schools in Freiburg, Innsbruck, and Oxford and received his medical degree from the University of Munich, Germany. Dr. Meyer completed his residency at the University Eye Hospital, Luebeck, Germany, and in 1999 pursued a 2-year vitreoretinal research fellowship at the Duke Eye Center, Durham, NC. He served as an Assistant Professor at the Department of Ophthalmology, Philipps-Universität, Marburg, and as a Professor at the Department of Ophthalmology, University of Bonn. Dr. Meyer is an editorial board member of several ophthalmic journals and serves as a reviewer for more than 65 peer-reviewed journals. He has published several book chapters and 240 articles in peer-reviewed journals. He received the AAO Achievement Award (2008), is a member of the Retina Society, Club Jules Gonin, EURETINA, and Deutsche Ophthalmologische Gesellschaft (DOG), and is listed in Marquis Who's Who and ORBIS. He is also a dedicated examiner on the panel of the European Board of Ophthalmology (EBO).

Eduardo Tomazoni is graduated in medicine from the University of Joinville Region and qualified as an ophthalmologist at Governador Celso Ramos Hospital, in Florianópolis-SC, Brazil. He is currently working as a Retina Fellow at Lumine Institute, Florianópolis-SC, Brazil. He is a member of the Brazilian Society of Ophthalmology and is also a reviewer for the *International Journal of Retina and Vitreous*.

Posterior Ocular Trauma

1

Müller Gonçalves Urias

Introduction

Ocular injury is an important cause of visual impairment on working-age individuals and is the leading cause of hospitalization of ophthalmologic patients (Négrel and Thylefors 1998). Ocular trauma is divided into open-globe injuries and closed-globe injuries. On both types, the posterior segment may be injured, and even with no direct eye injury, some posterior changes could be trauma-related—i.e., electrical burns, Purtscher retinopathy, and Valsalva retinopathy.

Some factors might impact on injury severity such as associated energy, occurrence of intraocular foreign body and also the foreign body characteristics, such as sharpness and size. These factors might lead to several damages and impact patient prognosis (Rahman et al. 2006; Knyazer et al. 2008).

The diagnosis—once there is a trauma history involved—is not as challenging (and controversial) as the treatment (Mittra and Mieler 1999). However, since there are many posterior segment features related to trauma, this chapter aims not only to describe those features but to present some of their proposed treatments and prognosis.

Epidemiology

An important data source on ocular injury was published in 1998 and collected data since 1971 (Négrel and Thylefors 1998). According to this source, 19 million individuals went blind or had some vision impairment due to ocular trauma, 55 million cases restricted their activities more than 1 day, and 750,000 required hospitalization each year. Besides that, authors mentioned a bimodal curve on age prevalence, with the first group at 5–25 years old and the second above 70 years old. They also mentioned that ocular trauma was more prevalent in male patients (four times higher). Moreover, according to the authors, there were 203,000 open-globe injuries within that period.

Incident prevention is crucial. Recognizing their patterns and areas with high prevalence is therefore equally important. The first care might represent a critical role in patients prognosis, so it might resemble why resources in providing adequate care and a well-trained professional are fundamental for those injuries.

Diagnosis

History

There are considerations beyond the presence of a trauma history. To link each patients clinical findings to their trauma history, might demands from the professional to understanding those findings and conclude if they are related or not to trauma. There are challenges, for example, when there is no related trauma history, but there are signs that might suggest otherwise, and also when the patient is unconscious. Table 1.1 describes some few examples of those situations.

Special attention is required on situations when there is no trauma history in a children—especially infants—with posterior segment findings, such as retinal and vitreous hemorrhage. Those findings might suggest child abuse, and the management involves a multi-professional approach with extreme caution (Duhaime et al. 1998).

Another important history related to elucidate is if the patient was handling anything during trauma and, if so, what kind and in which conditions it was. This in crucial in a way that, according to some materials, imaging for example, computer tomography, might be relevant, and this might not only change management but also prognosis (Loporchio et al. 2016).

M. G. Urias (✉)
Department of Ophthalmology, Federal University of Sao Paulo, São Paulo, Brazil
e-mail: urias@unifesp.br

E. B. Rodrigues et al. (eds.), *Trauma and Miscellaneous Disorders in Retina*, Retina Atlas, https://doi.org/10.1007/978-981-13-8550-6_1

Table 1.1 Examples of challenging diagnostic cases on different situations

Examples of challenging diagnostic situations			
Posterior clinical finding:	A)…not related to the trauma history related by the patient	B)… no trauma history referred	C)… no history at all
Example:	Hypotony and choroidal folds with hyphema or vitreal retinal hemorrhages	An unconscious infant with retinal or vitreal hemorrhages	An unconscious unattended 35-year-old victim of car crash with peripheral retinal hemorrhages
Consider for management:	Rule out hidden globe rupture	Rule out shaken baby syndrome	Rule out intracranial changes and verify other systemic disorders with retinal hemorrhages

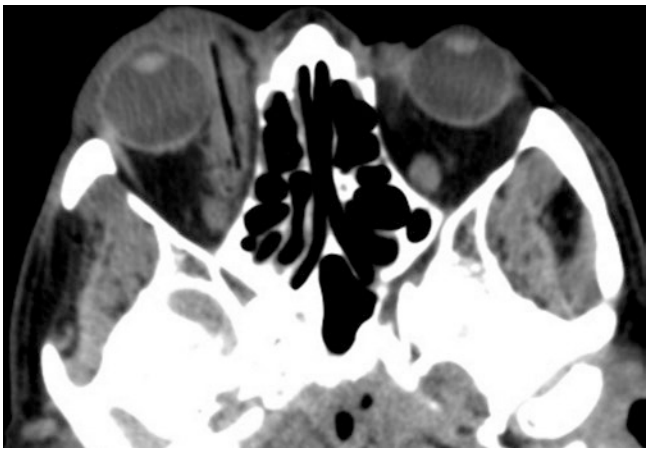

Fig. 1.1 Axial orbital CT scan. A patient who referred eyelid edema after falling from a stair in his garden. No signs of perforation or penetration. Peripheral choroidal folds in OD fundus exam. Intraorbital foreign body suspected in clinical exam and confirmed in CT scan

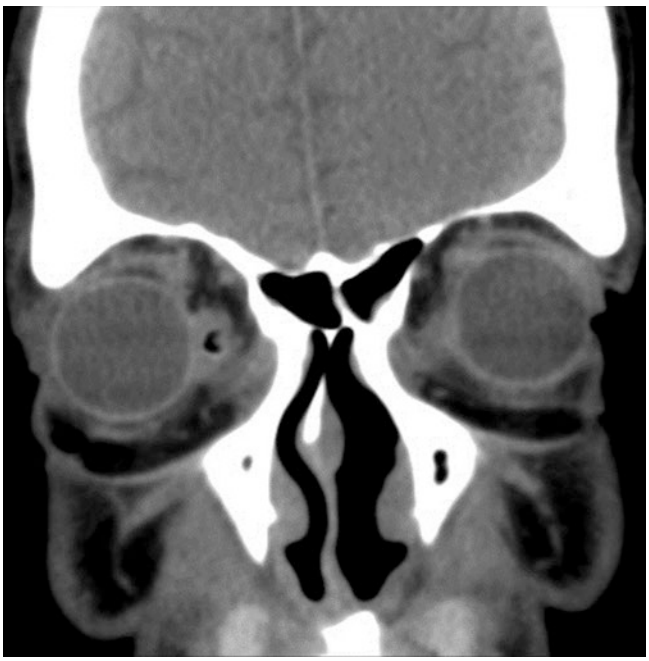

Fig. 1.2 Coronal CT scan from the same patient

Exam and Clinical Features

After identifying the complaints and their timing, the trauma mechanism, and other remarkable features, a careful ocular examination is fundamental for a following conduct. According to trauma severity, the evaluation may be laborious, but an error—i.e., obtaining visual acuity incorrectly or even demanding that in a stressful way, for example with a woman after an abbuse history – might represent a change in management that might not be adequate for that situation. In the same way, demanding a physical examination without proper anesthesia might result in undiserable effects, for example increased systemic blood pressure or even a Valsalva maneuver - which might lead to vitreal and uveal extrusion on an open-globe injury.

As the main prognostic predictor, visual acuity is usually the first and the main feature on the ophthalmological examination. Field testing and even an Amsler grid could be also helpful in some cases. After the visual quantification and qualification, pupils reflex and ocular extrinsic motricity should be evaluated. Slit lamp exam and intraocular pressure measurement, when possible, are also important. In some cases, both exams combined could suggest, for example, a hidden globe rupture (hyphema associated with hypotony) or even retinal detachment (*tobacco* dust, changed red reflex with decreased intraocular pressure).

As one of the crucial steps, a dilated fundus exam should be conducted, always with careful handling when opening the eyelids - especially if there is a suspected open-globe injury. With means opacity and suspected open-globe suspected, orbital imaging with computed tomography (CT) scan (Figs. 1.1, 1.2 and 1.3) or magnetic resonance imaging (MRI) should be performed (Kubal 2008). On close-globes injuries associated with means opacity—or after suturing a laceration in an open-globe injury—ocular ultrasound should be considered.

Posterior segment might be affected in an ocular injury with extensive possible findings, such as choroidal changes (folds, ruptures, hemorrhage, detachment), photoreceptor changes (*commotio retinae*), retinal hemorrhages (intraretinal or preretinal), vitreal consequences (vitreal extrusion with traction, macular hole, retinal tears, retinal detachment), optic nerve damage (extrusion, swelling), and even through a direct foreign body damage (Yan 2016).

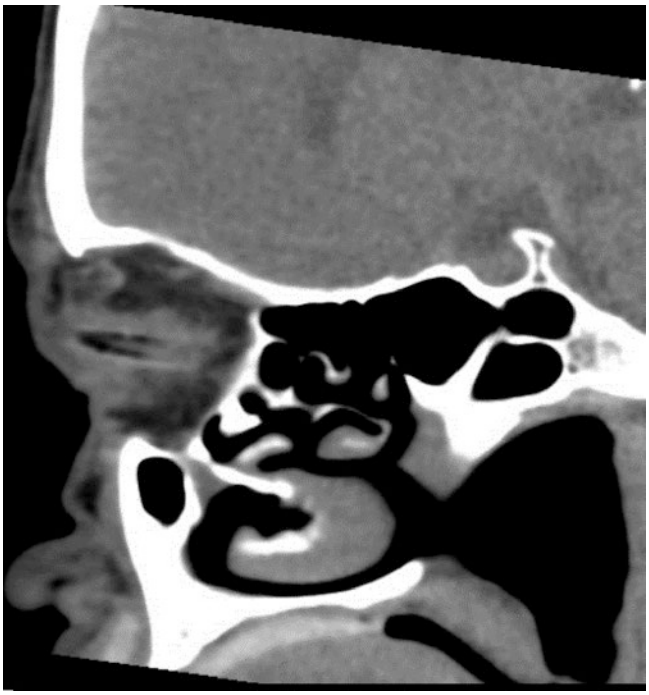

Fig. 1.3 Sagittal CT scan from the same patient

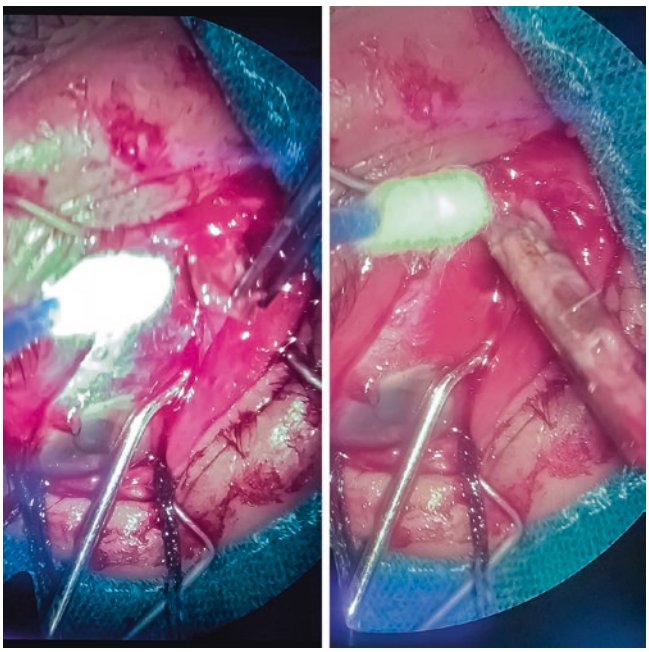

Fig. 1.4 Surgical exploration of the case above, with the removal of an intraorbital foreign body (courtesy of Allyne Kallaf MD)

Identifying those clinical features is crucial and their management will be discussed in the following chapters.

Management

Open-globe injuries (Figs. 1.4 and 1.5) must be recognized and surgically treated as soon as possible. Prevention of endophthalmitis must be addressed and considered when there is associated risk. The presence of a foreign body and its material must be identified, especially with regards to some materials toxicity, i.e., iron (Loporchio et al. 2016). However, factors such as a considerable vitreal and uveal extrusion along with an increased risk of endophthalmitis—and particularly in the absence of light perception—must also be considered in management. Unfortunately, in some cases, the evisceration or enucleation might be considered as an inevitable choice with regards to prognosis (Rahman et al. 2006; Knyazer et al. 2008; Man and Steel 2010; Zhang et al. 2010).

In closed-globe injuries with suspected hidden globe rupture, surgical exploration must be performed after imaging. Closed-globe injuries, with no sign of hidden globe rupture, management might change according to the posterior segment findings, and could vary from observation to a surgical approach. All those changes and their management will be addressed in the following chapters.

Fig. 1.5 Intraorbital foreign body, recognized as a wood stick (courtesy of Allyne Kallaf MD)

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Ocular Trauma Treatment Guideline

2

Ignatz Rohrbacher

Introduction

The classification of ocular trauma is fundamental for the communication between ophthalmologists and better management of the patient. This chapter will be guided by the Birmingham Eye Trauma Terminology System (BETTS), adopted by the International Society of Ocular Trauma (Kuhn et al. 1996) (Table 2.1). The visual prognosis can be estimated through a score created from the analysis of a broad database called Ocular Trauma Score (Unver et al. 2009; Kuhn et al. 2002) (Tables 2.2 and 2.3).

Management

The time and intervention strategy should be evaluated individually. Some authors advocate aggressive early surgical and drug intervention, while others opt for longer intervals.

Table 2.1 BETTS classification of ocular trauma

Closed-globe injury	
Contusion	There is no full-thickness wound
Lamellar laceration	Partial thickness wound of the ocular wall
Open-globe injury	
Laceration	Full-thickness wound by a sharp object
• Penetrating injury	Entrance wound
• IOFB	Intraocular foreign body
• Perforating injury	Entrance *and* exit wounds
Rupture	Full-thickness wound by a blunt object

BETTS Birmingham eye trauma terminology system, *IOFB* intraocular foreign body

Table 2.2 The ocular trauma score

Initial vision		
No light perception	=	60
Light perception	=	70
Light perception—<20/200	=	80
20/200–20/50	=	90
>20/40	=	100
Rupture	=	−23
Endophthalmitis	=	−17
Perforating injury	=	−14
Retinal detachment	=	−11
Relative aferente pupillary defect	=	−10

Table 2.3 Convert total points into % chance of vision outcomes

SUM	OTS	NPL (%)	LP (%)	LP—<20/200 (%)	20-200–20/50 (%)	>20/40 (%)
0–44	1	74	15	7	3	1
45–65	2	27	26	18	15	15
66–80	3	2	11	15	31	41
81–91	4	1	2	3	22	73
92–100	5	0	1	1	5	94

OTS ocular trauma score, *NPL* no light perception, *LP* light perception

Closed-Globe Injury

Hyphema caused by blunt trauma should be treated with the aim of avoiding corneal staining, increased intraocular pressure (IOP) and its consequences (Kearns 1991). Measurements such as shielding the injured eye, use of atropine, topical corticosteroids, and antifibrinolytic agents showed no significant change in final visual acuity but should be considered individually (Gharaibeh et al. 2013). Empirical criteria for surgical intervention may be adopted, such as the persistence of IOP >25 mmHg with total hyphema for more than 5 days, IOP >60 mmHg for more than 2 days in patients without sickle cell anemia, corneal bloodstaining, and persistent hyphema occupying more than half of the anterior chamber (Deutsch et al. 1984). The surgical technique should be the most familiar to the surgeon.

I. Rohrbacher (✉)
Department of Ophthalmology, Governador Celso Ramos Hospital, Florianópolis, SC, Brazil

E. B. Rodrigues et al. (eds.), *Trauma and Miscellaneous Disorders in Retina*, Retina Atlas, https://doi.org/10.1007/978-981-13-8550-6_2

The treatment of subluxation and dislocation of the lens is indicated in the case of cataract formation with visual impairment, possibility of amblyopia in children, pupillary block glaucoma, lens-cornea touch, and uveitis of difficult control. Cases with minimal lens instability can be treated with traditional phacoemulsification technique and may use capsular stabilization hooks and/or capsular tension ring. More complex cases should be treated with pars plana vitrectomy and lensectomy (Kazemi et al. 2000). Vitreous hemorrhage in blunt trauma without retinal lesion should be observed and monitored weekly with indirect ophthalmoscopy and ultrasonography.

Impact energy on blunt trauma can cause retinal lesions, such as tears, dialysis, and macular hole (Cox et al. 1966). Small lesions can be treated prophylactically with cryopexy or photocoagulation to prevent retinal detachment (Schachat et al. 2018).

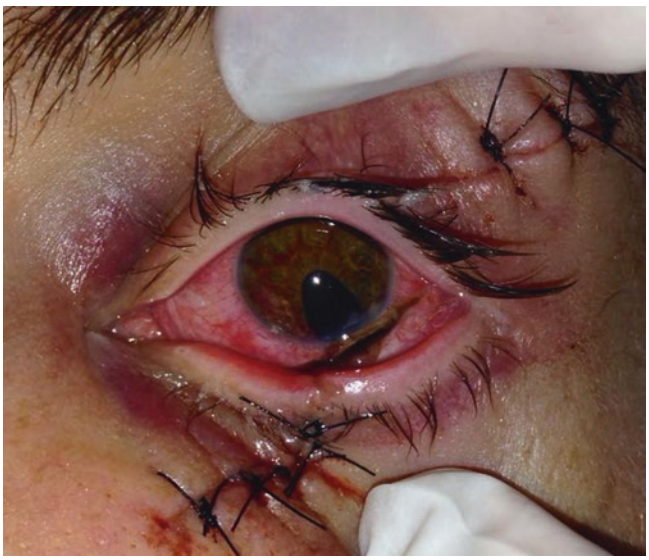

Fig. 2.1 Penetrating trauma of sclera. Presence of iris drop signal. Prolapse of uveal tissue and loss of vitreous by the wound

Open-Globe Injury

Initial care usually occurs in emergency rooms and should focus on life-threatening injuries before attention is focused on eye trauma. The eye should be protected with a shield, avoidance of any manipulation, toxicology screen, pain medication, antiemetic, broad-spectrum systemic antibiotics with high intravitreal penetration as vancomycin (15 mg/kg, maximum dose 1.5 g) and cetfazidime (50 mg/kg, maximum dose 2 g), administer tetanus toxoid, and perform non-contrast computer tomography of head and orbits to detect foreign body and any deformation of the globe (Schachat et al. 2018; Al-Omran et al. 2007; Ahmed et al. 2012). The use of intravenous antibiotics for 48 h in services with ocular trauma care was related to a rate of 0.9% of endophthalmitis (Andreoli et al. 2009; Faghihi et al. 2012). Magnetic resonance imaging is indicated for cases with suspected IOFB of wood or plastic. Ultrasonography can be performed delicately and with the eyelids closed to prevent prolapse of ocular contents, but this examination depends on the operator's ability to be satisfactory in the investigation (Kramer et al. 1995).

The treatment of wall lacerations should be performed with the objective of restoring the integrity of the eyeball and intraocular pressure, preserving the visual axis as much as possible. Corneal lesions reaching the limbus or sclera with indeterminate extent should be carefully explored through peritomy and dissection of the Tenon's capsule to determine size, involvement of other structures, and vitreous incarceration (Fig. 2.1). Sutures for eyelid retraction may be helpful in improving exposure and avoiding increased intraocular pressure. Corneal lacerations can be treated with single interrupted sutures using nylon 10-0 and with 2/3 depth of the corneal stroma. Other options are purse-string closure, tissue adhesive, and corneal patch. Prolapsed uveal tissue should be removed only when clearly necrotic or when exposed for more than 24 h. Scleral lacerations can be repaired with non-absorbable sutures 7-0. Incarcerated vitreous should be excised with sharp scissors, Vannas scissors, to avoid future vitreoretinal traction (Schachat et al. 2018; Colby 1999).

IOFB should be removed as soon as the patient is clinically stable and has access to a referral center in order to reduce the chances of endophthalmitis and inflammatory lesions caused by iron or copper. The IOFB can be removed using magnets, when metallic, or intraocular forceps. The use of forceps via pars plana presents better control of the situation and is the most available method for the procedure. Important remember to enlarge the sclerotomy to remove the IOFB and remove the posterior hyaloid during vitrectomy to avoid the formation of epiretinal membrane or vitreoretinal tractions (Slusher et al. 1982). In order to avoid future retinal detachments or traction, prophylactic encircling scleral buckles in all eyes receiving vitrectomy for management of penetrating injuries can be done (Schachat et al. 2018).

Patients with perforating trauma should undergo vitrectomy as early as possible to avoid fibrovascular proliferation. The anterior rupture should be readily sutured, while the posterior rupture becomes self-sealed around 7 days after the trauma (Schachat et al. 2018). Vitrectomy is only possible when the wounds are sealed, so it should be performed from 7 days of trauma. Some authors indicate prophylactic encircling scleral buckles in all eyes (Schachat et al. 2018). Another possible approach is to perform the chorioretinectomy, where the uveal and retinal tissue around the ruptures are destroyed with vitrector and diathermy (Weichel et al.

2010). The correct time to perform vitrectomy is still a controversial issue, but there is a consensus that it should be performed up to 14 days to reduce the risk of sympathetic ophthalmia and membrane formation (Winthrop et al. 1980).

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Choroidal Folds

3

Müller Gonçalves Urias

Introduction

The fundus image of "peculiar lines in the choroid"—as Nettleship first described in 1884 (Squirrell 2006)—the so-called choroidal folds (Fig. 3.1) are anatomically undulations in the retinal pigment epithelium (RPE), Bruch's membrane and the inner portion of the choriocapillaris—which may or may not be involved. If retina is involved, it is called chorioretinal folds. It differs from retinal folds since with retinal folds only neurosensory retina is involved and have a different presentation (Jaworski et al. 1999; Murdoch and Merriman 2002; Musetti et al. 2014).

Choroidal folds might be unilateral or bilateral, it is commonly related to intraorbital tumors, but it could be related to other several causes. For example, after an ocular injury, choroidal folds could be present-either as a result of the injury *per se* or as a consequence to the trauma, mainly associated with hypotony and inflammation (Squirrell 2006).

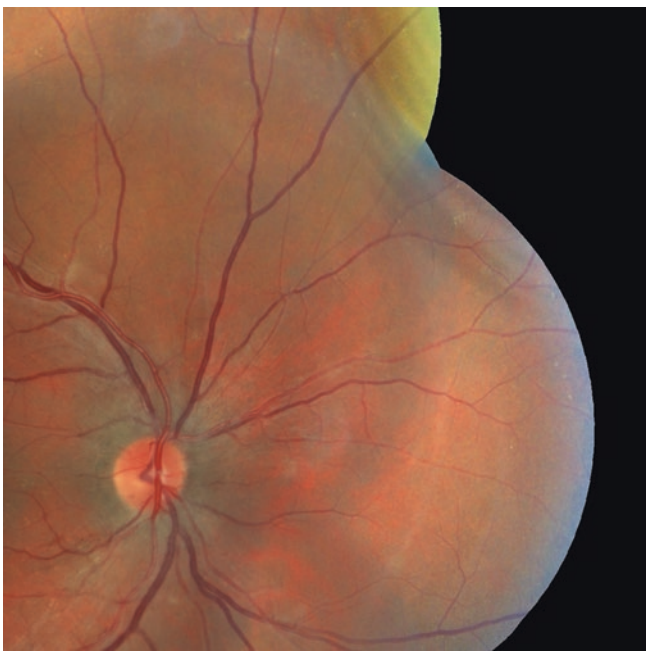

Fig. 3.1 Detail of a fundus composition of the right eye. Partial visualization of peripheral choroidal folds in a patient with intraorbital foreign body

Etiopathogenesis

The explanation of those folds is the excessive potential surface area of the choroid for the space that it must occupy, and it is commonly associated with either scleral shortening or choroidal congestion (Newell 1973; Bullock and Egbert 1974; Friberg 1989; Jaworski et al. 1999).

Regarding to ocular trauma, hypotony could be a result from an open-globe injury, from a hidden-globe rupture, or even from a ciliary body detachment. The hypotony, then, could present associated with choroidal folds. Other causes of trauma-related choroidal folds could be a persistent globe flattening after a blunt injury examined timely, an intraorbital but yet extraocular foreign body, and also inflammation of extraocular tissues (Lima-Gomez and Cornejo-Mendoza 2004; Ding and Zeng 2012).

Clinical Features

Patients with choroidal folds may present a sorts of symptoms. From no-symptoms to metamorphopsia, and even decreased visual acuity might from a hyperopia shift (Murdoch and Merriman 2002) might be present, so a detailed examination is necessary. Moreover, posterior biomicroscopy on a slit-lamp with retro-illumination on the area is the best way to observe the contrast between the ripples.

M. G. Urias (✉)
Department of Ophthalmology, Federal University of São Paulo, São Paulo, Brazil
e-mail: urias@unifesp.br

E. B. Rodrigues et al. (eds.), *Trauma and Miscellaneous Disorders in Retina*, Retina Atlas, https://doi.org/10.1007/978-981-13-8550-6_3

With this technique, the lighter areas are believed to be the stretched RPE and the darker areas, the concentrated RPE cells (Squirrell 2006).

Choroidal folds usually are present temporal to optic disc and rarely beyond the posterior pole. They could present in five varieties of patterns: horizontal, oblique, vertical, radial, or irregular. A unilateral or a bilateral occurrence and the fold pattern could suggest their cause and, then, their prognosis. Considering a posterior ocular trauma, irregular choroidal folds could be found—usually related to hypotony; radial folds could exist and help identify an abnormal intraorbital mass-effect compression; horizontal choroidal folds are characteristic from idiopathic cases (Newell 1973).

The folds are verified through the clinical exam, but ancillary exams might improve diagnostic accuracy. On fluorescein angiography (FA) (Fig. 3.2), there are hyperfluorescent

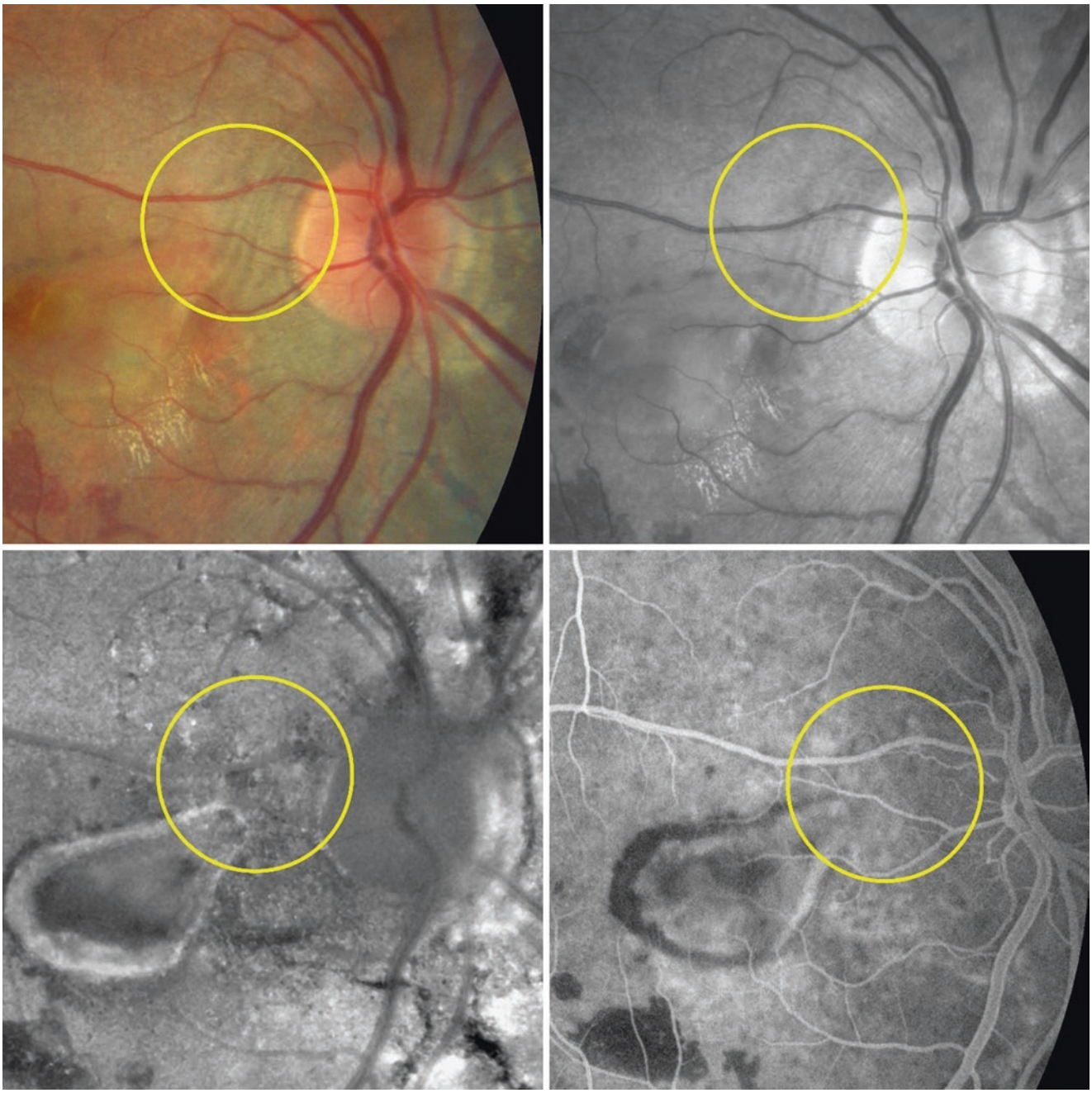

Fig. 3.2 Image composition of choroidal folds in a patient with history neovascular membrane associated with angioid streaks. Upper left: color fundus photo; upper right: infrared image; bottom left: autofluorescence image with almost no visualization of choroidal folds; bottom right: fluorescein angiography also with no visualization of choroidal folds. Anatomy aspect of RPE and retinal changes may affect the visualization of choroidal folds

lines—peaks—and hypofluorescent lines—valleys—visible from early stages until later stages without leakage or impregnation. This characteristic is also present on autofluorescence but in an inverted peak-valley pattern, according to the concentration of RPE cells and their lipofuscin pigment. Indocyanine green angiography shows a similar pattern of FA, and altought less accurate, might be helpful on excluding some differential diagnosis (Squirrell 2006). Optical coherence tomography (OCT) could especially identify when not only choroid but retina also folds, with a swept source OCT or an enhanced depth imaging useful in those cases (Shields et al. 2014). OCT angiography (OCTa) could be used to document those cases and be interesting for follow-up. However, the professional should be careful once chorioretinal folds usually interferes with the automated segmentation of external retinal layers and choroid. So, altought they might be initially present as a crackled image on an OCTa slab, probably they have segmentation issues that after manual correction might result in a different slab image. Moreover, as these folds are so evident that they could be identified on a fundus image, a fundus photo could be enough for follow-up. Ocular ultrasound imaging could be useful in verifying underlying causes of folds, such as ciliary body detachment, increased subarachnoid space, or choroidal tumors (Atta and Byrne 1988).

Management

The choroidal fold treatment is, when necessary, usually treat the fold cause. With regards to ocular trauma-related folds, it is essential for the ophthalmologist to rule out open-globe injuries, hidden-globe ruptures, and ciliary body detachments. It is also important to verify if there is no extraocular compression and treat trauma-related inflammation. After ruled out all differential diagnosis, management of idiopathic choroidal folds could be observed, without any medical treatment required (Squirrell 2006).

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Choroidal Rupture

4

Felipe Pereira and Gabriel Costa Andrade

Introduction

Choroidal rupture consists in the rupture of the retinal pigment epithelium (RPE), Bruch's membrane, and choroid. The main cause of choroidal rupture is blunt trauma, where up to 10% of individuals suffering from this type of trauma may present with this association (Williams et al. 1990).

Etiopathogenesis

Choroidal rupture can be classified as indirect or direct. Indirect choroidal rupture accounts for more than 80% of cases and is caused by a contrecoup mechanism, where the rupture occurs far from the impact site, i.e., in the posterior pole region. The most accepted theory is that after trauma there is a deformation of the eyeball in the anteroposterior direction with a force distribution from the optic nerve in concentric form. Since the retina has certain elasticity and the sclera shows great resistance, the region that suffers most from the deformation of the eyeball is the choroid, especially the more superficial region that is close to Bruch's membrane (Aguilar and Green 1984).

Direct choroidal rupture occurs near the trauma site and is usually oriented parallel to the ora serrata. The theory behind this type of rupture, for which there is no consensus among specialists, consists in triggering an inflammatory process with necrosis at the site of the trauma that eventually causes the defect in the choroid (Osterberg 1936).

Patients with angioid streaks deserve special attention because they have a naturally fragile Bruch's membrane and may have choroidal rupture after minimal trauma or even multiple ruptures (de Benedetto et al. 2012).

Clinical Findings

Choroidal rupture is more common in young men, with up to 80% male predominance, consistent with trauma epidemiology in general (Secretan et al. 1998).

Initial visual acuity may range from 20/20 to light perception. This variation occurs due to differences in the location of rupture (macular vs. peripheral), type of trauma that caused rupture (sport with ball vs. explosion with eyeball involvement) and due to some findings that may be associated with subretinal hemorrhage, vitreous hemorrhage, and retinal detachment (Ament et al. 2006).

The classic finding of choroidal rupture is a yellowish-white line, crescent-shaped, and concentric to the optic disc. The most common site is in the temporal region to the disc, in the posterior pole (Fig. 4.1). Direct trauma ruptures tend

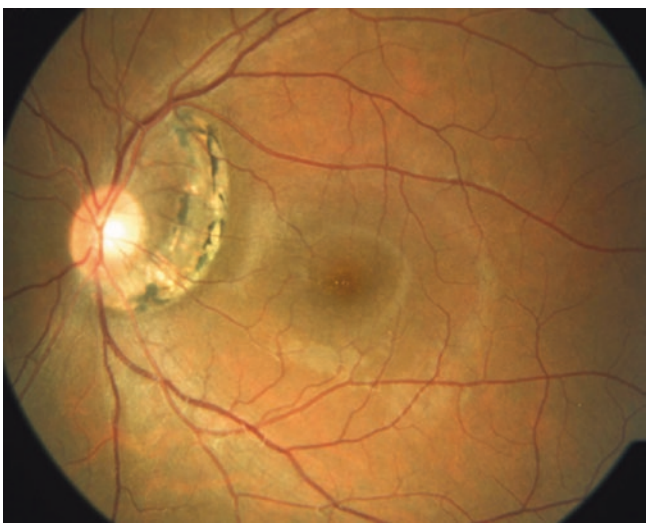

Fig. 4.1 Color fundus photograph shows choroidal rupture with crescent-shaped lesion concentric to the disc. Special thanks to Gabriel Costa de Andrade MD

F. Pereira (✉) · G. C. Andrade
Department of Ophthalmology, São Paulo Federal University, São Paulo, Brazil

E. B. Rodrigues et al. (eds.), *Trauma and Miscellaneous Disorders in Retina*, Retina Atlas, https://doi.org/10.1007/978-981-13-8550-6_4

to be more in the periphery and parallel to the ora serrata. Often there is association with subretinal bleeding due to the rupture of the choriocapillaris layer, which may make it difficult to observe the classic findings (Patel et al. 2013). After partial absorption of blood and deoxygenation of hemoglobin, there may be a yellowish subretinal lesion that should not be confused with vitelliform material (Gross and Freeman 1990).

The series published by Ament et al. (2006) collected 111 cases of choroidal rupture and found 68% of the ruptures occurring in the posterior pole, with 37% being in the foveal region and 31% in the extrafoveal region. The rest, 32%, were in a region outside the retinal vascular arcades. Approximately 61% of the patients in this series had only one rupture, 21% had two ruptures, 11% had three ruptures, and 7% had four or more ruptures.

Complementary Examinations

Autofluorescence

Autofluorescence imaging is able to detect the fluorescence emitted by lipofuscin, which is stored in the RPE. In choroidal rupture, the examination shows hypoautofluorescence in the rupture area due to the loss of RPE and the blockage that is caused in cases in which there is associated hemorrhage (Fig. 4.2). A hyperautofluorescent ring corresponding to RPE hyperplasia can be found at the edges of the rupture (Lavinsky et al. 2011; Shin et al. 2017).

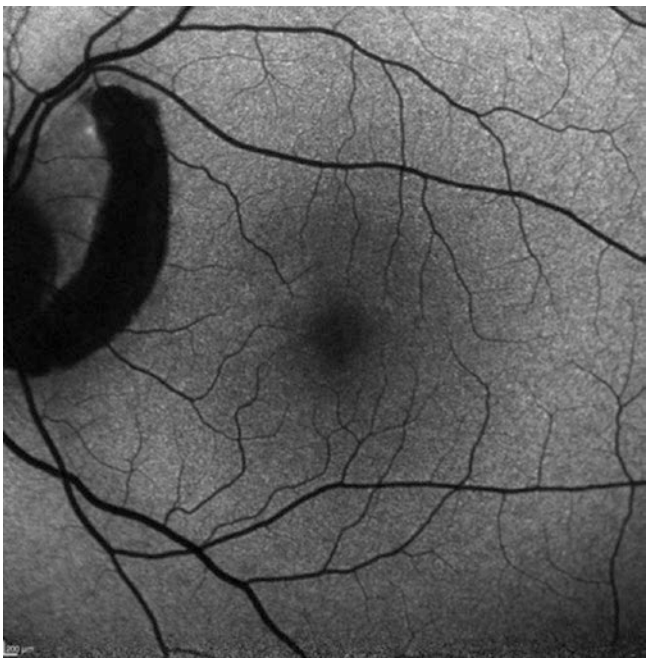

Fig. 4.2 Fundus autofluorescence demonstrates the hypoautofluorescence in the choroidal rupture lesion. In this case there is no hyperautofluorescence ring around the lesion. Special thanks to Gabriel Costa de Andrade MD

Fluorescein Angiography

Fluorescein angiography may be useful in cases of choroidal rupture as it may assist in the identification of rupture in areas showing bleeding, retinal edema, or other RPE abnormalities. The typical finding is hypofluorescence in the early phases of the examination, which becomes gradually hyperfluorescence due to the leakage of the choriocapillaris causing impregnation of the rupture margins (Shakin and Yannuzzi 1987).

Indocyanine Green Angiography

Indocyanine green classically shows a pattern of hypofluorescence at all stages of the examination. Thanks to the longer wavelength of the emitted light and less leakage of the contrast by the choroidal vessels, this examination has even greater sensitivity than fluorescein angiography and clinical examination to identify areas of choroidal rupture (Kohno et al. 2000).

Optical Coherence Tomography (OCT)

OCT findings in patients with choroidal rupture may include two different types. In type 1, we identified a dome-shaped protrusion of the RPE/choriocapillary complex toward the innermost layers of the retina, and possibly associated with RPE disruption. In type 2, there was a greater area of disruption of the RPE and choriocapillaris, besides a compromise of the photoreceptor layer and external limiting membrane, which was associated with a depression toward the choroid with a drop in tissues that are just above this defect in the concavity (Fig. 4.3) (Nair et al. 2013).

Choroidal Neovascularization (CNV)

CNV is a rare complication of choroidal rupture but shows serious implications in the visual prognosis of these patients. Soon after rupture formation, an inflammatory process occurs, with an increase in the concentration of vascular endothelial growth factor (VEGF) (Kim et al. 2015). If these factors persist long enough, neovessel formation may occur below the RPE (type 1 membrane) or they may gain subretinal space by rupturing the Bruch's membrane and RPE membrane (type 2 membrane).

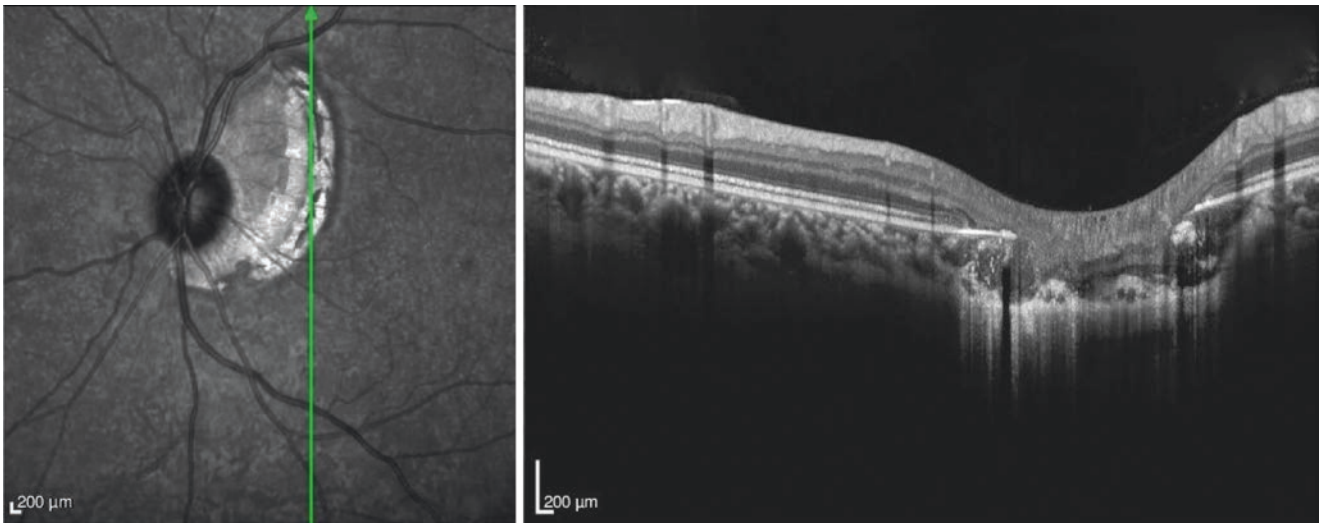

Fig. 4.3 Optical coherence tomography vertical scan showing type 2 choroidal rupture with extensive loss of RPE and choriocapillary complex and loss of photoreceptor layer. There is also a depression of retinal tissue toward the space created by the choroidal defect. Special thanks to Gabriel Costa de Andrade MD

The rate of CNV in patients with choroidal rupture ranges from 10 to 20% of cases and in 80% of cases they appear in the first year of follow-up, but may appear at any time after trauma (Secretan et al. 1998; Ament et al. 2006).

Clinically, the appearance of CNV occurs with sudden worsening of visual acuity or metamorphopsia, and in the examination we can identify new bleeding near the choroidal rupture with or without associated retinal edema. Fluorescein angiography demonstrates occult or classic leakage depending on where the membrane is located, below or above the RPE, respectively. OCT also helps in the diagnosis by identifying the presence of new hyperreflective material in the outer retina and facilitates the identification of intra- and subretinal fluid, which is of essential importance in the follow-up of these cases.

The onset of CNV after choroidal rupture is more common in those patients with long and macular ruptures as well as in older patients. There is no positive association between the onset of CNV and the width of the rupture or the number of ruptures (Ament et al. 2006).

Evolution

During the follow-up of these patients, ruptures may show the fibrosis and RPE hyperplasia at the margins. Another possible evolution may be the partial or complete closure of the rupture, which occurs more frequently in cases where there is no compromise of the deeper layers of the choroid or disruption of all layers of the retina above the rupture (Shin et al. 2017).

Patients with choroidal rupture usually do not evolve with good visual acuity. Overall, only 35% of patients recover visual acuity to 20/40 or better. In the subgroup of patients with macular rupture, this recovery is 22%, and in those with peripheral rupture, the rate is 60%. Patients who evolve with neovascular membrane have an even worse prognosis, with only 8% reaching a visual acuity of 20/40 or better (Ament et al. 2006).

Treatment

Patients with choroidal rupture without associated complications can only be observed. Follow-up should be done to identify the presence of CNV early, which should be done more frequently in the first year and then be more spaced out. The study can be done through fundoscopic examination and OCT, and can be confirmed with angiography if necessary (Patel et al. 2013).

Cases involving a large amount of sub-macular blood can be treated by intravitreal injection of an expandable gas for blood displacement. However, there are no studies that prove the true benefit of this action in relation to a natural course, so this approach should be taken with caution (Fig. 4.4) (Goldman et al. 2014).

When the patient presents with retinal detachment associated with choroidal rupture, the treatment of choice will depend on the location of the rupture and the presence or not of retinal dialysis. Another observation should be made regarding the lens of these patients, since the trauma may have caused traumatic cataract or subluxation. A combination of various techniques such as pars plana vitrectomy, conventional retinopexy, lensectomy, silicone oil, or gas implant should be part of the surgeon's arsenal when dealing with complex post-trauma cases (Mahapatra et al. 2018).

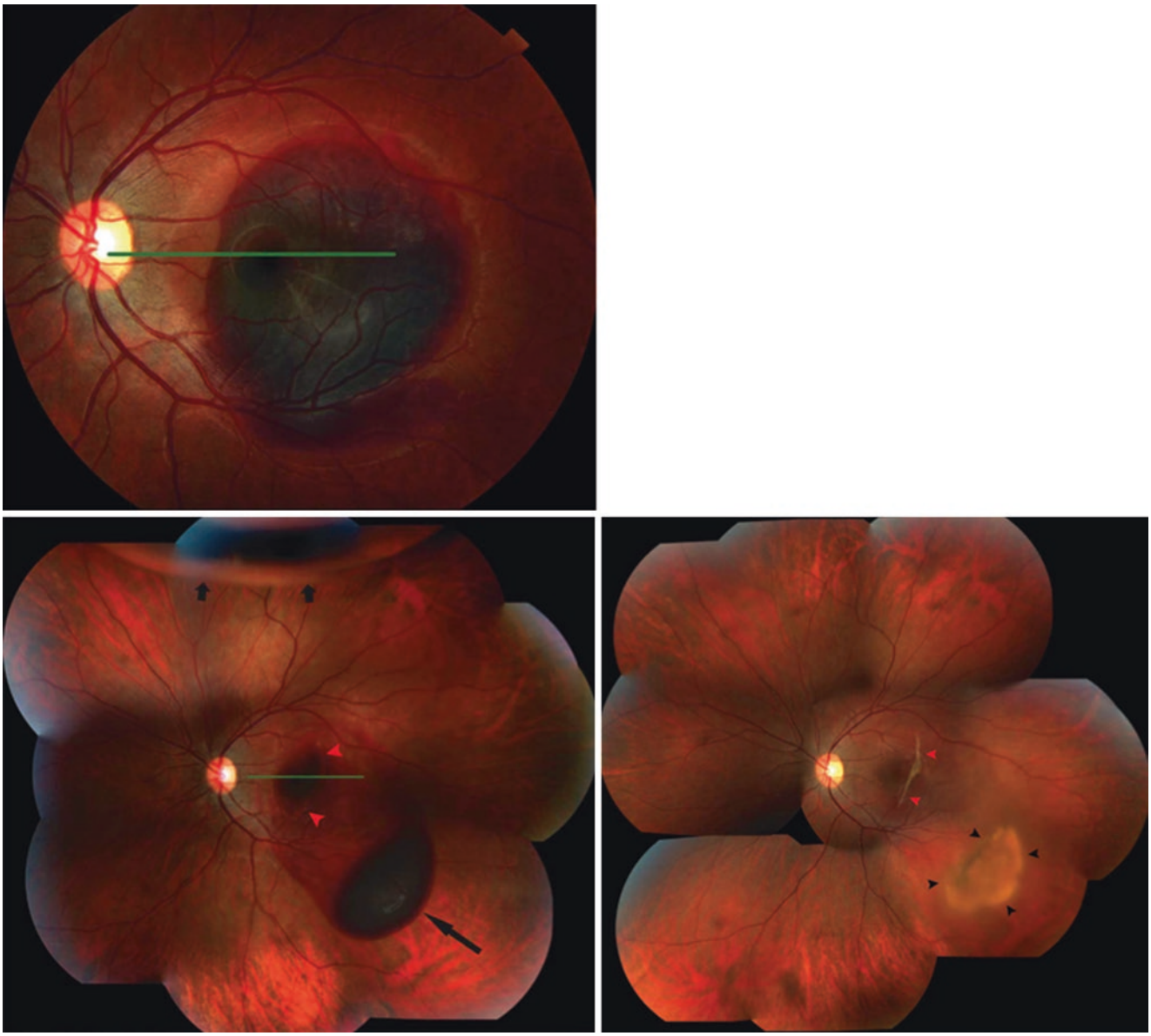

Fig. 4.4 Top image with extensive sub-macular hemorrhage. Bottom left image with 5 days after injection of 0.4 mL of 100% SF6. Bottom right with 6 weeks of presentation. It is possible to see the displacement of sub-macular hemorrhage. Final visual acuity was 20/50. Reproduced with permission of Elias Reichel MD from Goldman et al. (2014)

The treatment of CNV can be done with laser photocoagulation in cases in which the rupture and CNV are peripheral. In cases of rupture and macular or foveal CNV, the treatment of choice is anti-VEGF agents, since they are able to decrease vascular permeability, promote neovessels closure, and stimulate RPE. There are several case reports demonstrating improvement of visual acuity and decrease in intra- and subretinal fluid with stabilization of CNV after one injection and without the need for new applications in the vast majority of cases. Even with treatment and subsequent improvement of visual acuity, patients who developed CNV, especially in the macular region, still have inferior vision compared to those who did not develop CNV (Ozgur et al. 2009; Preziosa et al. 2018).

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Commotio Retinae

5

Felipe Muralha

Introduction

A commotio retina is a transient retinal lesion following direct ocular injury. It was first described by Berlin (1873) as whitening in the retina after a blunt trauma to the globe (Figs. 5.1, 5.2 and 5.3), usually appearing in the opposite site of the trauma (contrecoup injury).

Etiopathogenesis

When Berlin described the injury, he believed the trauma caused an extracellular edema, resulting in loss of retinal transparency, since then several studies contributed to elucidate the disease's pathogenesis.

Fluorescein angiography in animal models with commotio retinae lesions showed no leakage (François and De Laey 1970; Hart and Frank 1975; Gregor and Ryan 1982). Vitreous fluorophotometer analysis, a high sensitivity exam to detect leakage through the blood–retina barrier, did not show any difference between traumatized and non-traumatized eyes (Pulido and Blair 1987). In addition, immunohistochemical analysis showed minimal amount of albumin after trauma in the eye (Mansour et al. 1992). Collectively, these factors helped to prove that the blood–retina barrier is relatively intact and is not a major component in the disease's pathogenesis.

Histopathological studies in commotion retinae revealed the presence of extracellular and intracellular edema and photoreceptor outer segment disruption. Predilection for damage to the photoreceptor layer in commotion retinae may occur because of the absence of Muller cells support (Mansour et al. 1992; Hart and Frank 1975; Blight and Hart 1978; Sipperley et al. 1978), while some studies also showed additional damage in the retina pigment epithelium (RPE) cells (Mansour et al. 1992).

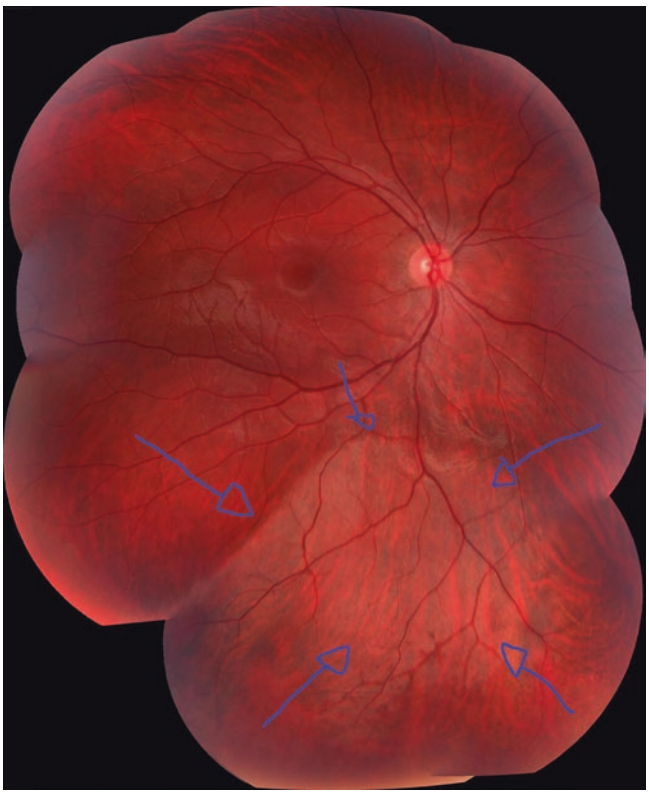

Fig. 5.1 Color fundus photography composite from the right eye of an 18-year-old male with a commotio retinae lesion after a traumatic injury from a volleyball. There's a whitening of the inferior retina. Courtesy of Dr. Renato Rosário

Clinical Features

Commotio retinae appearance on fundoscopic exam is a retinal pallor some hours after a traumatic injury to the globe, resulting in a gray–white opacification established predominantly in the outer retina (Mansour et al. 1992), with occasional RPE mottling. The size, color, and limits of the lesion will vary according to the severity of the trauma. The lesions

F. Muralha (✉)
Department of Ophthalmology, Federal University of São Paulo, São Paulo, Brazil

E. B. Rodrigues et al. (eds.), *Trauma and Miscellaneous Disorders in Retina*, Retina Atlas, https://doi.org/10.1007/978-981-13-8550-6_5

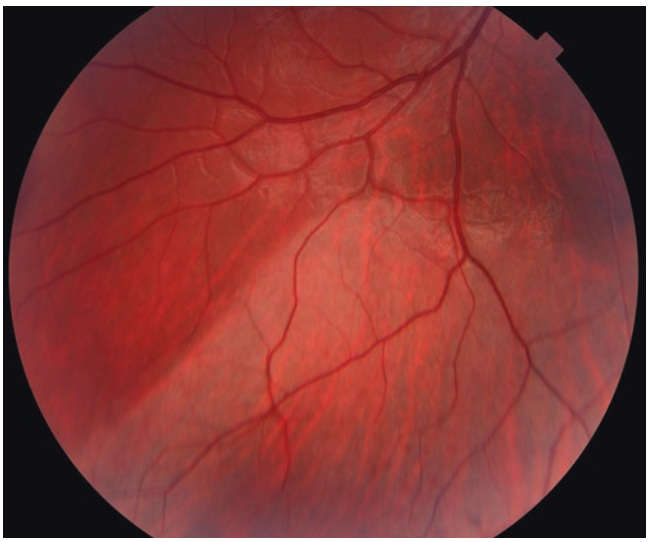

Fig. 5.2 Color fundus photography of the same patient focusing in the gray–white opacification of the retina's periphery. Courtesy of Dr. Renato Rosário

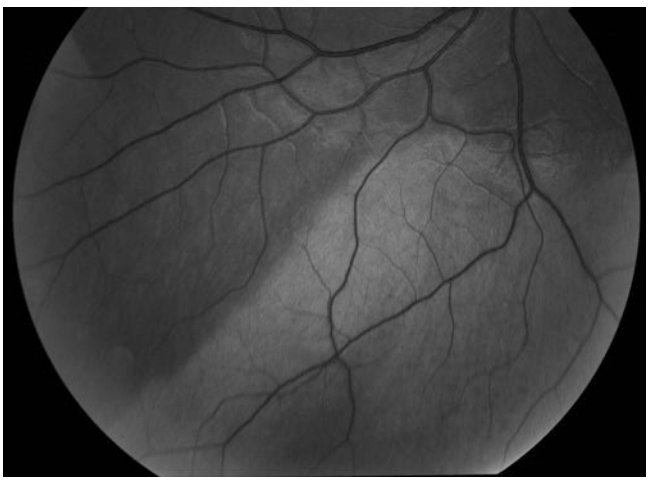

Fig. 5.3 Red-free image of the same lesion showing more precise limits. Courtesy of Dr. Renato Rosário

can happen in the periphery or involve the posterior pole, in which case they are referred as Berlin's edema. When it happens in the posterior pole, the fovea is often spared, resulting in a cherry-red spot. With macular involvement the visual acuity could be affected, but it usually goes back to normal after the opacification resolution. The lesions are transitory, normally disappearing spontaneously in days or weeks (Hart and Frank 1975); however, in some severe cases RPE mottling (Hart and Frank 1975) or intraretinal pigment deposits can happen (Crouch and Apple 1974), and a persistent visual impairment can result from cases with foveal involvement (Figs. 5.4 and 5.5).

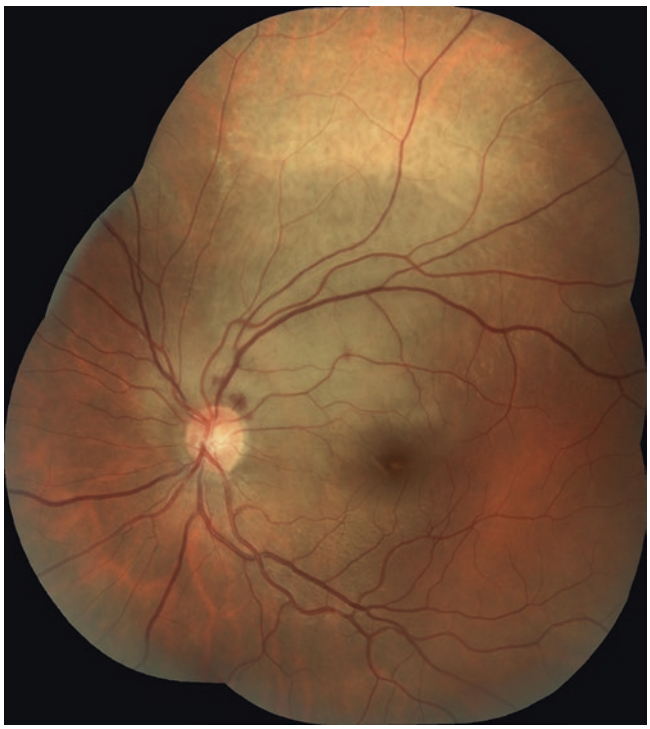

Fig. 5.4 Color fundus photography composite from the left eye of a 28-year-old male after a soccer ball trauma to the globe. There is a large lesion involving the macula and extending upward with some hemorrhages. Courtesy of Dr. Franklin Souza-Santos

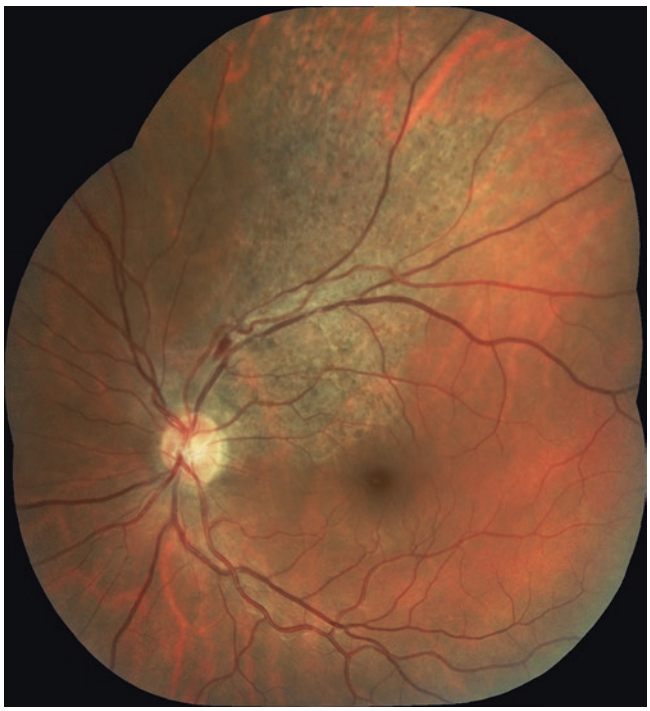

Fig. 5.5 Color fundus photography composite from the same patient 8 weeks later, the commotio retinae area evolved with RPE mottling. Courtesy of Dr. Franklin Souza-Santos

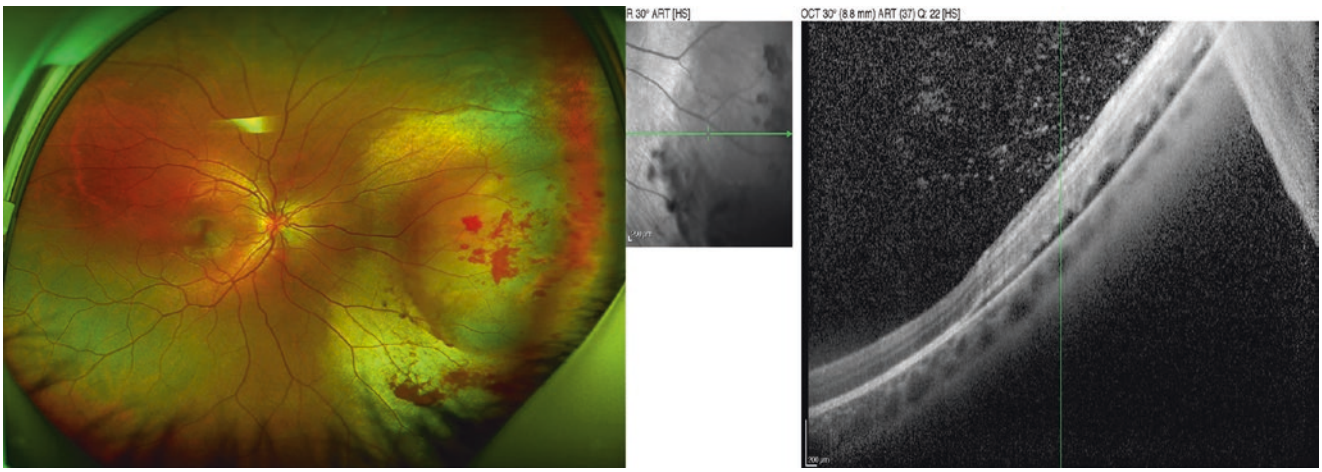

Fig. 5.6 Color fundus photography and the respective OCT of commotio retinae. Special thanks to Gabriel Andrande from BRAVS

Management

The commotio retinae prognosis is good, as most lesions disappear and, in most cases, there is no visual impairment (Eagling 1974). Because it has a self-limited nature, there is no approved or commonly used medical treatment for the entity. Most cases will evolve to complete resolution in 3–4 weeks, although some improvement can continue for up to 6 months (Eagling 1974). Some patients, however, may develop relative or absolute scotomas, or even permanent visual acuity impairment. About 26% of patients with macular involvement will have visual acuity worse than 20/30 permanently (Blanch et al. 2013). Other lesions as choroidal rupture and macular hole can happen in some cases due to the traumatic injury, limiting visual recovery. Optical coherence tomography imaging can help in predicting visual prognosis, an acute loss or attenuation of the ellipsoid layer and hyperreflectivity of the overlying retina are associated with pigment disturbance, retinal atrophy, and poor visual outcome (Fig. 5.6) (Souza-Santos et al. 2012).

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Valsalva Retinopathy

6

Nathalia Corbelli Roberti
and Eduardo Büchele Rodrigues

Introduction

Valsalva retinopathy was firstly described in 1972 by Thomas Duane as a particular form of retinopathy characterized by preretinal bleeding secondary to a sudden increase in intrathoracic pressure (Duane 1972, 1973). Hemorrhage typically occurs in the macular region (Fig. 6.1) and may resolve spontaneously within a few days or take months for complete reabsorption (Gass 1987). It usually affects young adults causing unilateral low visual acuity sudden and painless. Cases of bilateral involvement have been described (Durukan et al. 2008). The prolonged period of contact between the blood and the retina may lead to complications with risk of persistence of impaired visual acuity (Nili AhmadAbadi et al. 2009).

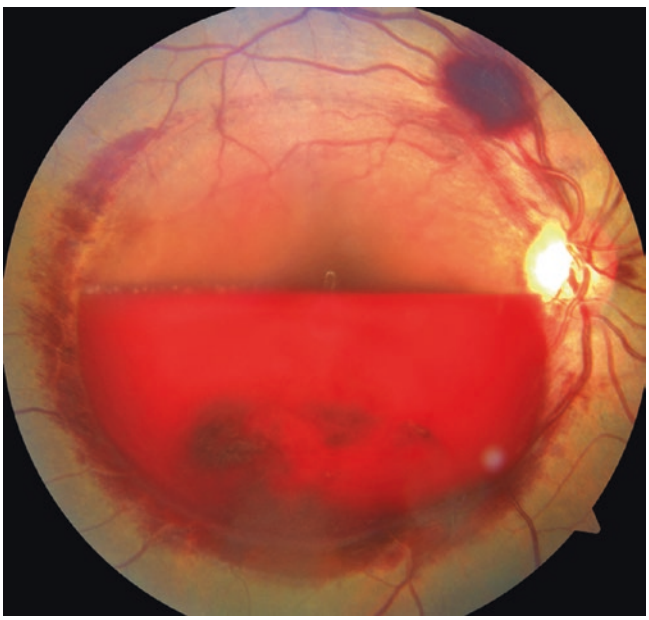

Fig. 6.1 Valsalva retinopathy. Color fundus photograph shows premacular hemorrhage

Etiopathogenesis

The valsalva maneuver occurs with an abrupt increase in intrathoracic or intra-abdominal pressure transmitted directly to intraocular veins. Incompetent valves or their absence in the venous system allows such an unexpected increase in the pressure that causes a decompensation in the retinal capillary bed. The rupture of the perifoveal capillaries leads to sublimitant internal hemorrhage, which may become subhyaloid or intravitreal (Duane 1972, 1973). The accumulation of blood between the posterior hyaloid and the inner limiting membrane (ILM) is responsible for the low visual acuity, which may occur less frequently between the ILM and the nerve fiber layer (Mennel 2007; Meyer et al. 2006). The macular region is the most affected due to the low adhesion of the internal limiting membrane in this region (De Maeyer et al. 2007). Valsalva retinopathy has been reported to be associated with cough, vomiting (Herr et al. 2004), pregnancy (Jayaprakasam et al. 2011; Li et al. 2018), labor (Ladjimi et al. 2002), intense physical activity (Sueke 2009), and compressive injuries (Chandra et al. 2005).

Clinical Features

The diagnosis is made by fundoscopy, showing sharply demarcated premacular dome-shaped hemorrhages (Fig. 6.2). Clinically, it is difficult to distinguish between

N. C. Roberti (✉)
Department of Ophthalmology, São Paulo Federal University, São Paulo, Brazil
e-mail: contato@dranathaliacorbelli.com.br

E. B. Rodrigues
Department of Ophthalmology and Visual Sciences, Federal University of São Paulo, São Paulo, Brazil

Department of Ophthalmology, Saint Louis University School of Medicine, Saint Louis, MO, USA

E. B. Rodrigues et al. (eds.), *Trauma and Miscellaneous Disorders in Retina*, Retina Atlas, https://doi.org/10.1007/978-981-13-8550-6_6

subhyaloid and sublimitant internal hemorrhages. The first one usually occurs in patients who did not have posterior vitreous detachment, while the last one commonly presents in patients with vitreous detachment with glistening reflex and fine striae on the surface of the hemorrhage (De Maeyer et al. 2007; Gibran et al. 2007; Meyer et al. 2006; Shukla et al. 2005; Tatlipinar et al. 2007). Optical coherence tomography (OCT) (Fig. 6.3) may help identify the location by assessing the segmentation of preretinal bleeding (Meyer et al. 2006; Shukla et al. 2005; Tatlipinar et al. 2007). Shukla et al. (2005) reported two cases with two membranes of different reflectivity. First, more hyperreflective and above bleeding would correspond to ILM; second, less reflective, irregular, and also above the hemorrhage would correspond to the posterior hyaloid. Hasegawa et al. (2011) described a case of valsalva hemorrhage, in which the posterior hyaloid and ILM were fused in the OCT. The method is susceptible to errors, since recent bleeding may present high reflectivity and impair interpretation (Blaise et al. 2006; De Maeyer et al. 2007). Currently, the most accurate method for such confirmation is vitrectomy with removal of the membrane and subsequent histopathological analysis (Gibran et al. 2007). Nevertheless, we propose six clinical and tomographic findings that may help to identifies sublimitant internal hemorrhages: (1) glistening reflex; (2) fine striae; (3) immobility of the blood; (4) sharply remarqued and dome shaped; (5) young patients; (6) high reflective blood immediately above the hemorrhage.

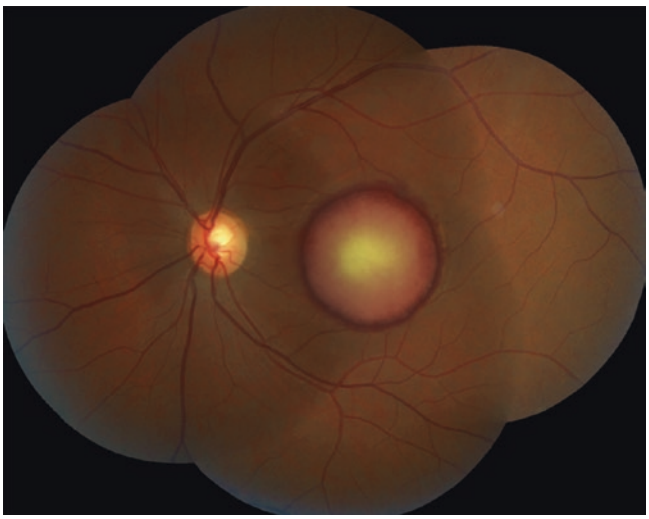

Fig. 6.2 Valsalva retinopathy. Color fundus photograph shows sharply demarcated premacular dome-shaped hemorrhage

Management

Observation

Spontaneous reabsorption of bleeding can occur in a few weeks to several months. In the severe cases, this process may take longer. Retinal toxicity by hemoglobin, epiretinal membrane formation, macular hole, retinal detachment, cat-

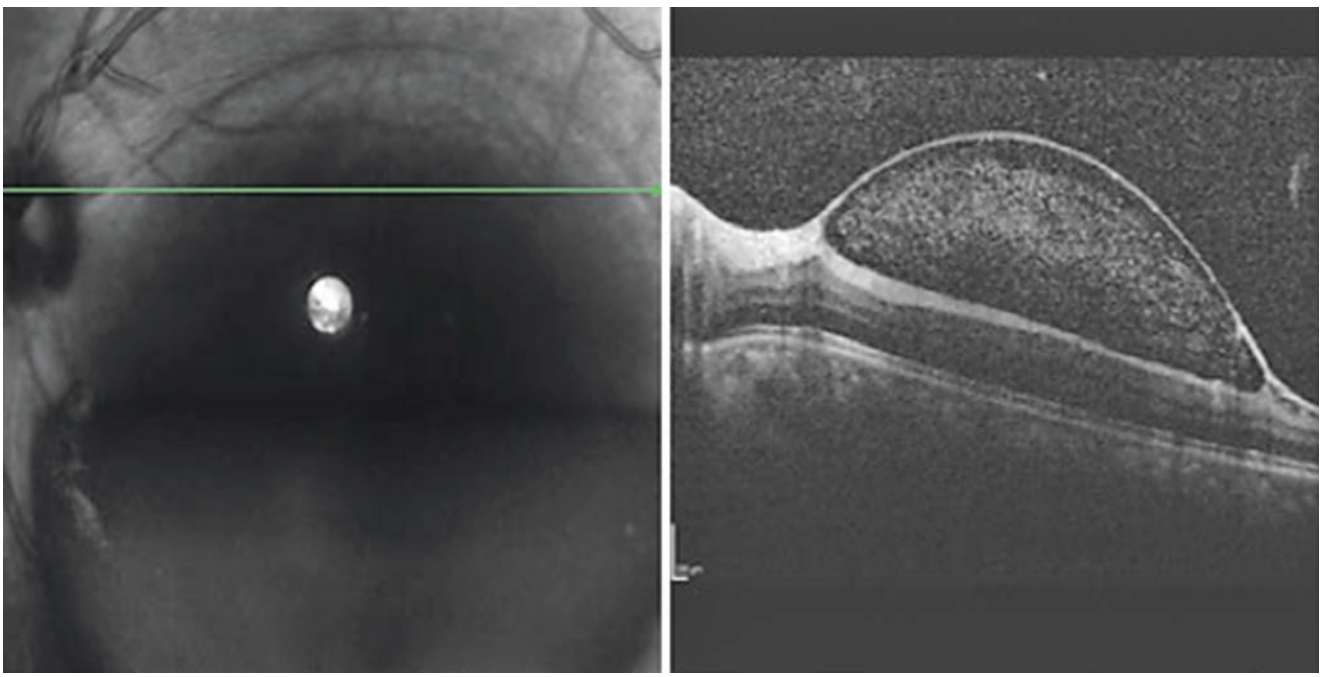

Fig. 6.3 Optical coherence tomography shows pre-internal limiting membrane bleeding in a patient with valsalva retinopathy

aract, glaucoma, and amblyopia in children are some complications that may occur due to the prolonged presence of intraocular blood (De Maeyer et al. 2007; Nili AhmadAbadi et al. 2009; Xie et al. 2014).

Laser Therapy

Neodymium-doped yttrium-aluminum-garnet (Nd:YAG) laser is an alternative treatment to promote immediate drainage of bleeding through the vitreous cavity by rupture of posterior hyaloid or ILM (Fig. 6.4), restoring visual acuity in a short time (Nili AhmadAbadi et al. 2009; Rennie et al. 2001). The best indications are hemorrhages greater than three-disc diameters treated until 3 weeks after bleeding (Durukan et al. 2008). The power of the laser can range from 2.5 to 10.5 mJ, preferably using the smallest amount needed, in order to avoid complications such as macular hole, macular tear, and retinal detachment (Ulbig et al. 1998). Studies with potency up to 50 mJ were performed without causing retinal injury. It is believed that preretinal blood acts as a protector of possible retinal lesions caused by laser energy (Gabel et al. 1989). Other known complications are epiretinal membrane formation and persistent premacular cavity (Goel et al. 2011; Kwok et al. 2003; Ulbig et al. 1998). Some factors limit the performance of this procedure, such as the presence of coagulated premacular blood and the proximity of blood to the retinal surface and to the fovea (García Fernandéz et al. 2012; Li et al. 2018).

Intravitreal Application Therapy

Another therapeutic option includes pneumatic retinopexy with or without plasminogen activating factor (rtPA) which removes hemorrhage from the foveal region. It is believed that the rtPA act by inducing the separation of the posterior vitreous, allowing drainage of the bleeding. In addition, the gas injection would aid in the diffusion of blood from the posterior pole (Hesse et al. 1999; Koh et al. 2000; Park and Seo 2004; Schmitz et al. 2000). This therapeutic modality presents low risk, but the increased chance of retinal ruptures induced by gas cannot be ruled out (Hesse et al. 1999).

Vitreoretinal Surgery

Currently, with the advancement of vitreoretinal surgeries with microincisions, this technique has been increasingly used. It is the treatment of choice in cases of preretinal hemorrhage for more than 4 weeks and in those that have not resolved spontaneously or with laser treatment (Kumar 2017). Vitrectomy has good visual results (Fig. 6.5) and a low rate of complications, preventing persistent vitreous hemorrhage, epiretinal membrane, and premacular cavity development (Durukan et al. 2008; Goel et al. 2011). Among the most frequent complications are cataract, increased ocular pressure, and retinal detachment (Schmitz et al. 2000).

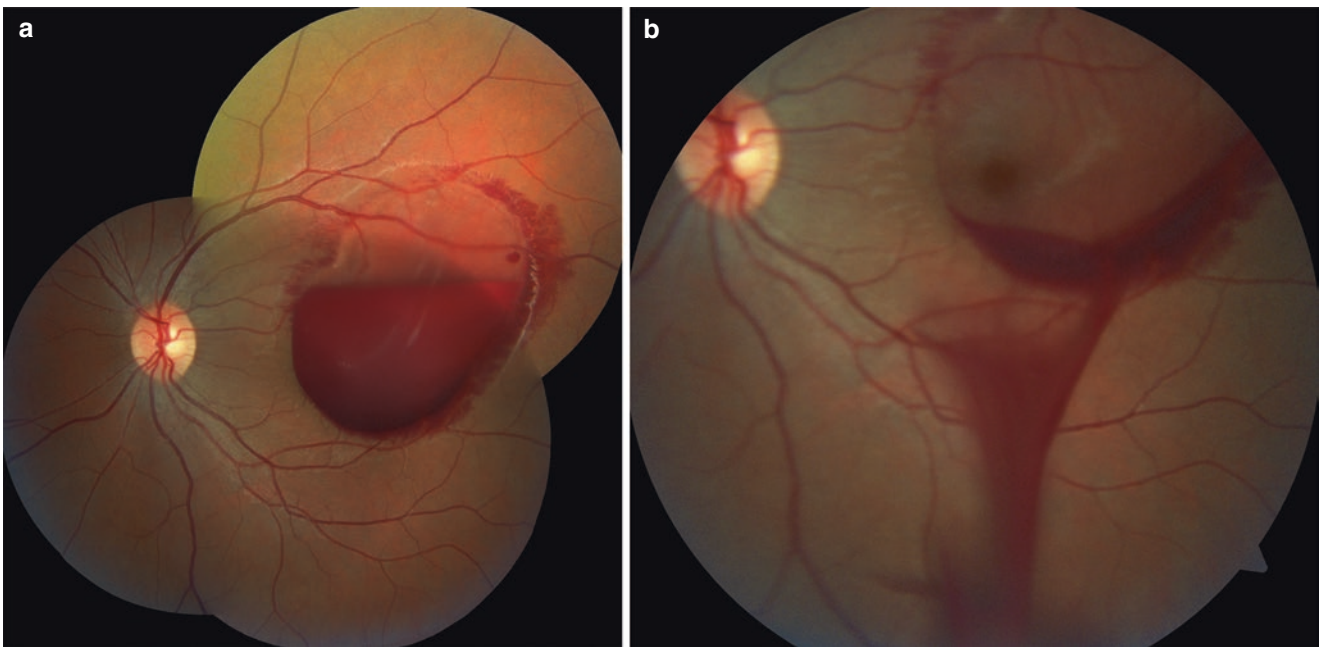

Fig. 6.4 (**a**) Valsalva retinopathy. (**b**) Color fundus photograph shows the drainage of bleeding through the vitreous cavity after YAG laser

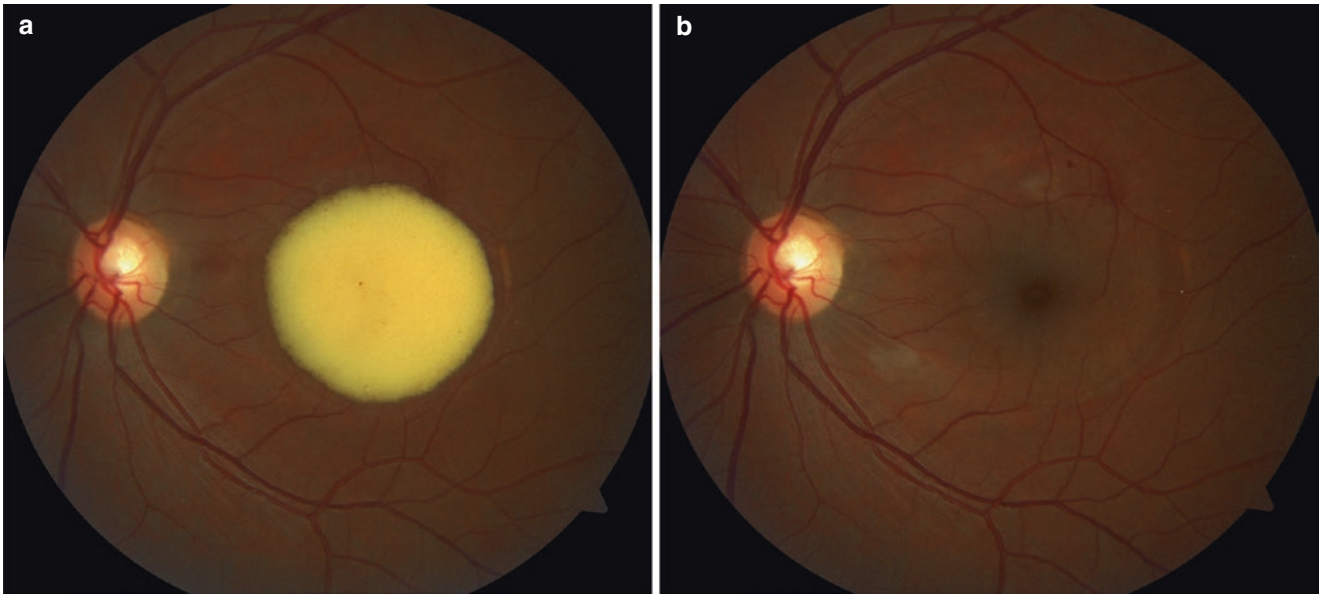

Fig. 6.5 (**a**) Color fundus photograph shows the change of hemorrhage coloration after 30 days of the beginning of the symptoms. (**b**) The same patient after ILM peeling by vitrectomy

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Purtscher Retinopathy

7

Débora Furuzawa Ribeiro, Eduardo Büchele Rodrigues, Ricardo Luz Leitão Guerra, and Carina Costa Cotrim

Introduction

Purtscher retinopathy is a microangiopathy characterized by loss of vision within hours to days after an indirect ocular injury, typically associated with cranial trauma or thoracic compression (Miguel et al. 2013). The incidence is an average of 2.4 persons per million per year (Medeiros et al. 2009; Xia et al. 2017). It was first described by Otmar Purtscher in 1910 in patients after severe head trauma with retinal findings as cotton-wool spots, retinal hemorrhage, non-capillary perfusion areas, and optic disk swelling (Fig. 7.1) (Buckley and James 1996; Agrawal and McKibbin 2007). "Purtscher-like retinopathy" is the term used to describe similar retinal signs without history of trauma.

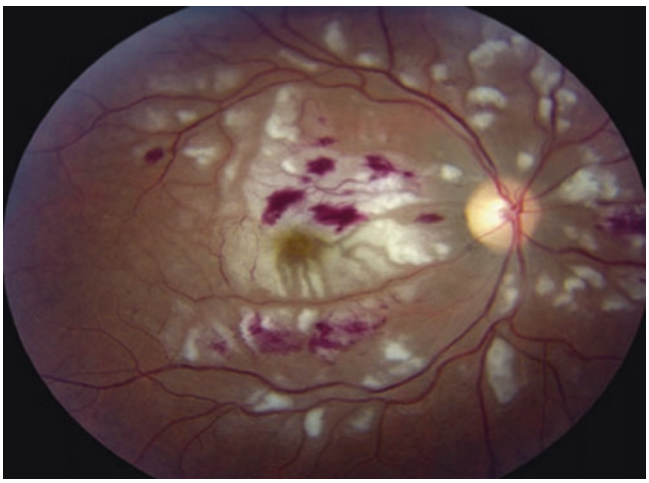

Fig. 7.1 Fundus color photography of patient with Purtscher retinopathy presenting cotton–wool spots and retinal hemorrhage

Etiopathogenesis

Purtscher first described five patients with similar retinal signs after a severe head trauma and postulated that the sudden rise in intracranial tension created extravasation of lymph from vessels, consequently, the superficial retinal whitening appearance (Eliott and Papakostas 2018). The severe head trauma is the most frequent cause of Purtscher retinopathy, but it has a different biologic mechanism compared to acute pancreatitis, the second most frequent case. The first is due to elevated intravascular pressure that leads to capillary endothelial damage—the same pathogeny of expansion of retinal veins and hyperviscosity (Miguel et al. 2013)—and the second by complement-mediated leukoembolization, caused by leukocyte aggregation and C5 activation (McCannel et al. 2017; Miguel et al. 2013). This mechanism was realized by an experiment with pigs, which used injection of fibrin clots into the ophthalmic artery and produced a similar Purtscher retinal image (Behrens et al. 1992), as well as other anterior experimental studies. Another mechanism is the arteriolar occlusion after air, fat, or amniotic fluid embolization. Presently, retinal arteriolar and capillary embolism with microinfarction is considered the most acceptable etiology for Purtscher retinopathy, and there is pathologic and experimental evidence to support this hypothesis (Harrison et al. 2011; Buckley and James 1996). Anatomically, the peripapillary capillary net is the principal zone of risk to ischemic injury after the arteriole's obstruction due to infrequent feed points and anastomoses in this place (Buckley and James 1996). Previous studies showed

D. F. Ribeiro (✉) · C. C. Cotrim
Department of Ophthalmology, Governador Celso Ramos Hospital, Florianópolis, SC, Brazil

E. B. Rodrigues
Department of Ophthalmology and Visual Sciences, Federal University of São Paulo, São Paulo, Brazil

Department of Ophthalmology, Saint Louis University School of Medicine, Saint Louis, MO, USA

R. L. L. Guerra
Department of Ophthalmology, Clínica de Olho Leitão Guerra, Salvador, BA, Brazil
e-mail: ricardo@leitaoguerra.com.br

E. B. Rodrigues et al. (eds.), *Trauma and Miscellaneous Disorders in Retina*, Retina Atlas, https://doi.org/10.1007/978-981-13-8550-6_7

choriocapillar involvement, especially in submacular choroid zone. Harrison et al. (2011) suggested a dysregulation in permeability of endothelial vessels by a rheological event in the microvasculature of the inner layer retinal posterior pole.

Clinical Features

The retinal findings of Purtscher retinopathy are: dot and blot hemorrhages, flame hemorrhages, Purtscher flecken, and cotton-wool spots, situated, respectively, in deeper retinal layers, nerve-fiber layer, retinal capillary, and superficial nerve-fiber layer (Harrison et al. 2011). The most common signs are cotton-wool spots (92%) and retinal haemorrhage (83%) found mainly in the posterior pole and corresponding into a size of 8-disc diameter. The bilateral involvement is the most frequent presentation (Miguel et al. 2013), even though bilateral involvement was reported in all cases of acute pancreatitis (Agrawal and McKibbin 2007). Purtscher flecken is a pathognomonic sign present as a consequence of the precapillary arteriole's occlusion and occurs in roughly 50% of cases (Agrawal and McKibbin 2007; Miguel et al. 2013) featuring a polygonal area of whitening in the inner retina between retinal arterioles and venules with a normal retina on either side of the vessels, extending an average of 50 μm. It has no prognostic value (Eliott and Papakostas 2018; Miguel et al. 2013). In Purtscher-like retinopathy, it is possible to find scattered intraretinal hemorrhages in the paramacular area, cotton-wool spots, and smaller fat embolism peripherally (McCannel et al. 2017). The range of visual loss varies from 20/20 to counting fingers. Occasionally, afferent pupillary defect, disc edema, and central or paracentral scotomas compose this pathology (Eliott and Papakostas 2018; McCannel et al. 2017; Xia et al. 2017).

Imaging Features

Fluorescein angiography may show variable findings, including normal choroidal filling, venous staining, disc leakage, blocked choroidal fluorescence following retinal arteriolar obstruction, and capillary nonperfusion (Figs. 7.2, 7.3, 7.4 and 7.5) (Eliott and Papakostas 2018; Kumar and Tomar 2017). The poor capillary perfusion of macular area was demonstrated by Kumar et al. in a study with swept-source optical coherence tomography (SS-OCT) with images of hyper-reflectivity of the inner retinal layers and ellipsoid layer defect. The optical coherence tomography angiography (OCTA) demonstrated that the total thickness of capillary plexus reduced. Furthermore, it is a helpful non-invasive tool to prognosticate the patient (Kumar and Tomar 2017). About visual field, central scotoma was observed in more than 90% of the cases in a systematic review (Miguel et al. 2013).

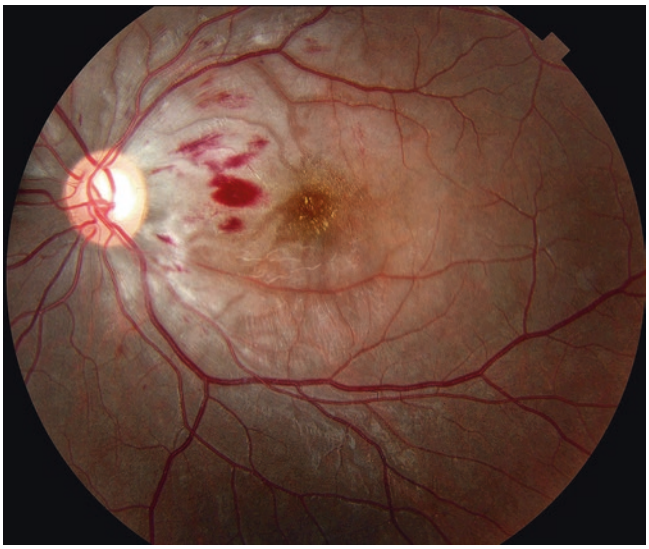

Fig. 7.2 The patient was a victim of pancreatitis and developed Purtscher-like retinopathy—findings are the same found in Purtscher's retinopathy. Fundus color photography demonstrating patches of superficial retinal whitening, cotton wool, and small hemorrhages in posterior pole. Special thanks to Ricardo Luz Leitão Guerra who gave the images of his article: Fundus autofluorescence in blunt ocular trauma, published in 2014 by Arq Bras Oftalmol (Guerra et al. 2014)

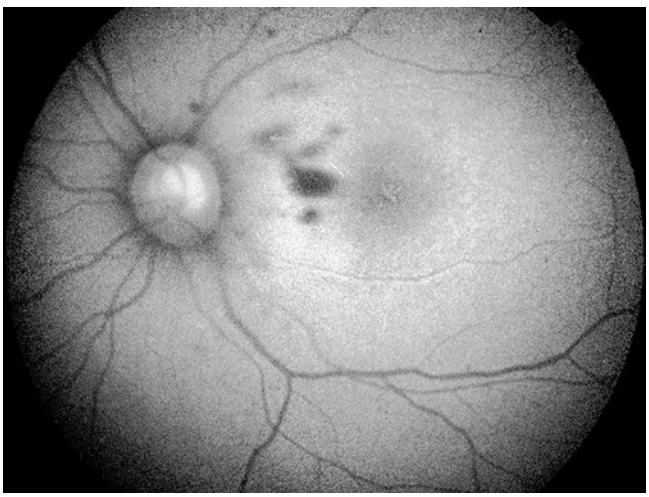

Fig. 7.3 The patient was a victim of pancreatitis and developed Purtscher-like retinopathy—findings are the same found in Purtscher's retinopathy. Autofluorescence exhibits hypoautofluorescence in the areas corresponding to intraretinal hemorrhage and mild perimacular hyperautofluorescence. Special thanks to Ricardo Luz Leitão Guerra who gave the images of his article: Fundus autofluorescence in blunt ocular trauma, published in 2014 by Arq Bras Oftalmol (Guerra et al. 2014)

Management

There is no protocol of treatment for Purtscher retinopathy. The treatment with corticosteroids is controversial, and many studies showed good results using expectant management (Miguel et al. 2013). The studies showed that no treatment for this reti-

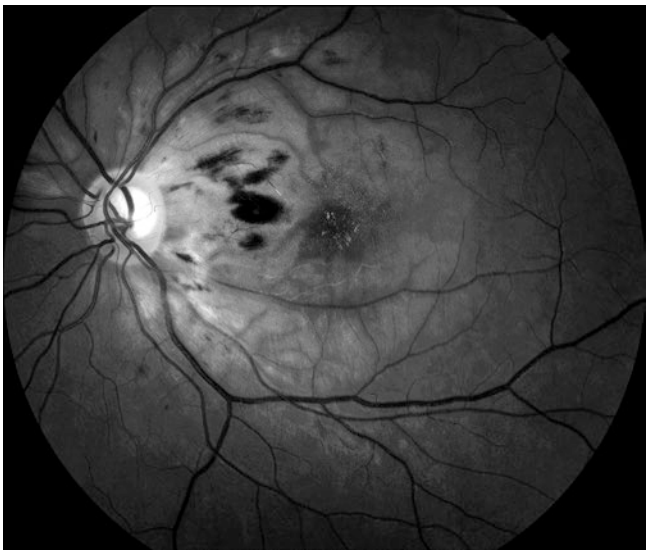

Fig. 7.4 The patient was a victim of pancreatitis and developed Purtscher-like retinopathy—findings are the same found in Purtscher's retinopathy. Red-free photography highlights hemorrhages and cotton wool. Special thanks to Ricardo Luz Leitão Guerra who gave the images of his article: Fundus autofluorescence in blunt ocular trauma, published in 2014 by Arq Bras Oftalmol (Guerra et al. 2014)

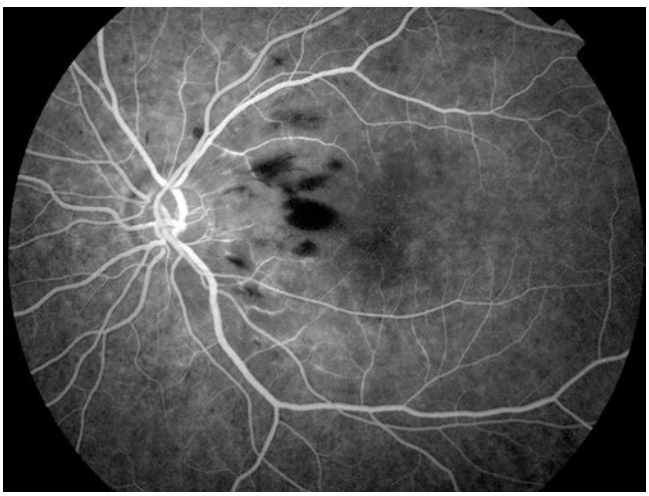

Fig. 7.5 The patient was a victim of pancreatitis and developed Purtscher-like retinopathy—findings are the same found in Purtscher's retinopathy. Fluorescein angiography exhibits blocked fluorescence in the areas corresponding to intraretinal hemorrhage. Special thanks to Ricardo Luz Leitão Guerra who gave the images of his article: Fundus autofluorescence in blunt ocular trauma, published in 2014 by Arq Bras Oftalmol (Guerra et al. 2014)

nopathy had a significant efficacy; in addition, the greater number of eyes had better visual function without any specific treatment (Buckley and James 1996; Agrawal and McKibbin 2007), except in cases of optic nerve involvement—usually with atrophy and bad visual acuity prognosis. Another possibility of permanent decrease in visual acuity occurs when vascular obstruction involves the capillaries of the macula foveolar arcade, observed a different time of improvement in visual acuity: earlier with corticosteroids (1–3 months) compared with no treatment (4–6 months and after 6 months) (Xia et al. 2017). Agrawal et al. demonstrated that 50% of participants improved at least two lines, 23% improved at least four lines, and only one of the studied eyes had worse vision at last follow-up. It was suggested that the etiology can affect the final visual acuity, with better results in pancreatitis and trauma cases. Clinical exam and image findings could not predict the patient prognosis, and these patients should be followed up for at least 6 months of the event (Miguel et al. 2013).

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Subhyaloid Hemorrhage, Traumatic Macular Hole, and Electric Burn

8

Thaís Sousa Mendes and Eduardo Büchele Rodrigues

Subhyaloid Hemorrhage

Definition

Subhyaloid hemorrhage can be described as the accumulation of blood and its confinement under hyaloid, which can cause severe visual impairment if located at macular region (Fig. 8.1).

Pathogenesis

Various diseases are related to subhyaloid hemorrhage occurrence such as trauma, retinal diseases (hypertensive retinopathy, macroaneurysms, valsalva retinopathy, age-related macular degeneration, diabetic retinopathy, and polypoidal vasculopathy.

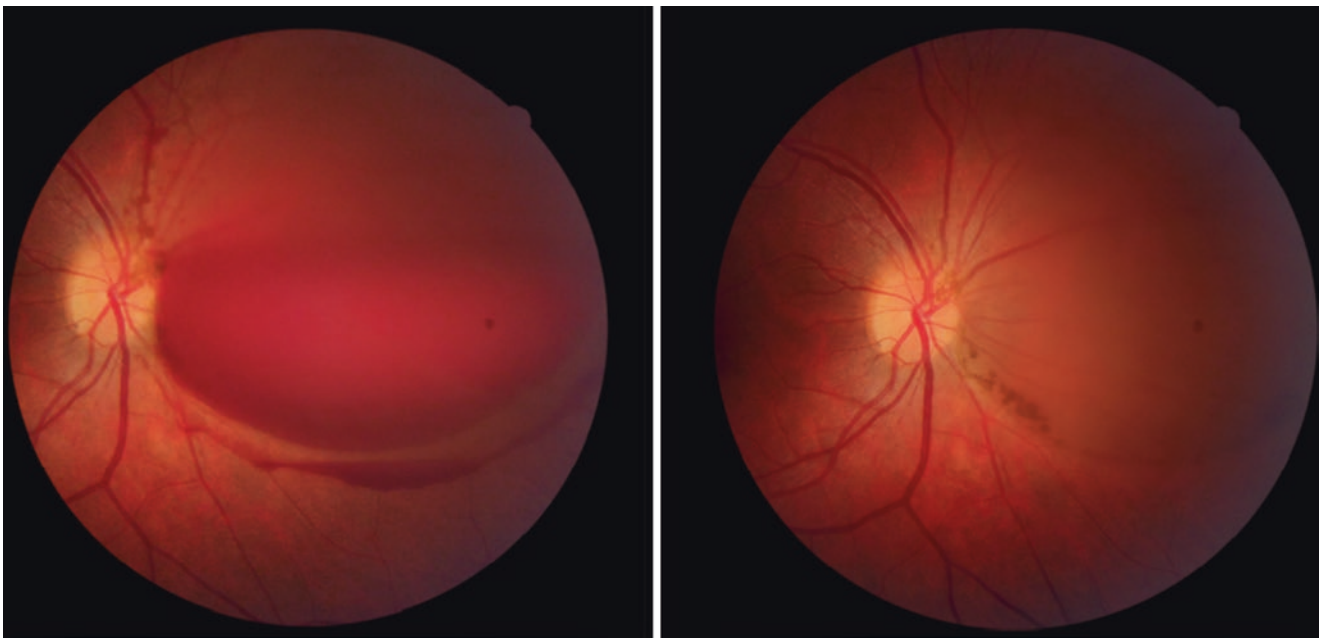

Fig. 8.1 Left: Subhyaloid hemorrhage nasal to disc caused by valsalva. Right: spontaneous resolution after 3 months. (courtesy by Edmundo Almeida, MD)

T. S. Mendes
Department of Ophthalmology, Federal University of Sao Paulo, São Paulo, Brazil

E. B. Rodrigues (✉)
Assistant Professor of Ophthalmology, Saint Louis University Eye Institute, Saint Louis University, Saint Louis, MO, USA

E. B. Rodrigues et al. (eds.), *Trauma and Miscellaneous Disorders in Retina*, Retina Atlas, https://doi.org/10.1007/978-981-13-8550-6_8

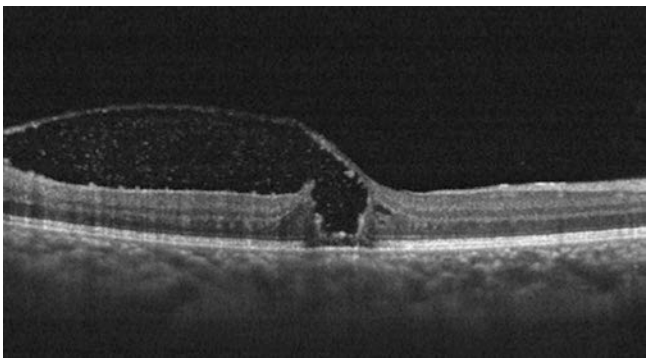

Fig. 8.2 Spectral domain optical coherence tomography (SD-OCT) of macular region, subhyaloid hemorrhage with pseudomacular hole. (courtesy by Alexandre Rosa, MD)

Prognosis and Treatment

Regarding treatment options, it can vary from observation, intravitreal gas injection to *pars plana* vitrectomy with or without tissue plasminogen activator (tPA).

Spontaneous resolution can occur after many weeks; however, blood accumulation can be toxic to retina (Fig. 8.2).

Most recently, authors have reported successful resolution of premacular hemorrhage with laser hyaloidotomy (Nd:YAG laser). Laser hyaloidotomy creates a drainage spot for the blood to reach vitreous cavity where absorption occurs. This maneuver must be performed before the blood is coagulated, and the hemorrhage size needs to be at least three-disc diameters to prevent retinal injury during laser (Alarfaj et al. 2018; Khadka et al. 2016; Rafa et al. 2017).

Traumatic Macular Hole

Definition

Traumatic macular hole (TMH) is a rare clinical condition, with a frequency of less than 10% in the related literature (Fig. 8.3). Blunt trauma is considered the most common cause, especially in sports accidents involving younger patients, which are subjects to ocular trauma.

Pathogenesis

Concerning pathogenesis, it is debated. Authors discussed that TMH is formed after induced cystoid alteration breakdown, abrupt vitreous detachment, or instantaneous tear caused by concussion and horizontal forces, leading to retinal full-thickness defect with severe central visual loss.

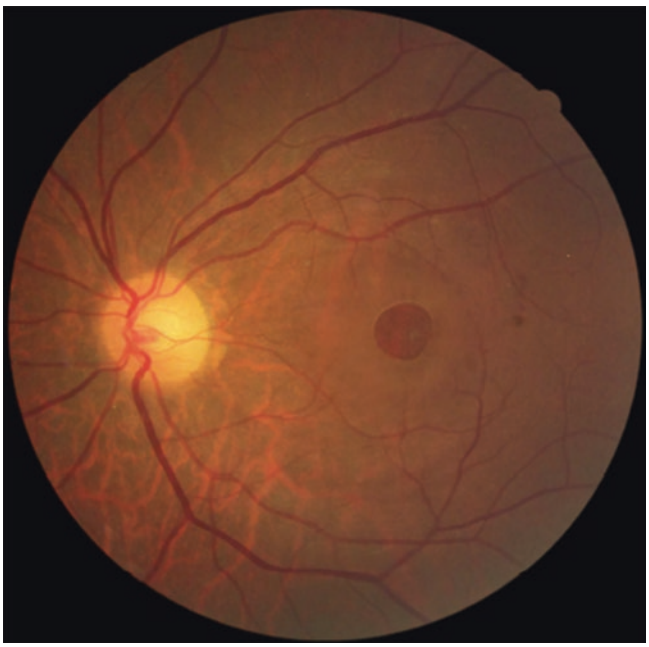

Fig. 8.3 Fundus photo of traumatic macular hole

Prognosis and Treatment

Spontaneous closure may occur in half of cases after 6 months, especially in younger patients. Recently, optical coherence tomography (OCT) predictors of TMH closure were reported by Chen et al. They believe that OCT can provide in vivo evaluation of retinal changes and helps predict TMH closure (Fig. 8.4) (Chen et al. 2015; Liu and Grzybowski 2017; Michi et al. 2012; Nasr et al. 2011; Sanjay et al. 2012).

These characteristics are small size hole at baseline, with less intraretinal cysts and young age. However, if TMH doesn't close after 6 months of observation, *pars plana* vitrectomy with internal limiting membrane (ILM) peeling and gas tamponade are needed.

Vitrectomy successful closure rate for TMH is approximately from 45% to 92.5%; however, visual outcomes are unlikely to be satisfactory because of previous retinal damage due to trauma such as submacular hemorrhage and choroidal rupture. ILM peeling using inverted flap technique has been reported as a treatment option for larger TMH (basal diameter 1300–2000 μm).

Electric Burn

Definition

Electric burn is defined by an electrical current that reaches the human body during a contact with voltaic arc. It is most

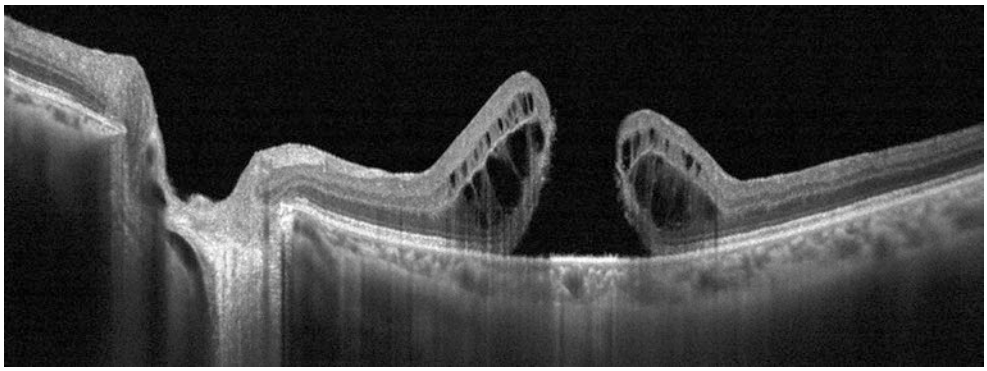

Fig. 8.4 Traumatic and chronic macular hole with irregular edges, intraretinal cysts, and photoreceptor disruption

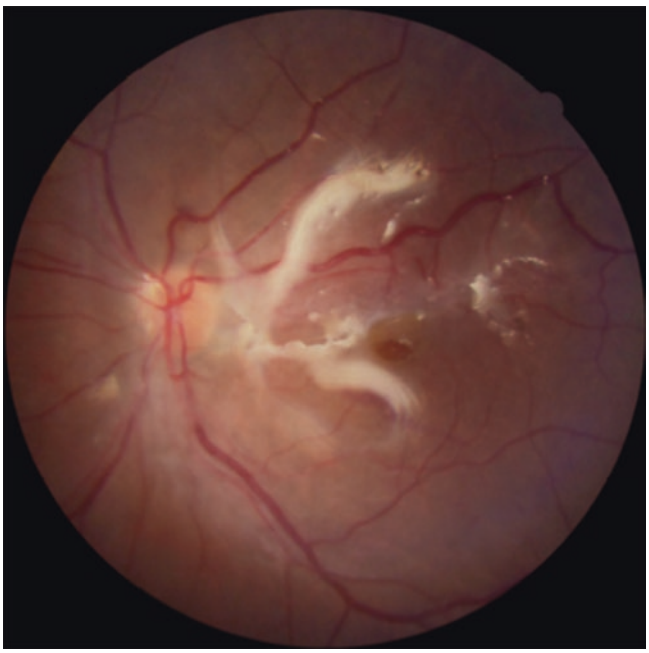

Fig. 8.5 Fundus photo after vitrectomy with silicone oil tamponed for retinal detachment induced by electric burn injury

common in young male adults, which can be affected during work, especially in industrial places where high-voltage equipment is present. There are several clinical forms due to electric burn and mortality rate is around 11% (Buja et al. 2010; Lakosha et al. 2009).

Pathogenesis

The mechanisms of burn can be direct (direct contact with ionized surface) or indirect (passage of electric current). Ocular damage is reported in half of the victims of lightning, mostly located in the anterior segments such as eyelid burns, thermal keratitis, hyphema, and cataract. Less commonly, uveitis can be induced by electrical shock (Luz et al. 2009).

Prognosis and Treatment

Most cases are accompanying with deep injuries with mandatory hospitalization.

Retinal injury is rare. Some authors reported macular hole formation and retinal detachment after high-voltage shock, leading to permanent visual loss and blindness (Fig. 8.5) (Faustino et al. 2014; Kotak et al. 2013; Rajagopal et al. 2010; Rao et al. 2009).

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Retained Intraocular Foreign Body

9

Pedro Felipe Mylla Boso
and Luis Felipe da Silva Alves Carneiro

Introduction

Ocular trauma is an important cause of preventable visual impairment worldwide. An estimated 2.4 million cases occur every year in the USA, with serious vision-threatening injuries ranging from 20,000 to 68,000 of these. Most cases presented in a study by the National Eye Trauma System Registry were seen in young male patients and happened in a home or workplace setting (Parver et al. 1993); therefore, the economic and social burden of this topic cannot be overstated. According to the Birmingham Eye Trauma Terminology (BETT) system, intraocular foreign bodies (IOFBs) or retained foreign objects are a subset of penetrating injuries, when there is an entrance wound without an exit wound (Kuhn et al. 2004). They account for 14–43% of open-globe injuries (OGIs) (Patel et al. 2012; Zhang et al. 2011; Shock and Adams 1985) and pose challenging surgical cases in which the goal is to remove the foreign body (FB) while attempting to preserve vision and restore ocular architecture (Greven et al. 2000). The most common site of entry is the cornea (65%), followed by the sclera (25%) and the limbus (10%) (Rathod and Mieler 2011). Most IOFBs are seen in the posterior segment (Katz and Moisseiev 2009; Zhang et al. 2011) but can also be found in the anterior chamber or even in the lens (Fig. 9.1). The composition of IOFBs differ, ranging from organic material such as wood and insect parts to inert substances like plastic and glass, but most of them are metallic in nature (Greven et al. 2000; Woodcock et al. 2006).

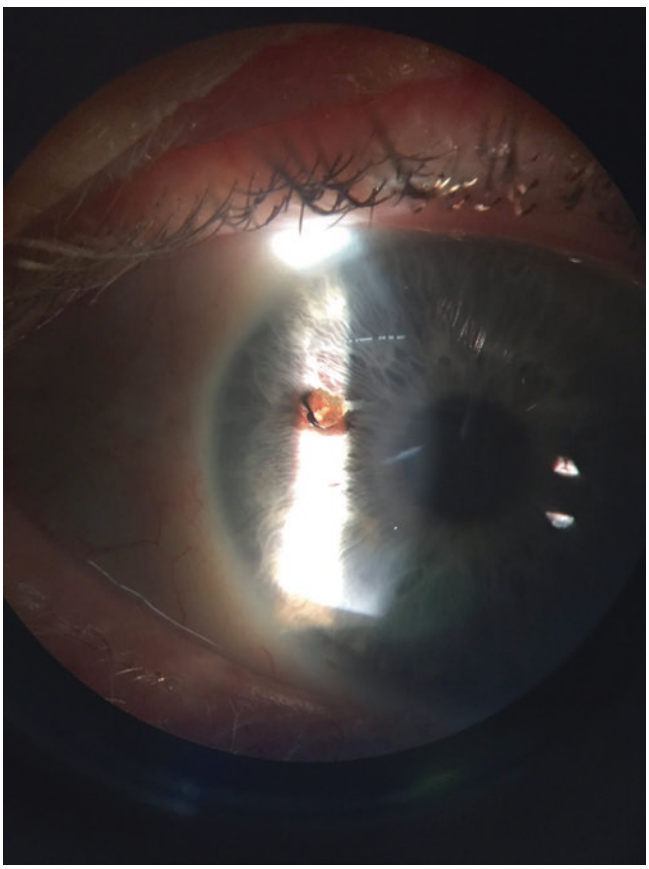

Fig. 9.1 Ophthalmic biomicroscopic exam of a patient with IOFB embedded in the iris. Courtesy of Dr. Andrei Gabriel Melo

Pathophysiology

Mechanical Damage

IOFBs can cause damage in more ways than one, and each must be considered when evaluating the patient and planning the treatment. There must be an entry wound through the cornea, sclera, or limbus, and direct mechanical damage to

P. F. Mylla Boso (✉)
Instituto de Olhos de Florianópolis, Florianópolis, SC, Brazil

L. F. da Silva Alves Carneiro
Department of Ophthalmology, Santa Casa de Belo Horizonte, Belo Horizonte, Brazil

E. B. Rodrigues et al. (eds.), *Trauma and Miscellaneous Disorders in Retina*, Retina Atlas, https://doi.org/10.1007/978-981-13-8550-6_9

intraocular structures ensues. Wound length can be used to assess the risk of retinal injury. When the IOFB passes through a shorter wound, it dissipates less kinetic energy and may travel further inside the eye, allowing it to cause greater damage to the retina. FBs entering through the sclera often cause more damage than those entering through the cornea (Loporchio et al. 2016; Kuhn and Pieramici 2002). The shape and mass of the FB can also influence on its ability to cause mechanical injury, as heavier objects and those described as blade-shaped achieve greater penetration to the posterior segment (Woodcock et al. 2006), and blunt objects are considered more destructive than sharp objects presumably due to an augmented transfer of energy at the time of the impact (Potts and Distler 1985; Kuhn and Pieramici 2002). The complications most often related to mechanical damage include traumatic cataract with or without breach of the capsule, vitreous hemorrhage, retinal detachment, and hyphema (Zhang et al. 2011; Colyer et al. 2007).

Intraocular Inflammation

Aside from mechanical damage, the FB can cause intraocular inflammation either from the presence of the object itself or secondary to trauma to the iris, hemorrhage, and exposure of vitreous or lens material to other ocular structures, among other lesions. This inflammatory response can lead to intraocular pressure (IOP) elevation, uveitis, and synechia formation and has been found to play a role in post-traumatic proliferative vitreoretinopathy (PVR) (Cardillo et al. 1997).

Metallosis

The presence of metallic IOFBs can cause damage because of chemical reactions following deposition and toxicity known as metallosis, particularly by iron and copper ions but also by lead, zinc, and nickel (Lit and Young 2002).

Siderosis Bulbi

The term siderosis bulbi is used to describe the condition caused by retention and oxidation of iron-containing FBs, and clinical findings include iris heterochromia, brownish pigmentation of the lens capsule, cataract formation, mydriasis, and retinal pigmentary degeneration, with presentation ranging from 3 weeks to 20 years after the injury, when it could be elicited (Sneed and Weingeist 1990; Dowlut et al. 2019). A characteristic finding in an electroretinogram (ERG) is an initial increased A wave followed by a progressive reduced B wave subsequently (Kuhn and Pieramici 2002), thus being an important investigative exam as electrophysiologic signs of siderosis can emerge earlier than clinical changes (Figs. 9.2, 9.3, 9.4, 9.5, 9.6, 9.7 and 9.8 refer to a 19-year-old patient, victim of metallic foreign body trauma presenting with visual acuity 20/20 and IOP 16 on the first day after trauma. The patient was submitted to PPV (without complications) and 90 days after the procedure, visual acuity reduced from 20/20 to 20/40 with exams suggesting ocular siderosis. Special thanks to Dr. Gustavo Barreto de Melo, from UNIFESP-SP, Brazil; Knave 1969).

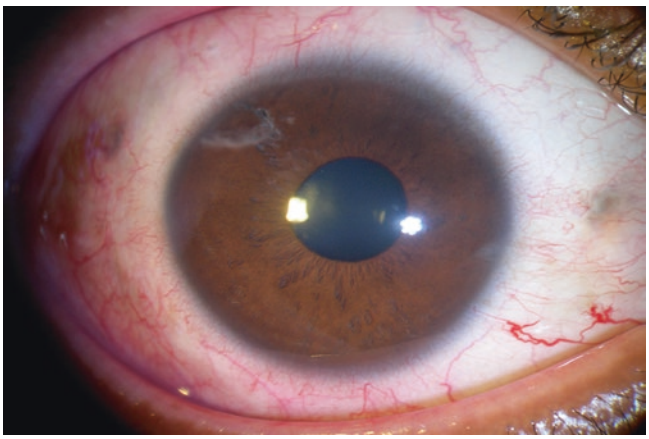

Fig. 9.2 Ophthalmic biomicroscopic exam with superior temporal corneal and iris lesion

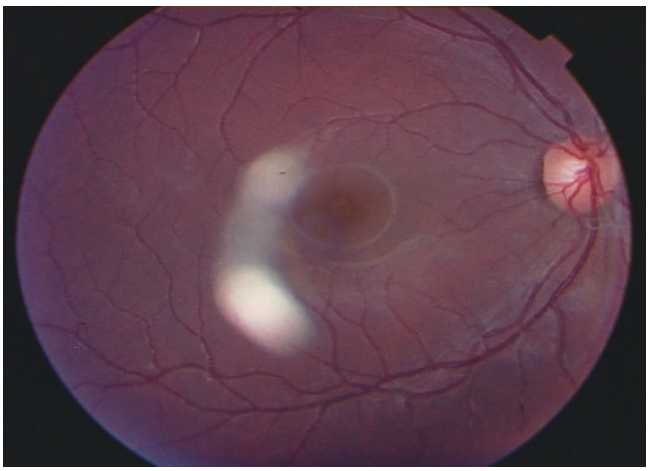

Fig. 9.3 Color fundus photography presenting with IOFB on vitreous body. The retina was perfectly attached

Chalcosis

The presence of a copper-containing IOFB can lead to a disease called chalcosis, the pathogenesis of which remains unclear (Loporchio et al. 2016) but may be related to a process of lipid peroxidation (McGahan et al. 1986). Chalcosis can present as a rapid endophthalmitis-like reaction with

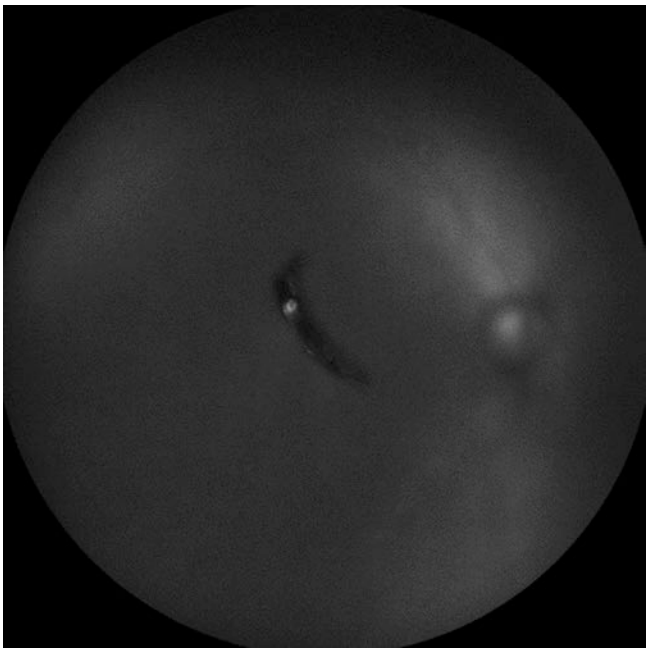

Fig. 9.4 Fundus autofluorescence delimiting the IOFB

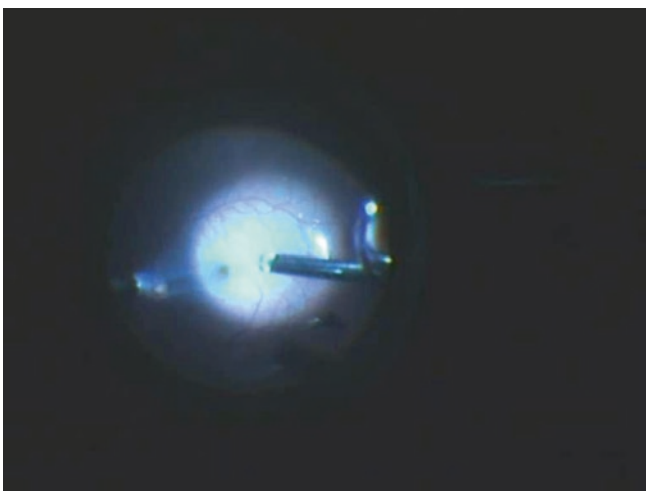

Fig. 9.5 Pars plana vitrectomy removing the IOFB

hypopyon formation and eventual phthisis bulbi or as more chronic reactions that can present similarly to those seen with hepatolenticular degeneration or high endogenous copper levels which can lead to the formation of Kayser–Fleischer ring, sunflower cataract, or presence of tiny refractile copper particles in the aqueous humor or iris (Lit and Young 2002).

Diagnosis and Investigation

Any injury deemed more severe or life threatening should be addressed prior to the ophthalmic lesion. When suspicion of an IOFB arises, some questions must be answered as to the presence or absence of an IOFB, location and number of FBs, circumstances of the injury, and associated conditions (Kuhn and Pieramici 2002). History taking is fundamental when considering the possibility of an IOFB and should be obtained whenever possible with special attention to the mechanism of the trauma (Yeh et al. 2008), as it can aid in the identification of the nature of the FB and its location (Lit and Young 2002). Other important history data that aids in the therapeutic planning and decision-making as well as in prognostic and medico-legal factors include setting and time of injury, use of safety equipment, occurrence of any interventions since the injury, when the patient last had anything to eat or drink, and the presence of allergies (Rathod and Mieler 2011). The physician must keep in mind that some patients do not experience pain and may present with normal vision, so the presence of an IOFB should be suspected on all OGIs, when the history points to high-risk activities (such as use of powered tools or hammering metal) or when a suggestion of an IOFB arises during examination (Lit and Young 2002; Kuhn and Pieramici 2002).

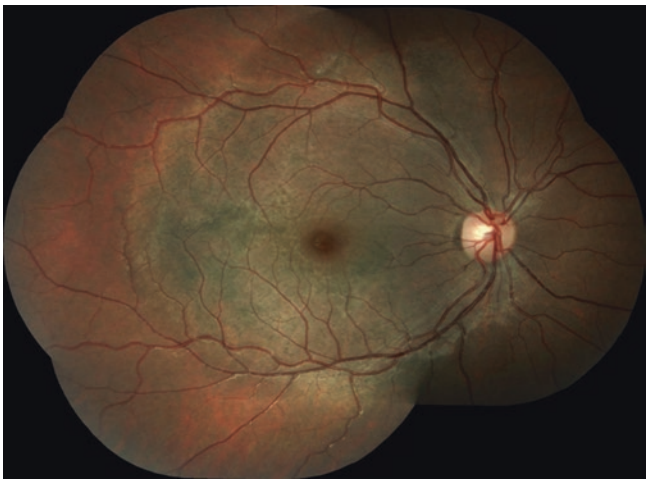

Fig. 9.6 Fundus color photography presenting with retinal degeneration in the posterior pole

After initial history taking, a thorough clinical examination must follow, including external inspection, visual acuity assessment, pupillary evaluation, slit lamp, and fundus examination when possible. When feasible a rough estimate of visual acuity before the traumatic event can help in characterizing the extent of visual loss due to the injury. Some important signs to look for during examination, apart from the obvious direct viewing of the IOFB or its trajectory, are focal lens opacities, IOP or pupil asymmetry, iris heterochromia, and small self-sealing wounds (Rathod and Mieler 2011). Until an OGI is ruled out, care should be taken not to exert undue pressure on the globe (Katz and Moisseiev 2009), avoiding scleral depression, gonioscopy, applanation tonometry, and B-scan, for instance. Clinical examination alone cannot identify all IOFBs (Patel et al. 2012); therefore,

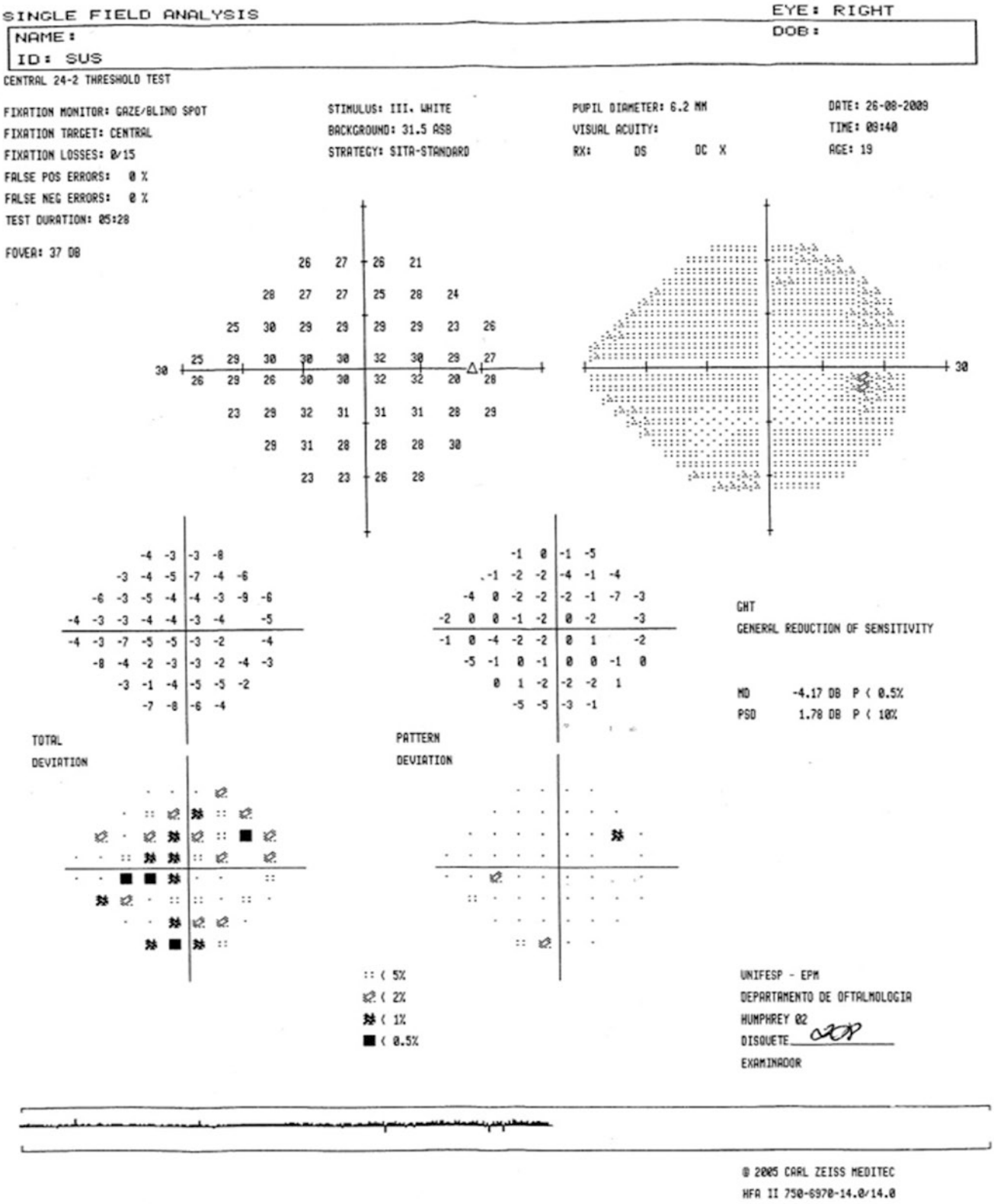

Fig. 9.7 Visual field of the right eye presenting with general reduction of sensitivity

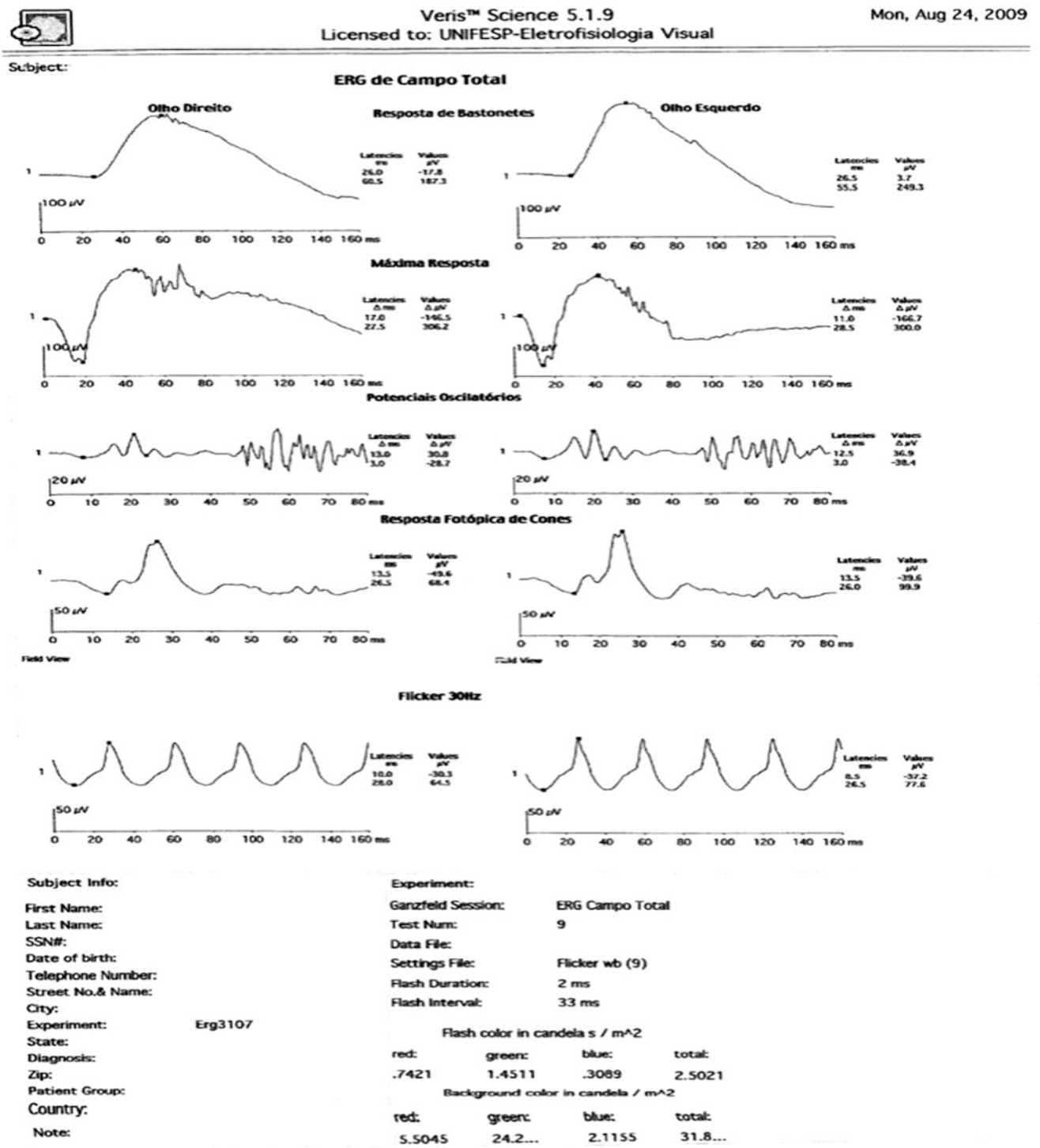

Fig. 9.8 Full-field ERG of the same eye presenting with a 17% decrease in rods and cones responses

ocular imaging should be employed even when one is identified due to the possibility of multiple IOFBs and to aid in its precise location (Woodcock et al. 2006). Ocular imaging modalities for the investigation of IOFBs consist mainly of radiography, computed tomography (CT), magnetic resonance imaging (MRI), and ultrasonography (USG). The choice of modality used for investigation will depend on some aspects such as availability, suspected location and composition of the FB, and overall condition of the globe.

Radiography

Plain film radiography (or X-ray) can be used as a screening modality for IOFBs and orbital FBs (Saeed et al. 2008), but it is being replaced by CT (Kuhn and Pieramici 2002) as it has been found unnecessary (Bray and Griffiths 1991) because it lacked sensibility in detecting IOFBs (Bryden et al. 1990) besides being a poor method to accurately localize IOFBs (Lit and Young 2002).

Computed Tomography

The mainstay of radiologic investigation is the CT scan (Fig. 9.9) (Parke et al. 2013; Rathod and Mieler 2011; Yeh et al. 2008; Lit and Young 2002). It has been found to be the most reliable method to identify IOFBs in patients with OGIs when compared to clinical examination and B-scan USG (Patel et al. 2012). CT with no contrast can detect up to 100% of metallic IOFBs greater than 0.05 mm^3, although for nonmetallic materials the sensitivity may be lower, with helical CT possibly being superior for smaller fragments (Chacko et al. 1997). The scan should be made with thin cuts (1.0–1.5 mm) in at least two planes. For metallic objects the bone windows are the most helpful, while less radiopaque materials are best seen on soft-tissue windows, but certain plastic, ceramic, or even wood can be difficult to visualize with CT scans (Lit and Young 2002).

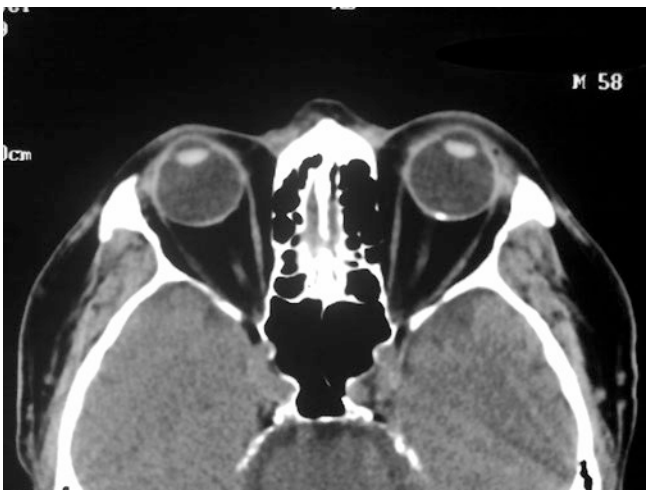

Fig. 9.9 CT scan of patient with metallic IOFB in the posterior pole of the left eye

Magnetic Resonance Imaging

MRI should only be used once the presence of a metallic IOFB is ruled out (Loporchio et al. 2016). That is because it can cause the movement of ferromagnetic FB leading to further intraocular damage, such as hyphema (Lawrence et al. 2015; Ta and Bowman 2000). For nonmetallic IOFBs, MRI shows superior detection when compared to CT, especially when they are small in size (Moisseiev et al. 2015). Tiny ferromagnetic IOFBs that cannot be identified by X-ray, CT, and B-scan USG may be detected by a 1.0 Tesla MRI without causing further intraocular damage (Zhang et al. 2009).

Ultrasonography

B-scan USG can be used when an IOFB cannot be visualized directly or through CT, but it must be performed extremely carefully in select eyes with OGIs, as there is a risk of extruding globe contents if pressure is exerted with the probe (Loporchio et al. 2016). It has been shown to be useful in examining the IOFB's relationship to soft-tissue pathology such as retinal detachment and when there is secondary vitreous hemorrhage (Coleman et al. 1987; McNicholas et al. 1995). Another modality of imaging that can be used is ultrasound biomicroscopy, which can be useful to detect IOFBs located in the anterior chamber angle, around the ciliary body or in the retrolental space (Deramo et al. 1998).

Management

Clinical Management

In many cases ocular injury is part of a multiple trauma setting (Dannenberg et al. 1992), so it must be confirmed that the patient is medically stable. Once an OGI is diagnosed or highly suspected, the eye should be protected with a rigid shield such as a Fox shield or other protection that does not compress the eye or lids. Anti-nausea medication should be administered if needed to reduce the chance of vomiting, which can cause expulsion of intraocular content. Pain medication should also be used as needed. Administration of tetanus booster or vaccine when needed is part of the considerations to be made. Another important aspect is

proper patient and family counseling as IOFBs pose a significant risk of permanent visual dysfunction and ocular morbidity.

Post-traumatic endophthalmitis is an important complication and is more commonly caused by gram-positive organisms, namely coagulase-negative staphylococci and streptococci, although fungal and gram-negative cases also occur (Yeh et al. 2008). *Bacillus cereus* is a common causative organism that presents a particularly poor visual prognosis (Bhagat et al. 2011). Prophylactic antibiotics are widely used, commonly penicillin, cephalosporins, vancomycin, or fluoroquinolones (Parke et al. 2013).

Surgical Management

Surgical considerations include the timing of IOFB removal and some factors that may influence this decision are the patient's overall health status, presence or absence of clinical endophthalmitis, the availability of trained personnel and equipment (Yeh et al. 2008), and the nature of the trauma, as wounds containing soil or vegetable matter pose greater risk of post-traumatic endophthalmitis (Essex et al. 2004). If clinical signs of endophthalmitis are present, globe repair with immediate IOFB removal is almost always indicated, except when concomitant life-threatening injuries preclude ophthalmic surgery. Early removal of IOFB (within 24 h after the injury) has the potential advantages of reducing the risk of endophthalmitis and PVR, and submitting the patient to a single procedure under anesthesia (Yeh et al. 2008; Thompson et al. 1993; Essex et al. 2004; Jonas and Budde 1999), so if the IOFB is fresh it is probably best that it is removed, preferably under general anesthesia (Kuhn and Pieramici 2002). Likely advantages of delayed removal include improved control of inflammation, ability to better assess intraocular structures, and possible development of spontaneous posterior vitreous detachment, making vitrectomy surgery easier (Colyer et al. 2007).

Surgical Management: Chronic Cases

If there is a chronic retained IOFB with no visual complaints, no signal of metallosis, and no electrophysiologic alterations, the decision to remove it must consider acute and chronic surgery-related complications versus the possibility of delayed development of IOFB complications (Kuhn and Pieramici 2002). Surgical approach is case-dependent, and closure of the entry wound should be performed initially to provide stability to the eye. Removal should be made in a manner that minimizes trauma, and generally the entry wound is not recommended as the site for this purpose (Rathod and Mieler 2011).

Surgical Management: Anterior Chamber IOFBs

Anterior chamber IOFBs are usually removed during primary repair (Kuhn and Pieramici 2002) and can usually be removed through a secondary limbal incision with forceps or a magnet (for ferromagnetic FBs) with attempts to dislodge it beforehand with saline or viscoelastic if located deep in the angle (Lit and Young 2002). Inducing miosis can protect the lens during removal. If the FB is located in the iris, an iridectomy may be required to remove the iris and the FB (Loporchio et al. 2016).

Surgical Management: Intralenticular FBs

Intralenticular FBs account for approximately 5–10% of all IOFBs and can lead to the development of glaucoma and severe intraocular inflammation due to the escape of lenticular proteins (Medina et al. 2006). Some authors recommend lens removal if cataract is present or inevitable (Kuhn and Pieramici 2002), and studies show that in some cases where endophthalmitis is not present cataract extraction with IOFB removal and intraocular lens (IOL) implantation is a safe option (Medina et al. 2006; Güler et al. 2010; Cazabon and Dabbs 2002).

Surgical Management: Posterior Segment IOFBs

The removal of posterior segment IOFBs should be conducted via pars plana vitrectomy (PPV) in most cases (Figs. 9.10, 9.11, 9.12 and 9.13 refer to a 56-year-old patient referred to a retinal specialist with retinal detachment and history of penetrating ocular trauma without IOFB found in initial ocular ultrasound. The first approach after trauma was to perform corneal suture associated with phacoemulsification and implantation of IOL. After that another ocular ultrasound was performed showing an IOFB and retinal detachment. Special thanks to Dr. Ricardo Luz Leitão Guerra, from Clínica de Olho Leitão Guerra, Salvador-BA, Brazil; Yeh et al. 2008) with advantages including direct visualization of IOFB for removal and ability to remove media opacities concomitantly, such as cataract and vitreous hemorrhage (Greven et al. 2000). Strategies for IOFB removal depend on size and composition. Small (<1.0 mm), metallic, ferromagnetic FBs may be removed with an intraocular magnet, while small nonferrous material can be removed with the vitreous cutter alone. Medium-sized IOFBs (1.0–3.0 mm) can be removed with basket forceps, while larger (3.0–5.0 mm) or glass FBs can be removed with the use of diamond-coated forceps. Small and some medium-sized FBs can be removed through the sclerotomy with enlargement of the wound, if needed, while FBs

larger than 4.0 × 4.0 × 4.0 mm may require a scleral tunnel removal (Yeh et al. 2008). After extraction of the IOFB, a thorough retinal examination with scleral depression should be performed to check for retinal tears, retinal detachment, or choroidal detachment. Laser photocoagulation is used to treat retinal tears, and retinal cryopexy is performed if blood precludes good laser treatment. Gas or oil tamponade can be used for retinal detachment. An encircling scleral buckle may be placed depending on the anatomy or anticipated likelihood of PVR development (Loporchio et al. 2016).

Complications include hyphema, cataract, lens luxation, corneal damage, metallosis, post-traumatic endophthalmitis, PVR, vitreous hemorrhage, retinal detachment, and sympathetic ophthalmia (Zhang et al. 2011).

Prognostic Factors and Outcomes

There are many factors involved in prognosis as injuries present in many variations with different extents and locations of damage, as well as distinctive treatment plans depending on several factors.

Visual acuity (VA) at presentation has been shown to be an important prognostic factor for final VA (Williams et al.

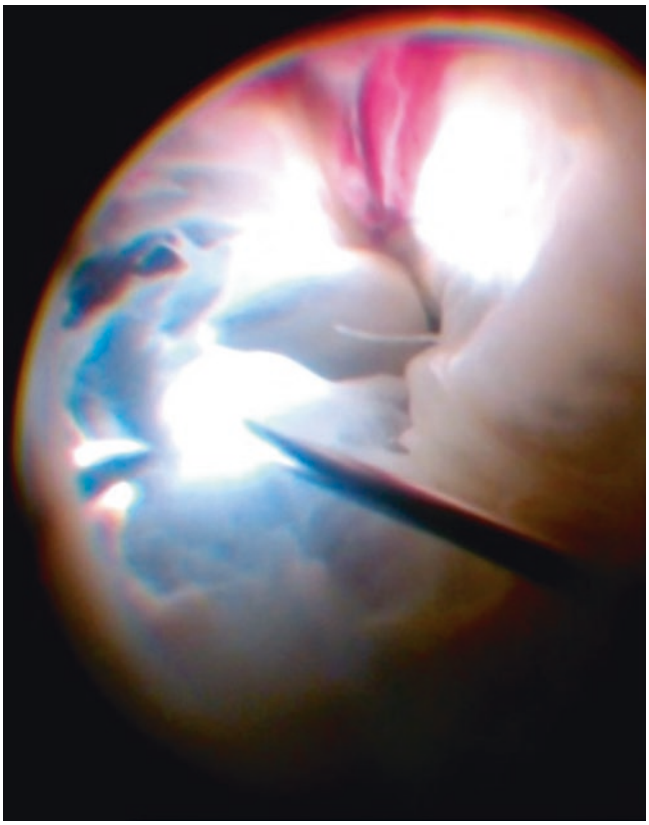

Fig. 9.10 PPV with retinal detachment and superior hemorrhage

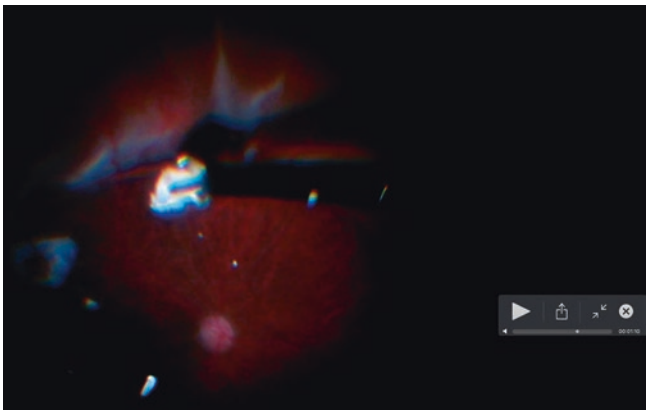

Fig. 9.12 Forceps grabbing IOFB in the vitreous cavity

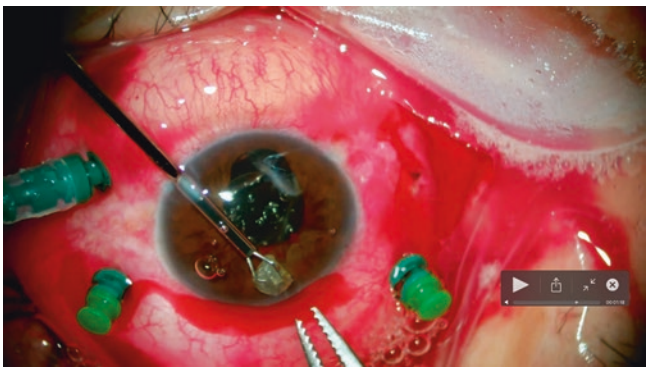

Fig. 9.13 Forceps removing IOFB through anterior chamber

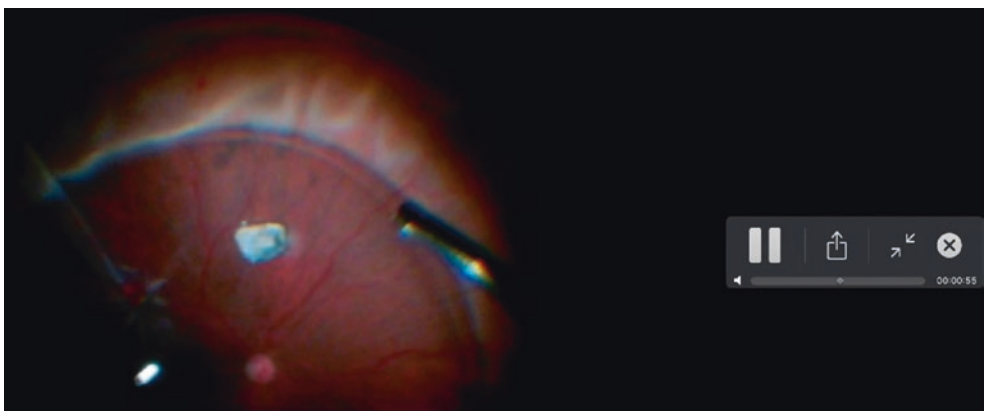

Fig. 9.11 In this step of the surgery liquid perfluorocarbon was injected, consequently elevating the IOFB

1988; Greven et al. 2000; Zhang et al. 2011). That may be because to some extent initial VA can reflect the severity of tissue damage in most cases (Zhang et al. 2011). Factors of poor prognosis are: worse presenting VA, larger wounds, IOFB in the posterior segment, endophthalmitis, wound larger than the IOFB in the largest length (Zhang et al. 2011), wound larger than 4 mm (Williams et al. 1988), vitreous hemorrhage, presence of afferent pupillary defect (Wickham et al. 2006; Greven et al. 2000), uveal prolapse (Ehlers et al. 2008), presence of retinal detachment (Wani et al. 2003), development of PVR (Szijarto et al. 2008), and IOFBs with increased mass (Woodcock et al. 2006). Better prognosis is seen in patients with normal lens at presentation, decreased wound length, anterior segment IOFB (Ehlers et al. 2008), need for only one or two operations in the treatment of the injury (Williams et al. 1988), absence of endophthalmitis, and absence of retinal detachment (Lieb et al. 2003).

To simplify an often confusing array of prognostic factors, the Ocular Trauma Score was developed to simply and quickly estimate visual prognosis based on initial examination (Kuhn et al. 2002). Visual prognosis in injuries with IOFBs is as diverse as is its presentations. A number of authors found fairly good final VA in case series, with VA of 20/40 or better in 53.4–71% of cases (Colyer et al. 2007; Greven et al. 2000; Williams et al. 1988), although a large case series in China showed 17.1% with VA of 20/40 or better and 57.7% with VA worse than 4/200 including 7.53% that were enucleated or eviscerated (Zhang et al. 2011). Visual outcomes are usually limited when complications such as PVR arise (Rathod and Mieler 2011).

Proper care can be initiated by the physician with thorough history and physical examination, coupled with appropriate imaging techniques. Upon suspicion of an IOFB, it is recommended empirically initiating intravenous antibiotics. Expedited surgery to repair the globe architecture and remove IOFB is warranted, and the choice of surgical technique must encompass FB composition, size, and location (Loporchio et al. 2016). It is important to have adequate follow-up as severe complications may arise. With the improvement in management approaches, development of novel surgical techniques, and refinement of diagnostic imaging, the outcomes for patients with IOFBs hopefully will improve, and the increase in protective equipment usage is of the utmost importance for the decrease in the number of such preventable injuries.

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Optic Nerve Avulsion and Hemorrhagic Choroidal Detachment

10

Luis Filipe Nakayama

Optic Nerve Avulsion

Introduction

Optic nerve avulsion (ONA) is a rare condition with high morbidity that is characterized by mechanic disinsertion of optic nerve from ocular globe, with total or partial nerve fibers lesion (Foster et al. 1997), mainly associated with high-energy blunt ocular trauma. ONA is reported in automotive accidents, falls, and sport traumas (Chaudhry et al. 2006).

Etiopathogenesis

The etiopathogenesis of ONA involves forced ocular torsion after blunt trauma and ocular retropulsion. Other possible mechanism is optic nerve avulsion from the cribriform lamina after a subtle increase in intraocular pressure (Foster et al. 1997).

Clinical Features

Patients with ONA present with subtle variable visual acuity decrease, according to portion of optic nerve lesion (Foster et al. 1997). In the fundus examination, vitreous opacities may be present due to hemorrhage. In the retinal exam, it is possible to find sub- or intra- or preretinal hemorrhage adjacent to the region where the optic nerve should be inserted and optic disc pallor (Foster et al. 1997) (Fig. 10.1). Due to afferent neurologic lesion, the pupillary exam shows direct reflex commitment in ONA eye.

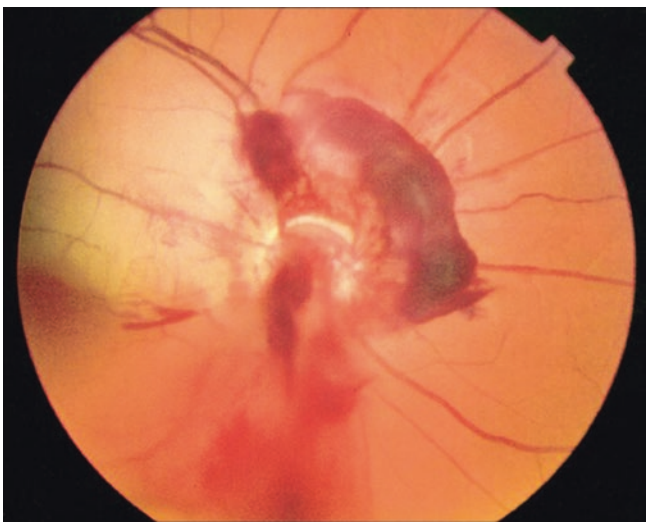

Fig. 10.1 Peripapillar hemorrhage in all retinal layers and adjacent retinal pallor, courtesy of S. Sandramouli

Management

Systemic

Other organs and systems should be evaluated according to trauma protocols because optic nerve avulsion is related to high-energy traumas.

Ophthalmological

Ocular ultrasonography may provide valuable information about ONA diagnosis when it is impossible to evaluate posterior segment due to vitreous hemorrhage (Sawhney et al. 2003).

Optic nerve avulsion does not have an effective ophthalmological treatment, and the prognosis is poor. Immediate recognition of the condition is important to avoid unnecessary procedures, exams, and treatment.

L. F. Nakayama (✉)
Department of Ophthalmology, São Paulo Federal University, São Paulo, Brazil

E. B. Rodrigues et al. (eds.), *Trauma and Miscellaneous Disorders in Retina*, Retina Atlas, https://doi.org/10.1007/978-981-13-8550-6_10

Hemorrhagic Choroidal Detachment

Introduction

Hemorrhagic choroidal detachment (HCD) is an uncommon condition characterized by blood accumulation at suprachoroidal space. It usually occurs after ophthalmological surgery, and it has poor prognosis.

The reported incidence of HCD is 0.003–0.06% after cataract phacoemulsification surgery, 0.15% after glaucoma filtering surgery, 0.45–1.08% after penetrating keratoplasty, and 0.17–1.9% after vitreoretinal surgery (Chu and Green 1999), and is directly related to surgery time and ocular hypotonia during procedure. In vitreoretinal surgery, hemorrhage may occur due to direct choroid or vorticosa vein trauma (Chu and Green 1999).

Etiopathogenesis

Hemorrhagic choroidal detachment etiopathogenesis involves choroid detachment from sclera due to hemorrhagic content. The proposed mechanism is short or long ciliary posterior artery rupture secondary to ocular hypotonia (Chu and Green 1999).

Proposed steps to the development of HCD involve: (1) choriocapillaris engorgement, (2) serous effusion at suprachoroidal space, (3) stretching and rupture of vessels at ciliary body base, and (4) bleeding from ciliary body and subsequent rupture of short and long posterior ciliary artery after blood accumulation.

Risk Factors

Hemorrhagic choroidal detachment risk factors can be divided into systemic, preoperative, and postoperative. Systemic risk factors include elderly, systemic arterial hypertension, arteriosclerosis, chronic renal disease, diabetes mellitus, and sanguineous dyscrasia.

Ophthalmological risk factors include primary open angle glaucoma, aphakia, increased axial length, intraocular inflammation, and vitreous loss (Chu and Green 1999; Wirostko et al. 1998; Hsiao et al. 2016). Perioperative risk factors are associated with conditions that lead to increase of episcleral pressure such as cough, nausea, vomits, and Valsalva maneuver. Postoperative risk factors include ocular hypotonia (Chu and Green 1999).

Diagnosis

Intraoperative diagnosis is based on ocular dynamic alterations such as increase of intraocular pressure, loss of red reflex, shallowing of anterior chamber, and cupuliform area (Fig. 10.2). Postoperative diagnosis is based on subtle decrease in visual acuity, ocular pain, shallowing of anterior chamber, loss of red reflex, and hemorrhagic choroidal detachment at retinal examination. Ocular ultrasound is a valuable ancillary exam when fundus examination is impossible due to vitreous opacities and to measure the choroidal detachment (Figs. 10.3 and 10.4).

Management

The most important intraoperative management consists of quickly closure of all incisions. Posterior sclerotomy is an alternative when it is impossible to replace the intraocular content, but with controversy results. Systemic hyperosmotic agents, sedatives, anti-hypertensive drugs to reduce systolic

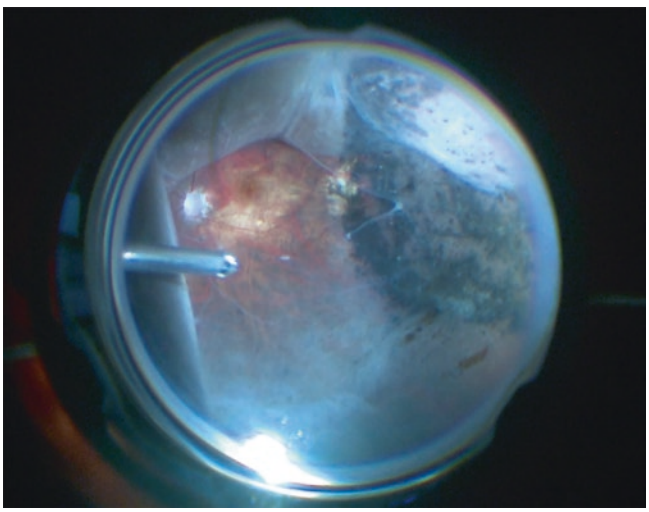

Fig. 10.2 Hemorrhagic choroidal detachment. Fundus with cicatricial retinal area, dense posterior vitreous and cupuliform lesion, courtesy of R. Brant

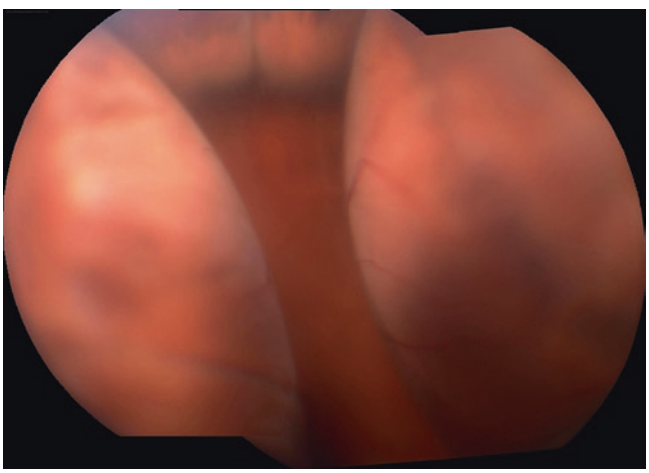

Fig. 10.3 Fundus color photography of choroidal detachment and almost appositional/kissing with serous and hemorrhagic content. Special thanks to Gabriel Andrande, from BRAVS

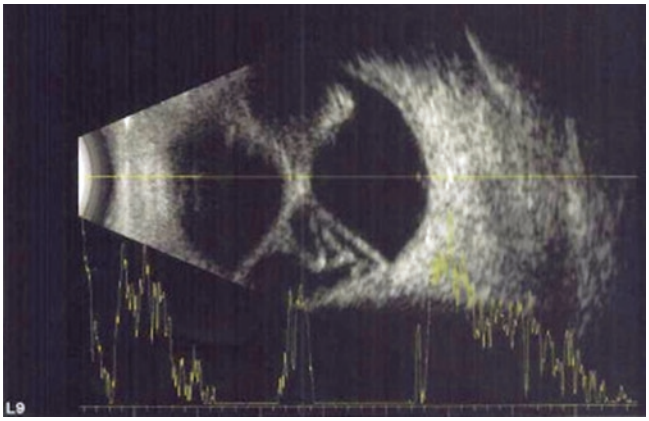

Fig. 10.4 Ultrasound scans of choroidal detachment and almost appositional/kissing with serous and hemorrhagic content. Special thanks to Gabriel Andrande, from BRAVS

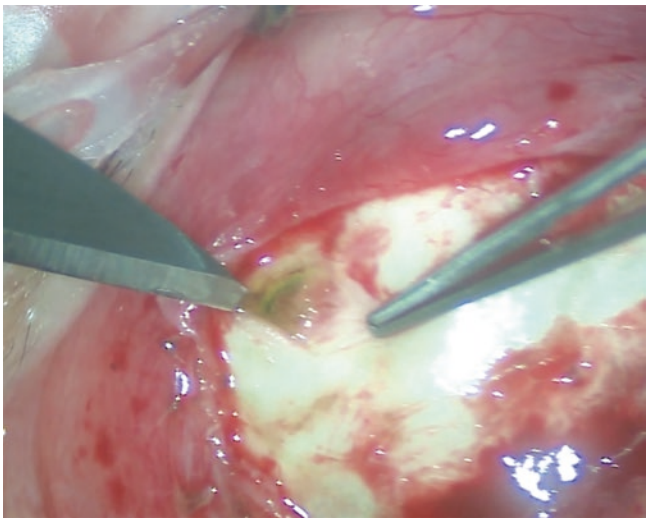

Fig. 10.5 Surgical drainage of hemorrhagic content—sclerotomy confection. Courtesy of R. Brant

arterial pressure may be used. Postoperative treatment consists of controlling the intraocular pressure, inflammation and pain. Corticoids could be used to control ocular inflammation.

Some signs contribute to surgical drainage approach decision such as concomitant retinal detachment, central retinal apposition, vitreous imprisoned at incision, increase of intraocular pressure, remain of lens material, uncontrollable ocular pain or when it is impossible to replace intraocular content (Chu and Green 1999; Wirostko et al. 1998; Figs. 10.5 and 10.6).

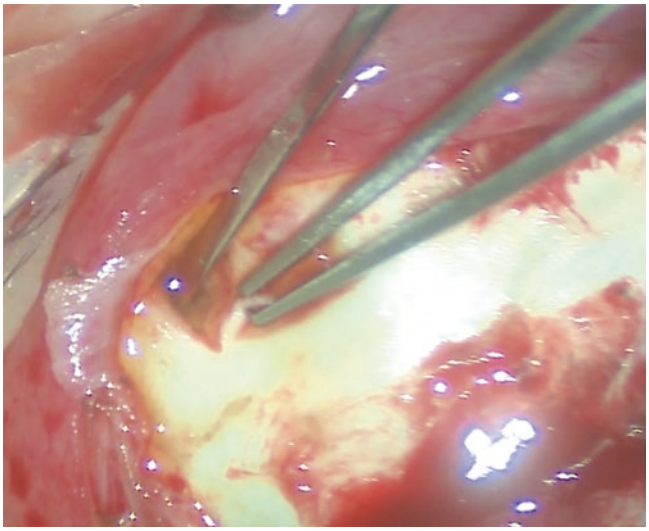

Fig. 10.6 Surgical drainage of hemorrhagic content—hemorrhage drainage. Courtesy of R. Brant

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Optic Disc Drusen

11

Nicolas Jean Paul Peres

Introduction

Optic disc drusen (ODD) are acellular bodies of hyaline deposits located both intracellularly and extracellularly first described by Muller in 1858 (Muller 1858; Hamann et al. 2018). The main clinical significance of optic disc drusen is that they can simulate true optic disc edema. Misdiagnosing drusen as true disc edema may lead to an extensive, invasive, and unnecessary systemic work-up, including neuroimaging and lumbar puncture. An ODD prevalence of 2.4% has been found in autopsy studies, though it has been reported to be significantly lower using in vivo imaging modalities. The prevalence of ODD is higher in women and involvement is usually bilateral (Lorentzen 1966; Friedman et al. 1975).

Etiopathogenesis

The pathogenesis of optic disc drusen formation has not been fully determined. The three classical theories on the formation of optic disc drusen postulate that they are caused by a disturbance in axonal metabolism with slowed axoplasmic flow (Tso 1981; Spencer 1978), vascular maldevelopment cause increased transudate release into the intercellular space which in turn acts as a nidus for the formation of ODD (Sacks et al. 1977), or a small scleral canal that physically compresses the optic nerve, causing ganglion cell death, with extrusion and calcification of mitochondria.

Clinical Features

Patients with optic disc drusen are frequently asymptomatic, and optic disc drusen are often discovered incidentally on ophthalmologic examination.

N. J. P. Peres (✉)
Department of Ophthalmology, Governador Celso Ramos Hospital, Florianópolis, SC, Brazil

Visual Field Defects

Optic disc drusen are associated with two major types of visual field defects. Axonal loss induced by direct compression or displacement of the fibers by the drusen could lead to arcuate defect or pseudopapilledema and the large protrusion induced by the optic disc drusen leading to an enlargement of the blind spot (Mustonen 1983; Wilkins and Pomeranz 2004; Katz and Pomeranz 2006).

The visual field defects may progress over time; however, it is slow. The prevalence of visual field defect varies greatly between studies: from 75% of patients showing no or minimal visual field defect (Lee and Zimmerman 2005) to 86% of patients showing visual field defects (Mustonen 1983). Such inconsistency may be attributed to the difference in the proportion of various types of ODD, with the highest occurrence in eyes with superficial optic disc drusen (Mustonen 1983; Savino et al. 1979).

Examinations Findings

The appearance of the optic nerve head is usually elevated, when the drusen are superficial they can be easily identified as round deposits with a lumpy bumpy appearance (Fig. 11.1); buried drusen are difficult to appreciate on fundoscopy examination but may sometimes be seen adjacent to vessels or the disc margin with oblique slit lamp illumination. Optic disc drusen are typically buried in the optic disc early in life and become more superficial later (Frisen 2008); therefore, in children, drusen are more likely to be buried and may be more difficult to detect (Erkkila 1977).

Diagnostic Technique

Various ancillary tests, including B-scan ultrasonography, fundus autofluorescence, fluorescein angiography, orbital computed tomography (CT scan), and more recently optical

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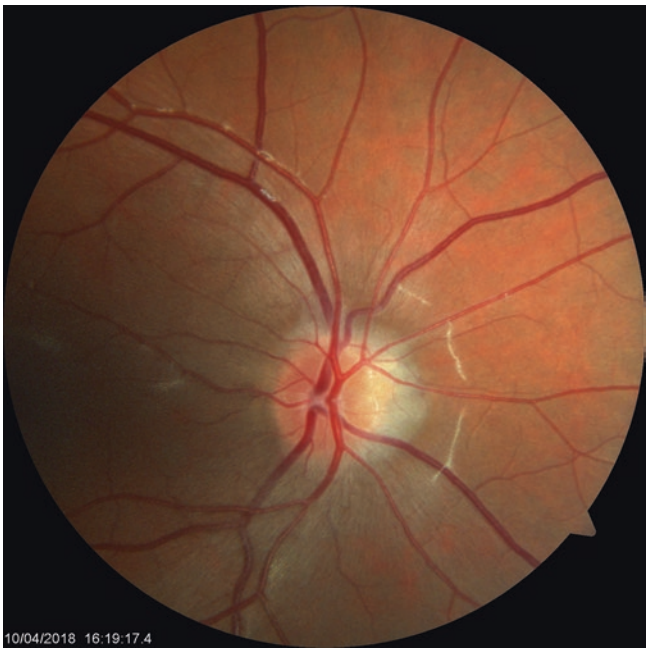

Fig. 11.1 Color fundus photograph of a 54-year-old man showing superficial optic disc drusen

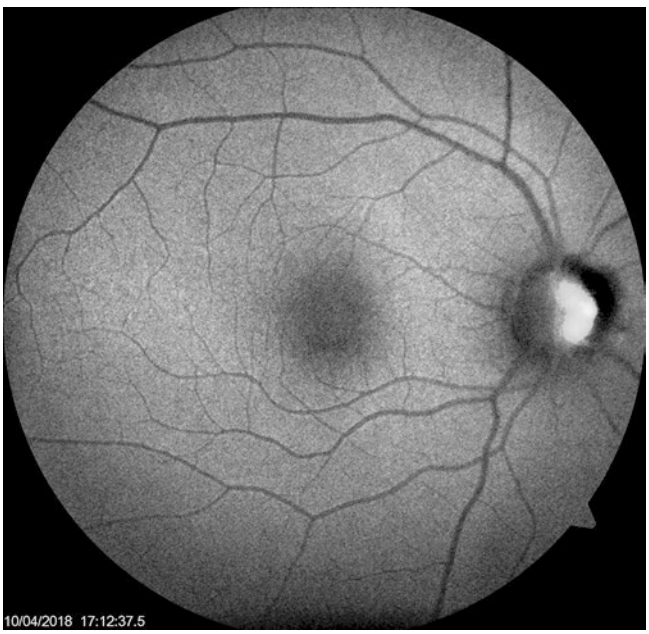

Fig. 11.2 Fundus autofluorescence showing hyperautofluorescence of optic disc drusen

coherence tomography (OCT), have been used to identify optic disc drusen (Mullie and Sanders 1985a, b; Sanders and Ffytche 1967; Pineles and Arnold 2012; Kurz-Levin and Landau 1999).

Fundus Autofluorescence

The autofluorescence characteristic of optic disc drusen (Fig. 11.2) can be helpful as a simple technique; however, it is only seen in superficial drusen and does not reliably detect buried drusen, possibly because of attenuation from overlying tissue. Kurz-Levin and Landau (1999) found that autofluorescence detected over 96% of superficial drusen but only 27% of buried drusen.

Fluorescein Angiography

Fluorescein angiography may be used to distinguish between optic disc drusen and true optic disc edema (Fig. 11.3a–c). Pineles was able to demonstrate that true disc edema is characterized by early or late disc leakage, while optic disc drusen displayed staining without leakage (Pineles and Arnold 2012).

B-scan Ultrasonography

B-scan ultrasonography is considered the gold standard imaging modality to detect optic disc drusen (Kurz-Levin and Landau 1999; Pineles and Arnold 2012; Atta 1988) (Fig. 11.4). Drusen characteristically appear hyperechoic with posterior shadowing on ultrasonography, due to the inherent high reflectivity of calcium. This technique is helpful in detecting buried drusen, but the echogenicity of drusen is dependent on the calcium content which also hampers the ability of CT scans to detect optic disc drusen.

Optical Coherence Tomography

A relatively new modality for imaging optic disc drusen is spectral-domain optical coherence tomography (SD-OCT) and, more recently, enhanced depth OCT. It became possible to image the acellular deposits in the optic nerve head with or without a border of high reflectance of calcification, and evaluate the interplay with other structures (Fig. 11.5a, b) (Heidary and Rizzo 2010; Kulkarni et al. 2014; Silverman et al. 2014).

Complications

Although optic disc drusen are typically considered benign, they may be associated with ocular complications.

Vascular anomalies appear to predispose patients with ODD to central and branch retinal artery occlusion and central retinal vein occlusion. Patients with ODD who suffer with these retinal vascular occlusions tend to be younger than individuals who do not have optic disc drusen. In most cases, these patients also had another systemic risk factor including hypertension, contraceptive use, atrioseptal defect, or migraine (Auw-Haedrich et al. 2002; Gifford 1895).

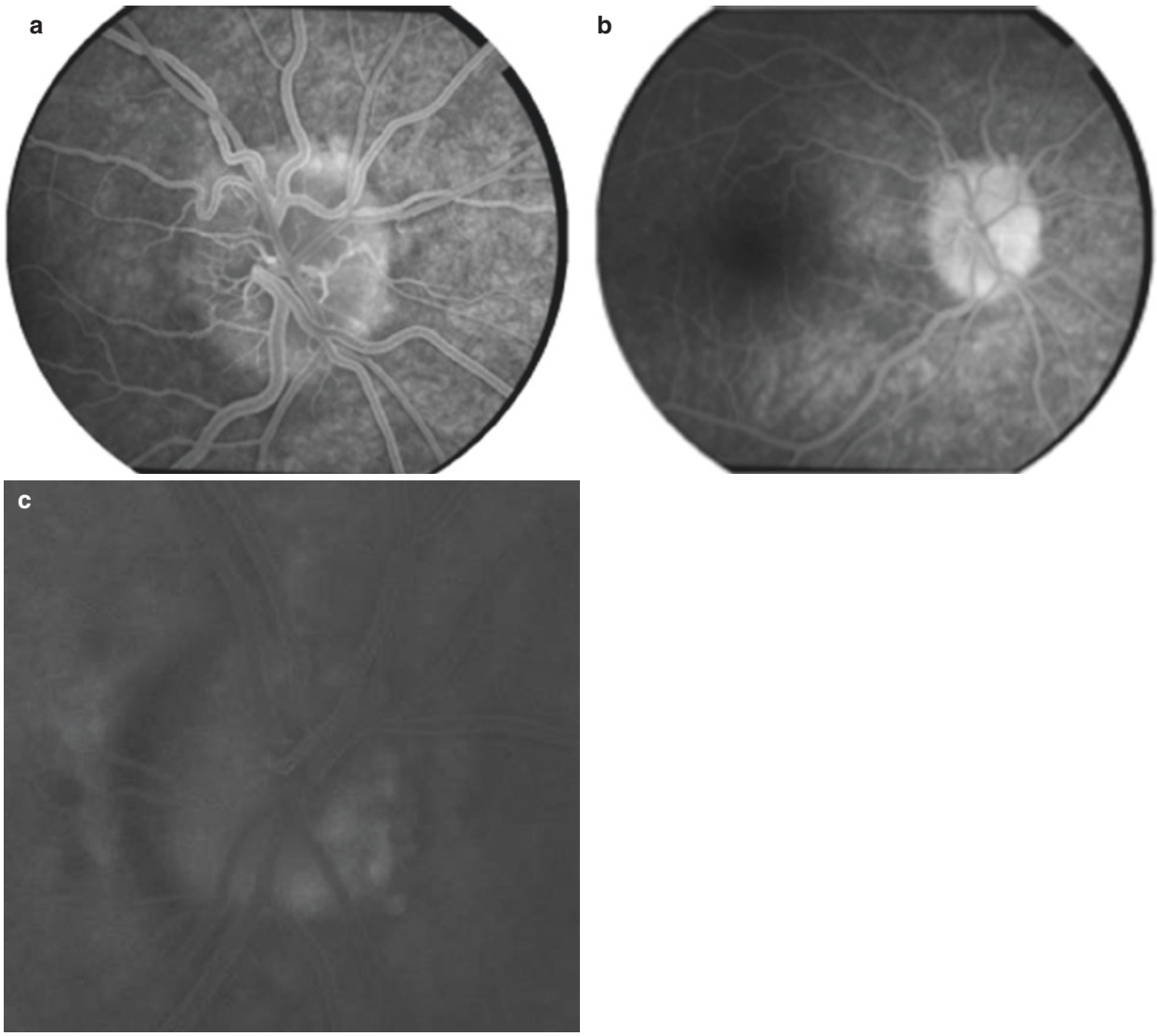

Fig. 11.3 (**a**). Early phase fluorescein angiography demonstrating nodular staining of the optic disc drusen. (**b**) Late phase fluorescein angiography demonstrating nodular staining of the optic discs with no leakage, same patient (**a**). (**c**) Late phase fluorescein angiography demonstrating nodular staining of the optic discs with no leakage, same patient (**b**)

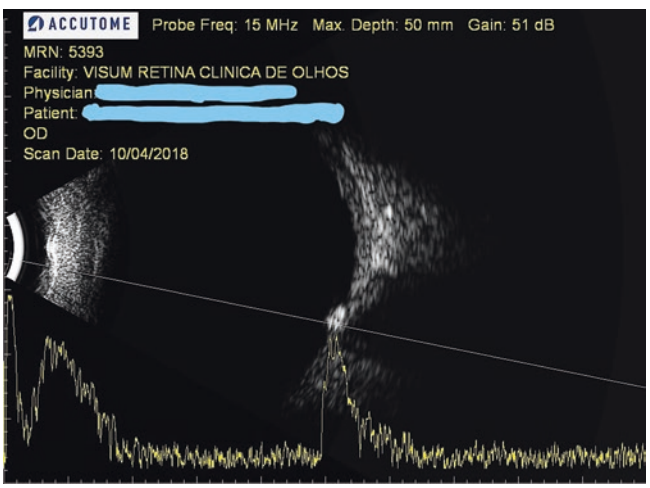

Fig. 11.4 Appearance of calcified optic disc drusen on ultrasound. The calcified drusen produce a hyperechoic signal, higher resolution is more successfully undertaken with lower decibel gain settings. Gain 51 dB

Choroidal neovascular membrane is also associated with ODD, the neovascular membrane is typically located in the peripapillary region, and is frequently associated with good visual acuity without treatment (Auw-Haedrich et al. 2002; Savino et al. 1979).

Management

If true optic disc edema has been ruled out, patients with asymptomatic optic disc drusen may be observed with serial visual field testing and regular examinations to ensure that treatable complications of ODD are appropriately managed. As the presence of ODD can predispose the patient to glaucomatous optic nerve damage at lower intraocular pressures and glaucoma may coexist with ODD, in the presence of visual field damage,

Fig. 11.5 (**a**) Color fundus, red-free, and OCT of a deep drusen and (**b**) another OCT view of the same patient

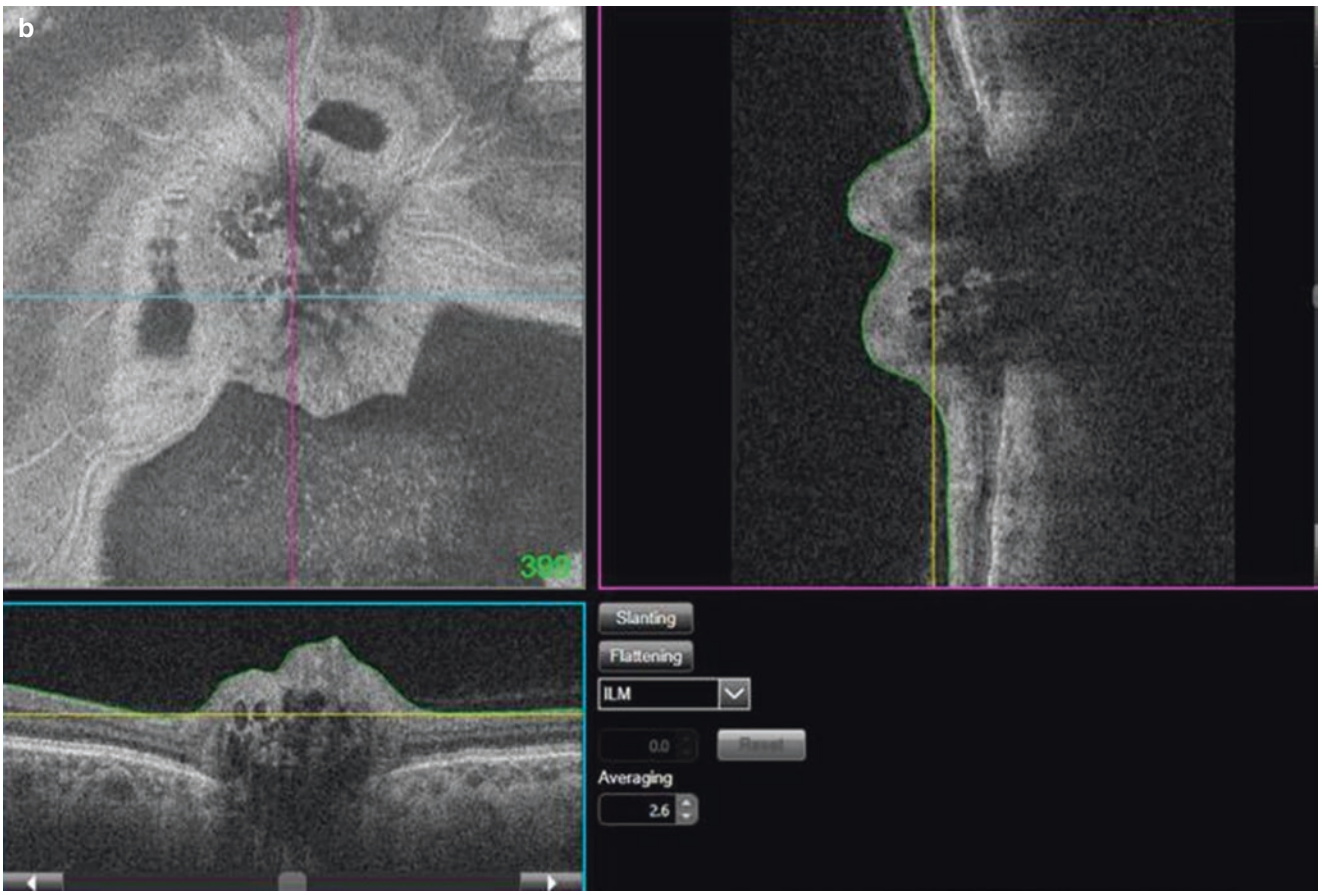

Fig. 11.5 (continued)

lowering the intraocular pressure should be considered (Samples et al. 1985).

Manage of vascular complications associated with ODD is similar to the management of these disorders in the absence of drusen. Treatment of neovascularization in ODD is not always required as the prognosis is usually good (Samples et al. 1985; Auw-Haedrich et al. 2002).

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Retinal Diseases in Pregnancy

12

Thaís Quisia Lima Gaspar, Eduardo Tomazoni, and Eduardo Büchele Rodrigues

Introduction

Pregnancy is a condition associated with high levels of estrogens that can cause a rapid progression of retinopathy in susceptible women or potentiate previous retinal diseases (Mallika et al. 2010, p. 1). Blood pressure typically declines early in pregnancy, peaks in the second trimester, with high levels of cortisol associated, and in the third trimester decreases slightly reaching supernormal levels. Another physiological alteration of pregnancy is the state of relative hypercoagulability of the pregnant woman, making her susceptible to the formation of pathological clots. These alterations can cause new retinal diseases or exacerbate pre-existing retinal disorders (Rosenthal and Johnson 2018, p. 2). Central serous chorioretinopathy may worsen with pre-eclampsia or occur without pre-eclampsia, probably because of increased levels of catecholamines during pregnancy. Increased levels of angiogenic factors also may contribute to the development of choroidal neovascularization (Errera et al. 2013, p. 5).

Retinal Diseases that May Be Induced by Pregnancy

Hypertensive Retinopathy and Choroidopathy

Pre-eclampsia is characterized by hypertension (systolic blood pressure ≥140 or diastolic blood pressure ≥90 mmHg, measured in a previous normotensive female patient), peripheral edema, and proteinuria, while eclampsia is defined as pre-eclampsia plus seizures (Andrada Paun et al. 2017, p. 6). Fundus findings of hypertensive retinopathy include arteriolar constriction, retinal hemorrhages, cotton wool spots, retinal edema, and lipid exudates. Severe cases may include subretinal fluid resulting from choriocapillaris infarction and/or optic disc swelling and ischemia. These findings on fundus examination indicate that it is necessary to conduct the treatment of hypertension immediately. In cases of ischemia, we can find retinal thinning or chorioretinal atrophy (Rosenthal and Johnson 2018, p. 2).

Exudative Retinal Detachment

May occur in HELLP syndrome (hemolysis, elevated liver enzymes, low platelets), disseminated intravascular coagulation, and thrombotic thrombocytopenic purpura (Rosenthal and Johnson 2018, p. 2). In these cases, retinal detachment is a consequence of choroidal vascular damage in the choriocapillaris layer, and then occurs the accumulation of subretinal fluid with posterior damage to the retinal pigment epithelium. As these situations are considered emergencies, a decision of cesarean along with a timely ophthalmologic approach would provide the best ophthalmological prognosis for the patient (Andrada Paun et al. 2017, p. 6).

Retinal Vascular Occlusive Disease

Hypercoagulability is a result of modifications of platelets, clotting factors, and blood flow hemodynamics fluctuations. The occlusion of the central retinal artery or cilioretinal artery is a possible condition during gestation with evident hypercoagulability and, if bilaterally occurring, there is a greater chance of being caused by hypercoagulable diseases. Occlusion of the central retinal vein is more common in diabetic and smoker patients who already have atherosclerosis (Andrada Paun et al. 2017, p. 6), and generally occurs in the

T. Q. L. Gaspar · E. Tomazoni (✉)
Ophthalmology, Retina Fellowship, Visum Retina, Florianópolis, Santa Catarina, Brazil

E. B. Rodrigues
Department of Ophthalmology and Visual Sciences, Federal University of São Paulo, São Paulo, Brazil

Department of Ophthalmology, Saint Louis University School of Medicine, Saint Louis, MO, USA

E. B. Rodrigues et al. (eds.), *Trauma and Miscellaneous Disorders in Retina*, Retina Atlas, https://doi.org/10.1007/978-981-13-8550-6_12

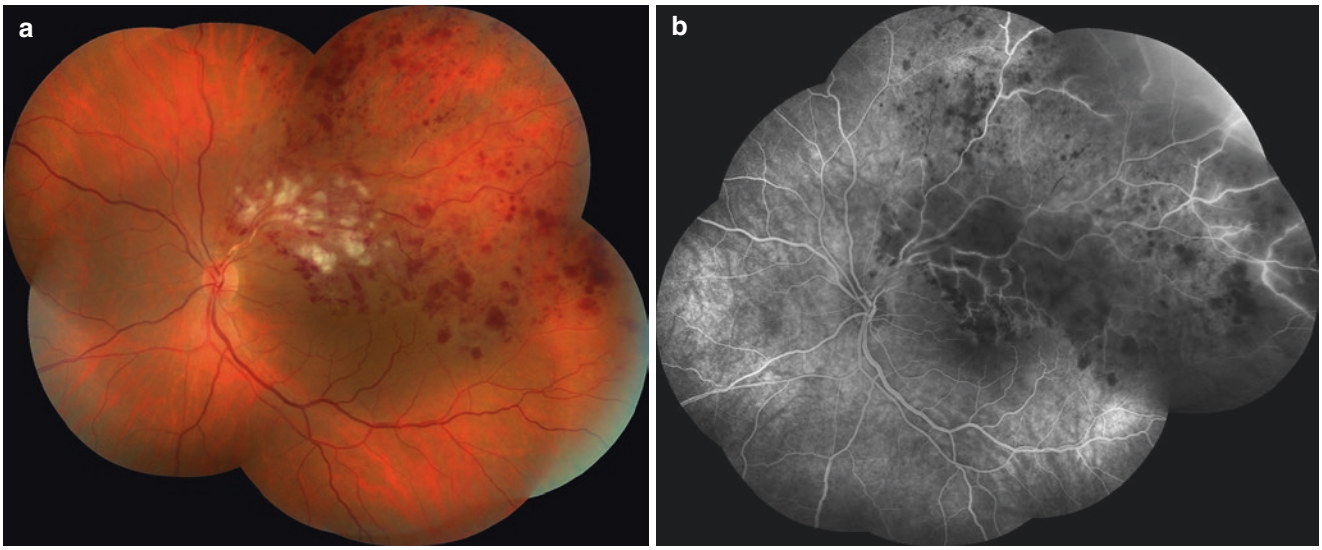

Fig. 12.1 (**a**) Retinography showing occlusion of superior temporal venous branch. (**b**) Fluorescein angiography of the same patient

third trimester or post-partum period (Rosenthal and Johnson 2018, p. 2) (Fig. 12.1a, b).

Retinal Diseases That May Be Worsened by Pregnancy

Central Serous Chorioretinopathy

Central serous chorioretinopathy (CSC) is characterized by an accumulation of subretinal fluid resulting in a circumscribed neurosensory retinal detachment. Pregnancy is a risk factor for active episodes of idiopathic CSC, probably due to the high serum cortisol levels of the pregnant (Errera et al. 2013, p. 5). The symptomatology consists in metamorphopsia, visual loss, scotomas, and light sensitivity (Andrada Paun et al. 2017, p. 6). In pregnant patients with idiopathic CSC, subretinal fibrin deposition occurs in up to 90%, compared to 20% of nonpregnant patients. Management of these patients can be challenging, as fluorescein angiography and photodynamic therapy are not well studied in pregnancy, and most physicians therefore prefer to avoid them. We recommend that if no fibrin is present in or near the fovea, and the patient is near term, the patient can be observed with an expectation of resolution after delivery. For a patient whose fovea is threatened by fibrin, it is reasonable to consider thermal laser photocoagulation guided by fundus examination and optical coherence tomography (OCT) imaging (Rosenthal and Johnson 2018, p. 2) (Fig. 12.2a, b).

Diabetic Retinopathy

Gestational diabetes (GDM) is defined as insulin resistance causing hyperglycemia that develops or is first diagnosed during pregnancy. Five to ten percent of GDM will persist following pregnancy and be classified as diabetes mellitus type 2 (T2DM). As the glucose intolerance in GDM is transient in most of cases, these women are not generally at risk of developing DR during pregnancy. However, a proportion of women with GDM do in fact have undiagnosed T2DM. This subgroup may develop DR during or following pregnancy, that is an independent risk factor for worsening of DR. Other risk factors for progression during pregnancy are like those for the general diabetic population (i.e., poor glycemic control, duration of diabetes, hypertension, and others; Morrison et al. 2016, p. 4). The severity of retinopathy at baseline significantly increases the risk for progression, especially in the second and third trimesters, and is uncertain in the postpartum. The long-term risk for progression of DR does not appear to be increased by pregnancy. In cases of progression to proliferative diabetic retinopathy (PDR) during pregnancy, regression after delivery may occur (Errera et al. 2013, p. 5).

The mechanisms underlying the possible exacerbation of DR during pregnancy are unclear, but hematologic, hormonal, metabolic, cardiovascular, and immunologic changes all play a role. Endothelial cell damage at the capillary level because of increased retinal blood flow may cause progression of retinopathy in pregnant diabetic women, even when good metabolic control is achieved, and baseline retinopathy is minimal. In addition, a number of studies have found that sudden improvement in glycemic control induces a decrease in retinal blood flow, secondary hypoxia, and thus possible worsening of retinopathy. During pregnancy, the synthesis of many angiopoietic factors is increased. Progesterone, for instance, may induce the local production of VEGF in addition to other intraocular angiopoietic factors involved in the development of the microvascular changes observed in DR during pregnancy (Errera et al. 2013, p. 5) (Fig. 12.3).

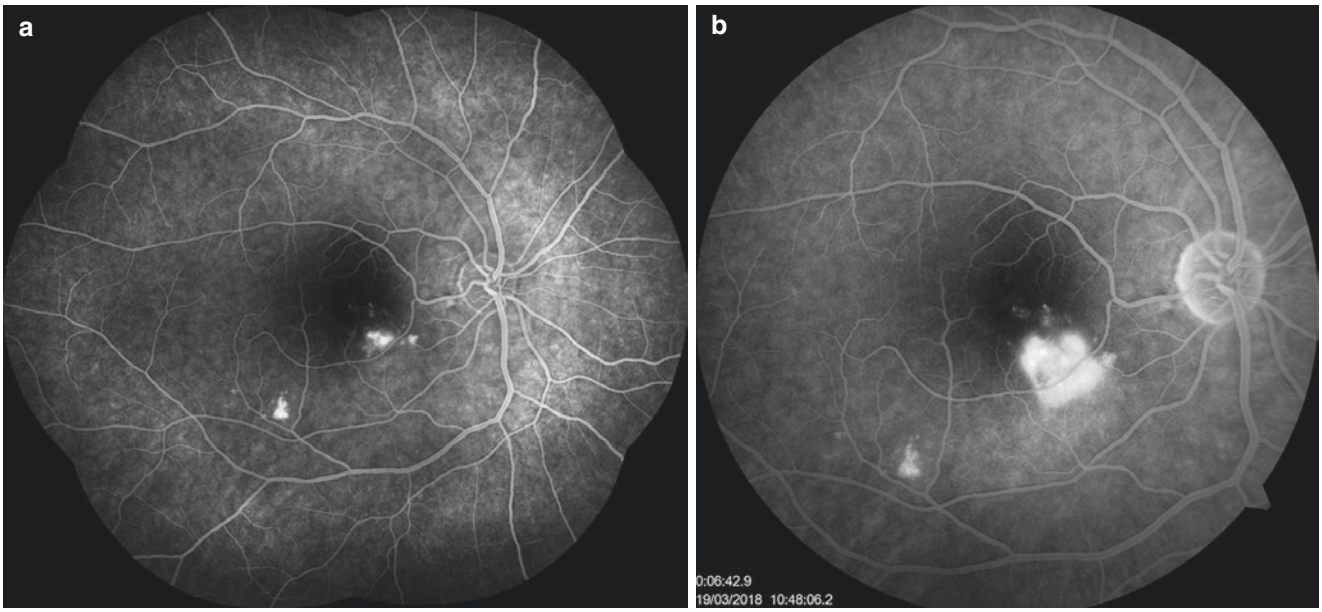

Fig. 12.2 (**a**) Fluorescein angiography at initial phase in a patient with central serous chorioretinopathy. (**b**) Fluorescein angiography at late phase

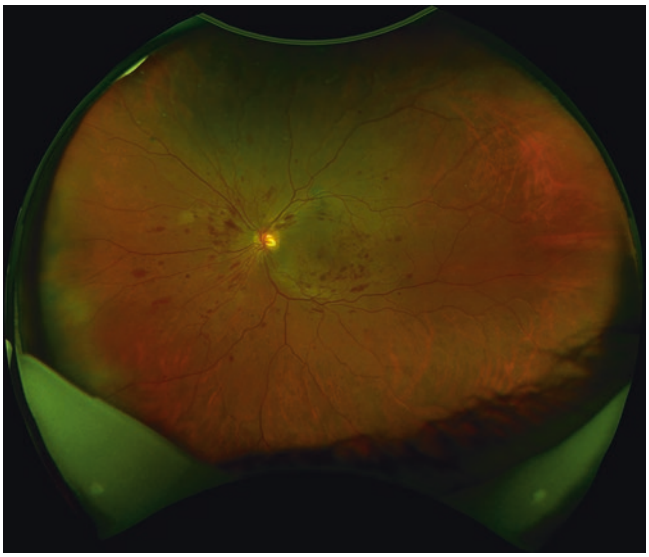

Fig. 12.3 Optomap retinal imaging showing NPDR in a pregnant patient

A preconception ophthalmic examination and a first trimester eye examination are recommended by all guidelines (Morrison et al. 2016, p. 4). Individuals with moderate nonproliferative diabetic retinopathy (NPDR) should be reexamined every 3–6 months and those with severe NPDR or worse should be examined every 1–3 months. Heightened surveillance should continue during the first year postpartum (Rosenthal and Johnson 2018, p. 2).

In patients who present with severe NPDR and proliferative retinopathy during pregnancy, pan-retinal photocoagulation (PRP) is safe and has been found to be effective to reduce visual impairment (Morrison et al. 2016, p. 4). Observation is a reasonable management option for pregnant patients with mild diabetic macular edema (DME), since the edema may well resolve after delivery. In cases of DME requiring treatment, Rosenthal and Johnson believe that anti-VEGF therapy should be avoided in favor of focal laser photocoagulation or intravitreal corticosteroid injections, but Morrison et al. reported that the anti-VEGF medications ranibizumab (Lucentis) and Aflibercept (Eylea) given intravitreally have been assigned pregnancy category C by the FDA. If the intravitreal administration limits systemic exposure, the treatment may pose a risk to malformations and fetal abnormalities. It is therefore recommended that women wait at least 3 months after their last treatment with these agents prior to conceiving. The same results have been seen with bevacizumab (Avastin).

Recommendations

Angiography in Pregnancy

Fluorescein dye may cross the placenta and is present in breast milk for up to 72 h after the procedure. However, there are no reports of adverse fetal or infant effects, but is prudent use OCT and/or OCT angiography in place of invasive angiography when is possible (Rosenthal and Johnson 2018, p. 2). Fluorescein dye should be particularly avoided in the first trimester (Morrison et al. 2016, p. 4).

Ocular Medications

Due to most of the topical medications did not have a conclusive study about the teratogenicity, these medications are generally avoided. Some ocular medications, however, are known to be deleterious in pregnancy, for example, acetazolamide has been associated with neonatal acidosis, prostaglandin analogues are considered pregnancy category C (insufficient research data to determine safety), and beta-blockers may be teratogenic (Rosenthal and Johnson 2018, p. 2). We suggest good medical practice, using recent research on the drug before prescribing it. For cases where the drug is prescribed, patient can minimize systemic drug absorption by compressing nasolacrimal duct and closing the eyelids for some minutes.

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Cancer Therapy-Associated Retinopathy

13

Valeska Aparecida Riguette Costa, Eduardo Tomazoni, and Eduardo Büchele Rodrigues

Introduction

There are two types of cancer therapy-associated retinopathy: associated with radiation or caused by drugs. Treatment in most cases involves suspension or substitution of the drug and depends on the type of the cancer and how advanced it is.

Radiation Retinopathy

Radiation retinopathy is a slowly progressive delayed-onset occlusive microangiopathy of the retinal vasculature that occurs with variable latency after exposure of the retina to ionizing radiation. The term encompasses all retinal vascular changes, including ischemic and proliferative radiation retinopathy and radiation maculopathy (Fig. 13.1). It is a potentially devastating sequela of exposure of the eye to any source of radiation, including local plaque radiation treatment (brachytherapy, Fig. 13.2), external-beam radiation treatment, proton beam radiation, helium ion radiotherapy and gamma knife radiotherapy of the eye, ocular adnexa, orbit, and head and neck structures (Mittra 2013). Gupta et al. (2007) reported that the incidence of radiation retinopathy was significantly higher in patients with diabetes mellitus, hypertension, or who received previous chemotherapy. Factors that influence radiation retinopathy are total dosage and daily fraction size.

V. A. R. Costa · E. Tomazoni (✉)
Ophthalmology, Retina Fellowship, Visum Retina, Florianópolis, Santa Catarina, Brazil

E. B. Rodrigues
Department of Ophthalmology and Visual Sciences, Federal University of São Paulo, São Paulo, Brazil

Department of Ophthalmology, Saint Louis University School of Medicine, Saint Louis, MO, USA

Drug-Induced Toxic Retinopathy

A series of oncology drugs may induce secondary retinal changes. In this chapter, we are going to focus on those better studied so far.

Tamoxifen

Tamoxifen is one of the most common chemotherapy systemic drugs that ophthalmologists encounter as a source of ocular toxicity. It is an antiestrogenic drug used as a coadjutant treatment in breast cancer. The risk of ocular toxicity exists in 1–2% of patients on standard tamoxifen dosing. There are two types of presentation: acute form or long-term use form. Acute form presents with white intraretinal crystalline deposits, macular edema, and punctate retinal pigmentary changes. This form is reversible upon discontinuation of the drug. Long-term use of tamoxifen-induced retinopathy occurs in patients taking a cumulative daily dose exceeding 100 mg or more than 1 year of therapy. This other form presents with crystalline maculopathy, consisting of crystalline refractive intraretinal deposits as a result of axonal degeneration in the perifoveal area, and may remain even with the suspension of the drug. Spectral-domain OCT can be of value as a screening tool (Gagnon 2013; Mittra 2013).

Interferons

Interferons (IFNs) are a class of cytokine with ability to inhibit cell proliferation and modulate viral replication functions of the immune system. IFN is the most relevant adjuvant treatment for patients with cutaneous melanoma. Although generally well tolerated, incidences of IFN toxicity are commonly reported. Interferon-associated retinopathy is typically characterized by retinal hemorrhages and cotton wool spots in the posterior pole with visual function usually maintained. Macular edema with reduced visual acuity may occur, as well asymptomatic patients may develop ischemic retinopathy. These retinal changes are usually reversible with discontinuation of therapy. To identify retinal toxicity at its

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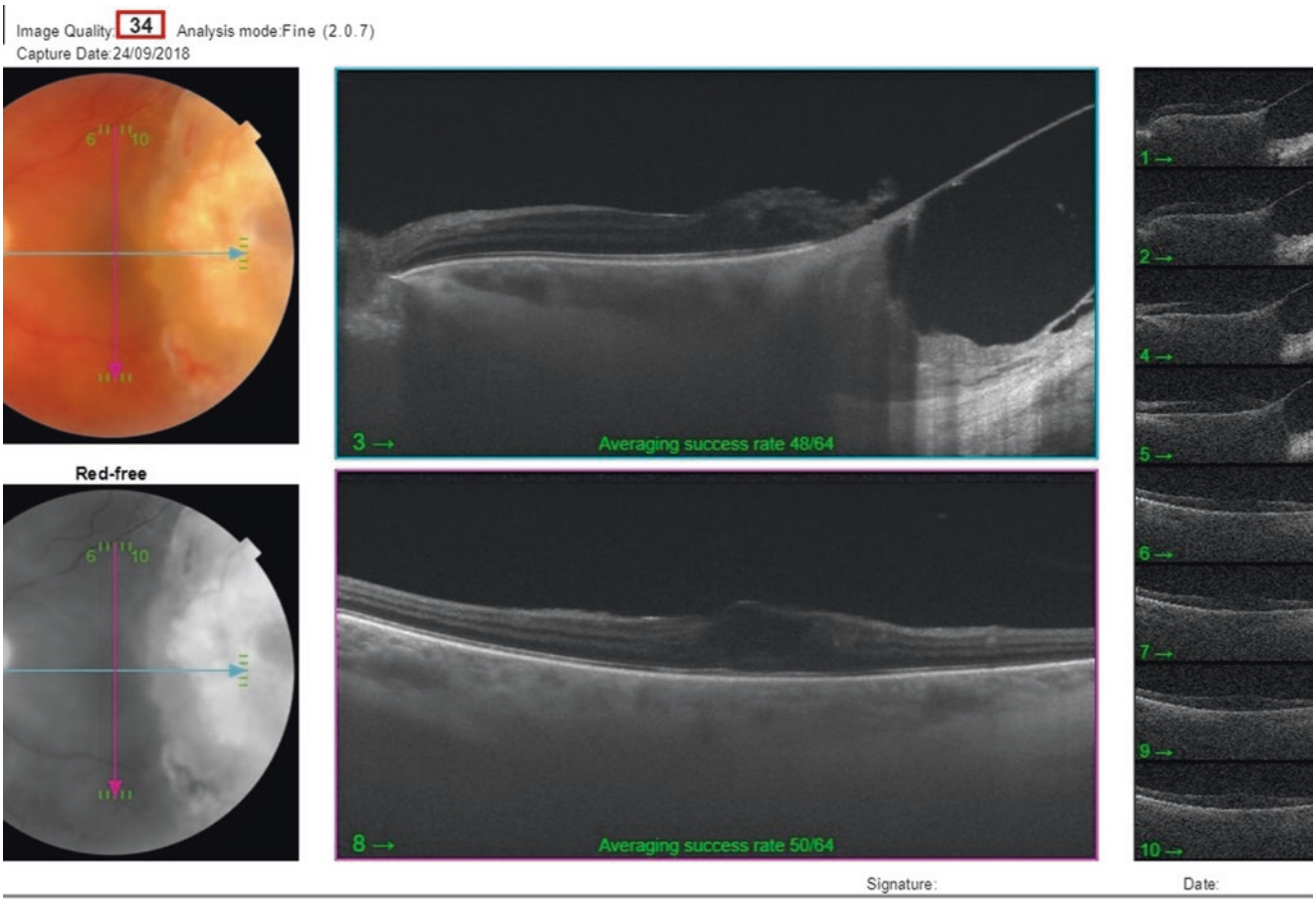

Fig. 13.1 Residual radiation macular edema as consequence of brachytherapy for management of choroidal melanoma—optical coherence tomographic

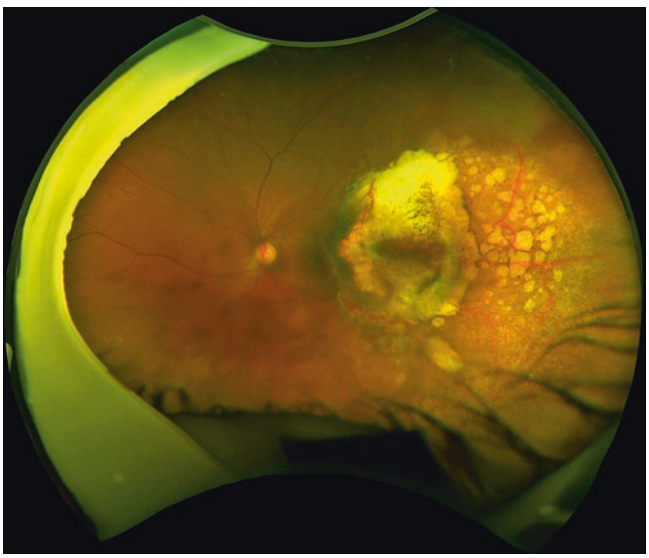

Fig. 13.2 Same patient of Fig. 13.1 showing devastating sequela—color retinography

earliest stages, fundoscopic exam is necessary at baseline and during follow-up every 3 months (Borrego-Sanz et al. 2014; Omoti and Omoti 2006).

Cisplatin

Cisplatin is a chemotherapy drug for a variety of solid tumors, metastases, and small cell cancers with unknown primary tumors (Borrego-Sanz et al. 2014). Visual loss may be bilateral and irreversible, and visual fields show bilateral central scotomas. Visual evoked response and electroretinogram are used to document the retinotoxicity of these agents (Omoti and Omoti 2006; Mittra 2013).

Etiopathogenesis

Radiation Retinopathy

Studies have demonstrated that radiation exposure results in the preferential loss of vascular endothelial cells, while spar-

ing pericytes. This may be related to the direct exposure of endothelial cells to high ambient oxygen and iron from the blood that generates free radicals and damages cell membranes (Murray 2013).

Drug-Induced Toxic Retinopathy

Tamoxifen

Tamoxifen apparently contribute to accumulate crystalline retinal deposits that cause axonal degeneration. This hypothesis is supported by the intracellular location of the retinal lesions in the nerve fiber layer and inner plexiform layer of the retina, which reacted on tests with glycosaminoglycans. It has also been postulated that tamoxifen binds with polar lipids, inhibiting normal catabolism of this substance and resulting in accumulation of drug–lipid complexes in lysosomes (Srikantia et al. 2010).

Interferons

It is believed that interferons cause an increase in immune complex deposition and activated complement C5a with leukocyte infiltration. EOG testing may become abnormal in early toxicity (Borrego-Sanz et al. 2014).

Cisplatin

Cisplatin apparently induce platinum-containing agents to bind DNA causing conformational changes that interfere with transcription and replication resulting in apoptosis (Langevin et al. 2017).

Clinical Features

Radiation Retinopathy

The clinical features of radiation retinopathy include microaneurysms, cotton wool spots, capillary dilation, telangiectasia, and capillary closure (Fig. 13.3). The posterior retina is more sensitive to radiation than peripheral retina. Vascular compromise may result in retinal edema. Ischemia may lead to disc neovascularization which in turn can cause vitreous hemorrhage and retinal detachment. Histologically, there is a thickening of the wall of the arterioles and capillaries and loss of endothelial cells, sparing the pericytes.

A reported safe dose is 3000 rads/30 gray, 1000 rads/10 gray per week in five fractions (200 rads/2 gray per session), although cases have been reported with lower doses of radiotherapy. The time of onset of radiation retinopathy is between 6 months and 3 years, with a few cases occurring earlier or later.

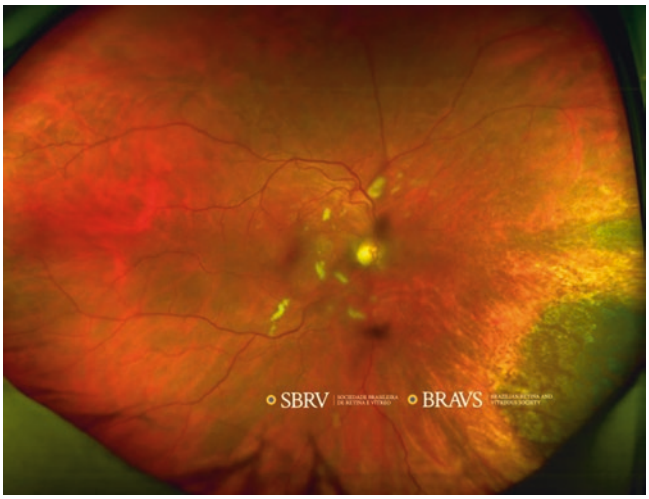

Fig. 13.3 Right eye of a 72-year-old patient with choroidal melanoma treated with brachytherapy 1 year ago. Retinal atrophy nasal inferior corresponding to the treated area. In posterior pole, we find hemorrhages, macular edema, and cotton wool spots—color retinography. Special thanks to Gabriel Andrade

Factors which exacerbate radiation retinopathy include chemotherapy, vascular diseases (like diabetes and hypertension), and possibly by pregnancy.

Cataracts and radiation retinopathy are the most common visually limiting complications seen after ophthalmic plaque radiation therapy. The cataracts are amenable to surgical treatment mostly leading to improvement in vision. However, retinopathy can lead to permanent and severe visual loss (Gupta et al. 2007).

Drug-Induced Toxic Retinopathy

Tamoxifen

Tamoxifen acute retinal toxicity consists of decreased visual acuity and color vision with white intraretinal crystalline deposits, macular edema, and punctate retinal pigmentary changes. Crystalline maculopathy usually occurs in patients who use long-term and high-dose tamoxifen and consists of refractile intraretinal crystalline deposits concentrated primarily in the perifoveal area (Fig. 13.4). Visual acuity decreases are usually secondary to foveal cyst development. Less extensive retinal changes may occur in patients receiving low doses for long periods, and isolated retinal crystals may be observed in patients without visual symptoms (Mittra 2013; Srikantia et al. 2010).

Interferons

Interferon therapy has been associated with the development of multiple cotton–wool spots, retinal hemorrhages, optic

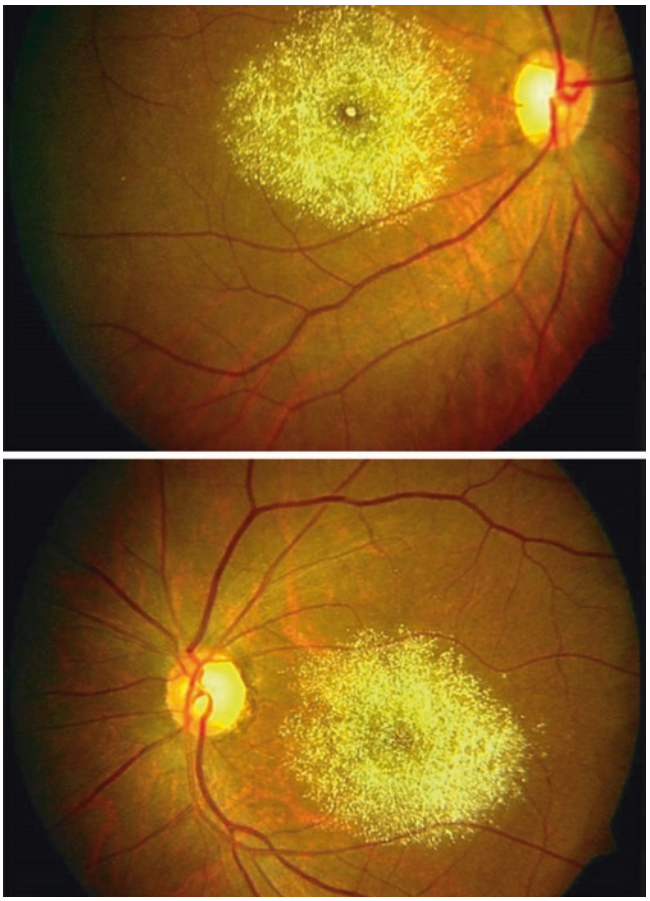

Fig. 13.4 Crystalline maculopathy: refractile intraretinal crystalline deposits concentrated primarily in the perifoveal area

disc edema, branch arterial and venous occlusion, central retinal venous obstruction, anterior ischemic optic neuropathy, and cystoid macular edema. Visual acuity usually is not affected in mild cases presenting with cotton wool spots and intraretinal hemorrhage. Changes are noted within the first 4–8 weeks of therapy and are seen more frequently in diabetic and hypertensive patients.

Cisplatin

Cisplatin may cause three main different types of retinal toxicity. First type consists of a pigmentary retinopathy of the macula with markedly decreased visual acuity and frequently abnormal electrophysiologic testing. This pigmentary change has been reported after administration of intra-arterial cisplatin for malignant glioma treatment (Mittra 2013). A second type of retinopathy has been described and consists of cotton wool spots, intraretinal hemorrhages, macular exudate, and optic neuropathy with disc swelling. This condition was reported in cases with high dose of chemotherapy with cisplatin, cyclophosphamide, carmustine, and autologous bone marrow transplantation for metastatic breast cancer. The third type of change involves a vascular retinopathy or optic neuropathy, which can include arterial occlusion, vasculitis, and papillitis. Type three retinal toxicity has been seen in approximately 65% of patients receiving intra-arterial carmustine isolated or combined with cisplatin for malignant glioma. These changes are associated with a profound visual loss that begins about 6 weeks after the start of the therapy. Other ocular effects may include orbital pain, chemosis, secondary glaucoma, internal ophthalmoplegia, and cavernous sinus syndrome.

Management

Radiation Retinopathy

Currently no guidelines or treatment exists for radiation retinopathy. Pan retinal photocoagulation is performed in the proliferative stage to prevent further visual loss, although studies have shown that earlier intervention may be more beneficial in preserving vision. There are studies which have reported a temporary improvement in vision after using intravitreal triamcinolone or focal laser treatment for cases with radiation maculopathy. Spontaneous improvement can occur, but this is infrequent (Gupta et al. 2007).

Drug-Induced Toxic Retinopathy

Tamoxifen

The development of actual crystals by tamoxifen is likely not associated with vision loss, so cessation is not required. Any associated vision loss that ensues is an indication to discontinue therapy. The presence of microcystoid foveal changes on OCT and significant color vision loss are both indications to cease tamoxifen (Gagnon 2013). For persistent macular edema, which has been noted after prolonged high-dose therapy after cessation of the drug, antivascular endothelial growth factor injection may be beneficial (Mittra 2013).

Interferon

After discontinuation of interferon therapy, most of the retinal lesions have spontaneously disappeared, given the reversibility of IFN-associated side effects reported in multiple studies (Borrego-Sanz et al. 2014).

Cisplatin

The visual loss with cisplatin usually is progressive, and no treatment is known (Langevin et al. 2017).

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Asteroid Hyalosis and Synchysis Scintillans

14

Mariana Batista Gonçalves

Asteroid Hyalosis

Introduction

Vitreous opacities may be the result of developmental abnormalities, degenerative processes, inflammation, intraocular bleeding, and trauma (Kador and Wyman 2008). Among the vitreous opacities, we can mention asteroid hyalosis (AH), characterized by the presence of small spherical opacities of yellowish-white color, that move according to the ocular movements, returning to its original position after the interruption of the movement (Figs. 14.1, 14.2 and 14.3; Potter 1983).

The origin of the term "asteroid hyalosis" comes from 1894, when Benson described the case of a patient with yellowish-white spheres in the right eye, whose appearance of the particles in the vitreous resembled the appearance of "stars on a clear night." He then used the term "asteroid" to refer to stars and called the condition "asteroid hyalitis" (Benson 1894; Potter 1983).

Demographic Data

Two large population studies, the Blue Mountain Study and the Beaver Dam Eye Study, were executed to determine the prevalence of AH as well as other demographic characteristics of this condition. In these studies, the overall prevalence of asteroid hyalosis ranged from 1% to 1.2%, increasing significantly with age. In the Blue Mountain Study, which included patients over 49 years of age living in the Blue Mountain region of Australia, the prevalence of AH was 0% in people under 55 years of age, 0.4% in persons aged 55–64 years, 1.1% in people aged 65–74 years, and 2.1% in people aged 75 years and over (Mitchell et al. 2003; Moss

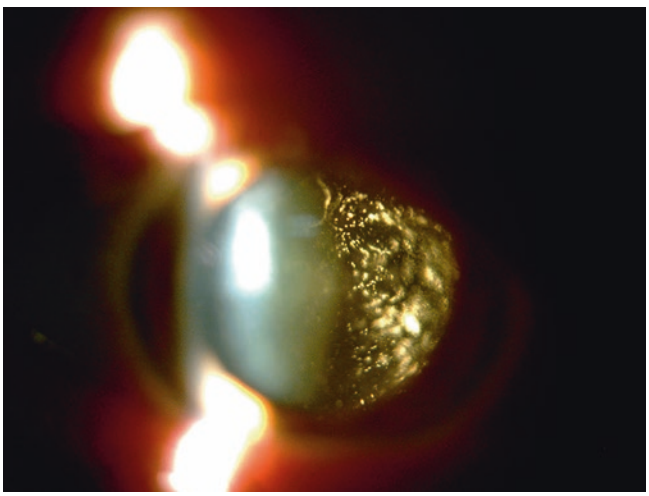

Fig. 14.1 Slit-lamp photograph of a patient with asteroid hyalosis demonstrates small spherical opacities, of yellowish-white color in the anterior vitreous

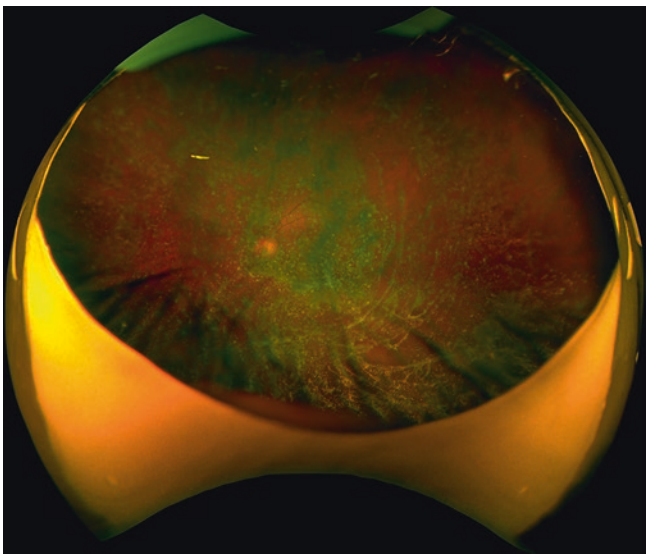

Fig. 14.2 Optomap imaging shows asteroid bodies within the vitreous. Although this condition often makes difficult the fundus examination, typically this disorder does not produce reduction in visual acuity

M. B. Gonçalves (✉)
Department of Ophthalmology, São Paulo Federal University, São Paulo, Brazil

E. B. Rodrigues et al. (eds.), *Trauma and Miscellaneous Disorders in Retina*, Retina Atlas, https://doi.org/10.1007/978-981-13-8550-6_14

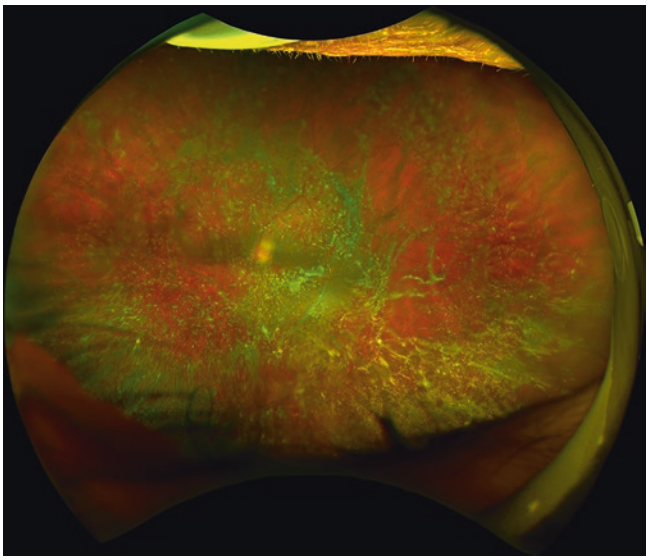

Fig. 14.3 Another patient with the same condition, with imaging also captured by Optomap

et al. 2001). Another study conducted at the University of California analyzed the presence of asteroid hyalosis in eyes undergoing autopsy and found a prevalence of 1.96% as well as a statistically significant correlation of this condition with the age of the patients (Fawzi et al. 2005).

Asteroid hyalosis is more common in men and usually has a unilateral presentation. A significant difference was observed in the Blue Mountain Study with regard to gender, with a prevalence of 1.4% in men and 0.6% in women. Regarding laterality, the Beaver Dam Eye Study reported a prevalence of 9% of bilateral cases, but this rate varies from 7% to 22% in the literature (Mitchell et al. 2003; Moss et al. 2001).

Pathogenesis

The etiology of asteroid hyalosis remains uncertain, but previous reports associate this condition with hyperopia, diabetes mellitus, hypercholesterolemia, hypertension, gout, and increased serum calcium levels. Most of these studies were case series or case–control studies. Because they do not always represent the general population, such studies may contain biases, and are not as reliable as population studies to establish such associations (Moss et al. 2001).

In the Beaver Dam Eye Study, an association between AH and body mass index, alcohol consumption, and gout history was observed, but associations with diabetes mellitus and hypertension were not observed (Moss et al. 2001). Similarly, the Blue Mountain Study found no associations with hypertension and diabetes (Mitchell et al. 2003). The UCLA study found a statistically significant association between asteroid hyalosis and age-related macular disease, hypertension, atherosclerosis, and posterior vitreous detachment (PVD). However, after adjusting this data for age in a multivariate logistic regression analysis, only association with vitreous detachment was maintained (inversely correlated). Association between asteroid hyaloid and diabetes mellitus was also not observed in this study (Fawzi et al. 2005).

Composition

The asteroid bodies are composed of lipids, calcium, and phosphorus. A study conducted by Miller et al. (1983) showed, through transmission electron microscopy, that such particles are composed of a regular arrangement of parallel lamellae, arranged in different orientations in different regions of the asteroid body. Such an arrangement, typical of the liquid crystalline phases of the lipids in the water, motivated the researchers to call the asteroid bodies of liquid crystals of lipids of the vitreous humor. Through X-ray microanalysis, this study also confirmed the presence of calcium and phosphorus in the asteroid bodies, as well as suggested that the lipids present in the composition of these spheres would be the phospholipids.

In 2001, Winkler and Lunsdorf demonstrated, through electron spectroscopy images, a homogeneous distribution of calcium, phosphorus, and oxygen in the asteroid bodies, as well as a similarity of these particles with hydroxyapatite. These researchers also observed, through immunofluorescence, that chondroitin-6-sulfate was present on the surface of asteroid bodies and that part of the internal matrix of the asteroid bodies was composed of specific carbohydrates of hyaluronic acid. In 2003, Komatsu et al. confirmed the structural similarity between asteroid bodies and apatite (Kador and Wyman 2008; Komatsu et al. 2003; Winkler and Lunsdorf 2001).

Clinical Features

At the ophthalmological examination, the asteroid bodies can have white coloration when directly illuminated or golden color when they are under diffuse illumination (Galveia et al. 2013). These particles may be distributed randomly or organized along chains, surrounded by an adherent network of vitreous fibrils (Allison et al. 1991; Moss et al. 2001; Winkler and Lunsdorf 2001). Such deposits typically produce no symptoms or reduced visual acuity, but occasionally patients may report the presence of floaters (Yannuzzi et al. 2017).

The vitreous of the patient with asteroid hyalosis has particular characteristics, such as a lower vitreous liquefaction and a lower incidence of posterior vitreous detach-

ment, with the reported rate of PVD in 24–38% of these eyes. In order to investigate the anatomical characteristics of the vitreoretinal interface in these patients, Mochizuki and colleagues conducted an interventional case series in which patients with asteroid hyalosis and vitreoretinal diseases underwent vitrectomy assisted by triamcinolone. Through the preoperative optical coherence tomography (OCT), they observed that in some cases the vitreous cortex remained adhered to the surface of the retina even after visualization of the Weiss ring. Such a finding suggests that the prevalence of PVD in patients with asteroid hyalosis may be even lower than that reported in the literature, since most of the previous studies determined the occurrence of PVD through slit-lamp biomicroscopy (Mochizuki et al. 2008, 2009).

Another characteristic of the vitreous in patients with AH is the presence of anomalous vitreoretinal adhesions, which may be associated with residual vitreous cortex on the retinal surface, vitreoschisis, anomalous PVD, and diseases of the vitreoretinal interface, such as macular hole (MH) and epiretinal membrane (ERM). In the study by Mochizuki et al. (2009), preoperative OCT revealed abnormal vitreoretinal adhesions in eight of the ten eyes submitted to vitrectomy.

Despite the diagnosis of asteroid hyalosis is clinical, complementary exams can bring additional information and thus help to determine the best way to conduct the case.

Fluorescein Angiography

When asteroid hyalosis makes the fundoscopic visualization difficult and furthermore some retinal pathology is suspected, fluorescein angiography can reduce the distortion caused by hyalosis and facilitate the diagnosis, since the barrier filters of the machine help to screen out reflected asteroid blue light scatter (Hwang et al. 2006).

Optical Coherence Tomography

In the context of asteroid hyalosis, the OCT may help with the diagnosis of some macular changes. In a series of cases conducted by Hwang et al., OCT was performed in seven patients in whom asteroid hyalosis did not allow adequate fundoscopic visualization. In this study, the OCT permitted the definitive diagnosis of conditions that included ERM, MH, traction retinal detachment, and drusen. In cases where the AH is dense, there may be loss of signal and shading of certain regions of the retina, which can be minimized through maneuvers such as better patient positioning, focus adjustment, use of polarization function, and by improving corneal lubrication (Hwang et al. 2006).

Ocular Ultrasonography

In A-mode ultrasonography, asteroid hyalosis manifests as high reflectivity peaks. In B-mode, discreet moving points

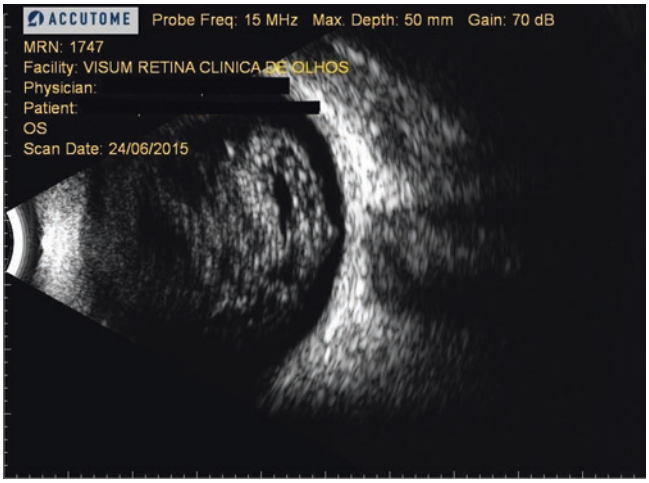

Fig. 14.4 B-scan ultrasonography shows intense reflectivity of the asteroid bodies without posterior acoustic shadow

are observed as bright echoes not associated with posterior acoustic shadow (Fig. 14.4; Kachewar and Kulkarni 2014).

Biometry and Automated Refraction

The importance of these two tests in patients with asteroid hyalosis lies in the reading error that can occur due to the media opacity present in these eyes. In the case of automated biometry, a false decrease of the axial length of up to 6.23 mm may occur. One of the explanations for this phenomenon is that the asteroid deposits in the vitreous would cause an interface with enough reflectivity to be interpreted as the anterior surface of the retina by the automated biometry unit (Allison et al. 1991). Modern autorefractors require a minimum degree of transparency of media for their realization. In patients with asteroid hyalosis, it is believed that there is a decrease in the instrument sensitivity and an abnormality in the retinoscopy reflex, which originates from the middle of the vitreous rather than in the retina, making the eye appear more hyperopic than it is (Wong and Sampath 2002).

Management

Treatment is rarely necessary, but occasionally vitrectomy may be required to remove visually significant opacities or to facilitate the treatment of underlying retinal abnormalities, such as proliferative diabetic retinopathy (Parnes et al. 1998).

When vitrectomy is indicated, two aspects should be considered. First, due to the presence of abnormal vitreoretinal adhesions, patients with AH have an increased risk of retinal tears during surgery. In addition, in some cases, there is an anomalous PVD associated with vitreoschisis or residual vitreous cortex adhered to the retina. In these patients, the use of triamcinolone to remove the entire vitreous can be especially beneficial (Yamaguchi et al. 2007).

Synchysis Scintillans

Introduction

The presence of cholesterol crystals in the vitreous cavity was first described by Parfait-Landrau in 1828 (Kumar 1963; Potter 1983). This condition, known as synchysis scintillans (SC) or cholesterolosis bulbi (CB), is characterized by the presence of highly refractive vitreous opacities, flattened, brown, or gold colored, dispersed in the vitreous cavity (Figs. 14.5 and 14.6; Banc and Stan 2015; Silva and Farah 2016).

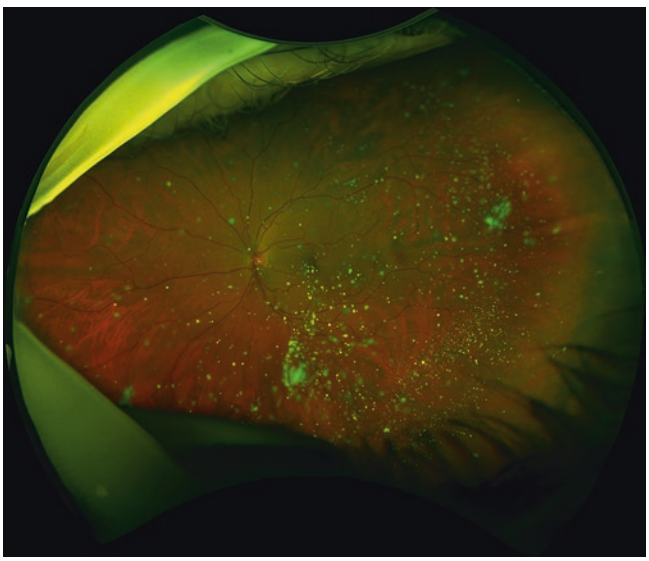

Fig. 14.5 Optomap imaging of a patient presenting with vitreous synchysis scintillans

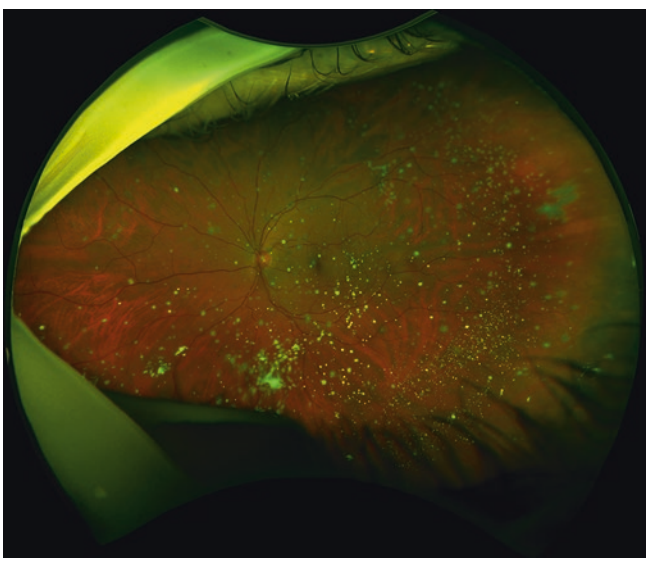

Fig. 14.6 Same condition of synchysis scintillans in another patient, captured by optomap imaging

Demographic Data

Although there are few studies about the epidemiology of SC, some authors claim that this condition occurs at an earlier age than asteroid hyalosis, typically affecting patients in the third decade of life (Potter 1983; Wand et al. 1975). In addition, unlike asteroid hyalosis, which can be unilateral in up to 93% of cases, synchysis scintillans is usually bilateral (Mitchell et al. 2003; Moss et al. 2001; Potter 1983; Wand et al. 1975; Winkler and Lunsdorf 2001).

Pathogenesis

Cholesterol deposits have been demonstrated in many ocular tissues, with the most common sites being the lens, the vitreous, and the retina. The occurrence of synchysis scintillans is associated with intraocular hemorrhages, traumas, degenerations, inflammations, particularly those of the uveal tract, as well as chronic retinal detachments and Coats' disease (Kumar 1963; Park et al. 2011).

Some studies have shown that, under normal conditions, the concentration of cholesterol in the vitreous is much lower than the blood one. According to these studies, the main source of intraocular cholesterol crystals would be degenerating extravascular blood. This mechanism would explain the cases of synchysis scintillans associated with intraocular hemorrhages. Another mechanism proposed to explain the presence of cholesterol crystals are the changes in the blood–aqueous barrier, present in cases of intraocular inflammation. Pathologies such as chronic retinal detachment or exudative retinal detachment caused by Coats disease may be associated with synchysis scintillans, since the subretinal fluid is rich in cholesterol, which can diffuse into the vitreous chamber through retinal breaks (Park et al. 2011).

Synchysis scintillans is usually observed in the vitreous cavity, but may also be present in the anterior chamber, which was first described by Schmidt (1831). Through techniques of wet field microscopy, electron microscopy, and histopathology, a study conducted by Kennedy in 1996 analyzed the pathogenic mechanism of three different cases of synchysis scintillans in the anterior chamber. In the first case, the cholesterol crystals were originated from a vitreous hemorrhage, while in the second case the origin of the cholesterol in the anterior chamber was a chronic retinal detachment. The aphakia, present in the two cases mentioned above, as well as the lens subluxation, can facilitate the migration of the cholesterol crystals to the anterior chamber. In the third case, the phacolysis and degradation of intralenticular cell membranes would be responsible for the appearance of cholesterol crystals in the anterior chamber of a patient with morgagnian nucleus. The cases of SC caused by phacolysis usually have a better prognosis when compared to cases of

chronically diseased eyes, such as cases of vitreous hemorrhage and retinal detachments (Kennedy 1996).

Composition

By chromatographic analysis of aqueous humor and vitreous humor, Andrews et al. (1973) observed that the crystals present in the eye of a patient with cholesterolosis bulbi were composed of pure cholesterol. In addition, after comparing the aqueous humor of a patient with synchysis with a normal aqueous humor, they proposed that the normal eye would have mechanisms to keep cholesterol in solution, which could be damaged in the eye with synchysis scintillans.

Clinical Features

Synchysis scintillans is characterized by the presence of crystals with flattened and shiny appearance, which freely disperses according to the ocular movements, depositing inferiorly when the movement is interrupted, causing a snow globe effect (Potter 1983; Silva and Farah 2016; Yannuzzi et al. 2017). One of the explanations for the inferior location of cholesterol crystals is a higher incidence of vitreous liquefaction in the eyes, unlike what occurs in patients with asteroid hyalosis, in whom the vitreous has a lower PVD rate and a higher incidence of anomalous vitreoretinal adhesions (Potter 1983; Wand et al. 1975).

Complementary Exams and Management

The diagnosis of synchysis scintillans is made through clinical examination, but in cases of intense vitreous opacity, complementary examinations such as optical coherence tomography, fluorescein angiography, and ocular ultrasound can give more details about retinal condition and help in the follow-up of underlying pathologies, such as diabetic retinopathy.

Patients with SC usually have little or no symptomatology, so surgical treatment is usually not necessary. Vitrectomy may be performed in cases of significant vitreous opacities, attempted visual improvement, or to facilitate the monitoring and treatment of other retinal diseases (Silva and Farah 2016).

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Myelinated Retinal Fiber Layer

15

Bruno de Queiroz Alves

Introduction

Myelinated retinal nerve fiber (MRNF) layers are a relatively common ophthalmoscopic finding and were first described as "chalk-white spots" by Virchow in 1856 (2). It appears in 0.57–1% of the population, typically present at birth as static lesions, and can occur bilaterally in 7.7% of affected patients (Gradle 1921; Virchow 1856).

MRNF commonly appear contiguous with the optic nerve as gray–white well-demarcated patches with feathered or fibrillated borders along the retinal nerve fiber layer, obscuring underlying retinal vessels. Although, MRNF may also have be seen discontinuous with the optic nerve head and along the vascular arcades (Gradle 1921; Virchow 1856).

Despite MRNF-affected patients are commonly asymptomatic, in some cases the vision can be compromised by axial myopia, amblyopia, and strabismus in the affected eye (Kodama et al. 1990; Straatsma et al. 1981). Myelination of the MRNF is often congenital but can be acquired or even progress in childhood or adolescence (Jean-Louis et al. 2000).

Etiopathogenesis

The myelin is produced by a complex process mediated by neuro-hormonal signals that stimulate the oligodendrocytes, and the axonal myelination commonly occurs in the direction of the impulse conduction down the axon. In MRNF, the myelination of retinal ganglion cell seems to take on the opposite way from the optic tract toward the eye (Magoon and Robb 1981). Despite the myelin formation process is well understated, the pathophysiology of MRNF remains largely unknown (FitzGibbon and Nestorovski 1997).

Clinical Features

The diagnosis is usually made accidentally by fundoscopy, in asymptomatic and healthy patients. It is observed on ophthalmoscopy often around the optic nerve disc some white striated patches with feathered borders involving from one-disc diameter in size to larger areas of the retina. The MRNF is more prevalent in the superior sector, although the myelinization may be disconnected from the optic disc head (Kodama et al. 1990; Figs. 15.1, 15.2, 15.3 and 15.4).

Diagnosis and Imaging

The myelin in MRNF consists in a high lipid content affecting imaging modalities. This sensitivity to lipids results in whitish appearance of MRNF on infrared and red-free image. Myelin blocks the detection of fluorescent material appearing dark on fundus autofluorescence and also is responsible for the blocking effect created on fluorescein angiography (Fig. 15.5a–d). On optical coherence tomography (OCT),

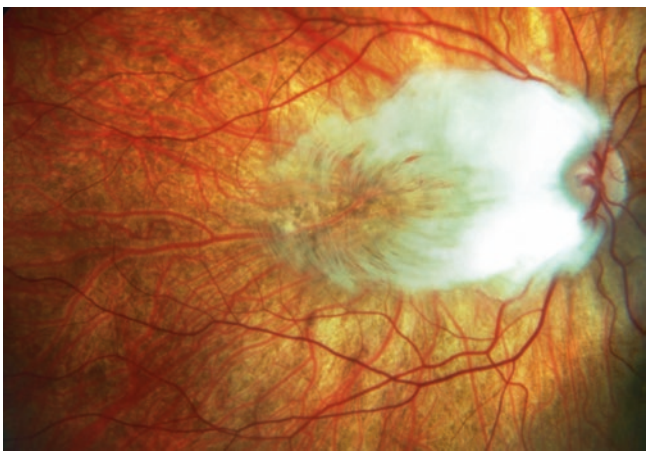

Fig. 15.1 Myelinated retinal nerve fiber layers from the optic disc until the foveal region. Color fundus photography

B. de Queiroz Alves (✉)
Department of Ophthalmology, São Paulo Federal University, São Paulo, Brazil

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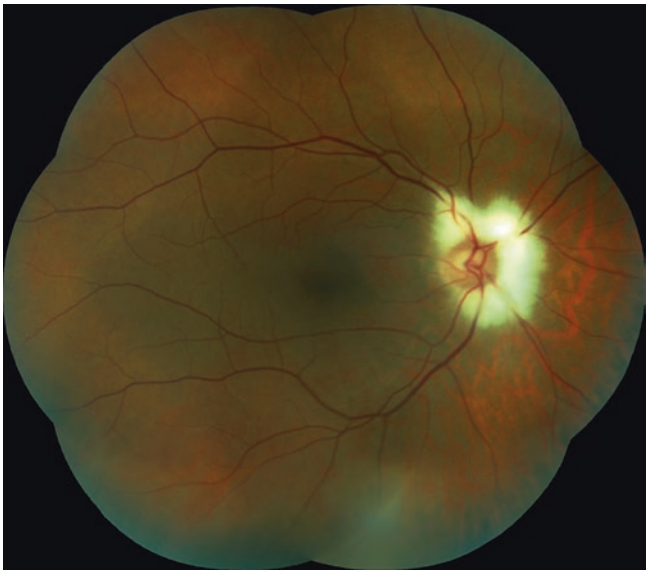

Fig. 15.2 Myelinated retinal nerve fiber layers around the optic disc. Color fundus photography

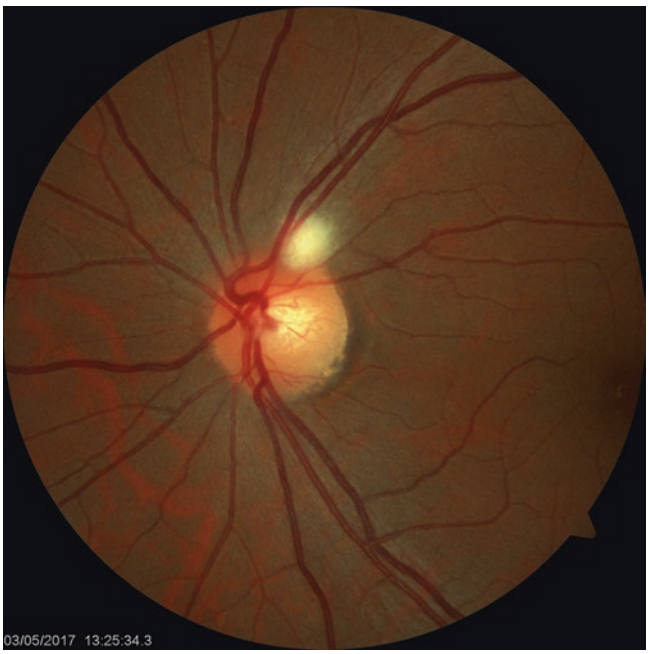

Fig. 15.3 Myelinated retinal nerve fiber layers superior to the optic disc. Color fundus photography

MRNF is thickened and hyper-reflectivity causing back-scattering and decreasing visibility of retinal layers below myelin (Fig. 15.6; Shelton et al. 2013).

Conditions Associated to MRNF

Myelinated retinal nerve fibers have been linked with numerous ocular abnormalities, including craniofacial abnormalities, neurofibromatosis, coloboma, polycoria, and keratoconus (Duke-Elder 1963), and strabismus was found in 66% (Tarabishy et al. 2007). An autosomal recessive syndrome of growth retardation, alopecia, pseudoanodontia, optic atrophy (GAPO syndrome) and MRNF associated with hypertelorism, severe glaucoma has been described in familial cases (Bozkurt et al. 2013). Other systemic syndromes have been described to be associated: Turner syndrome (Aaby and Kushner 1985) and Down syndrome (Straatsma et al. 1981).

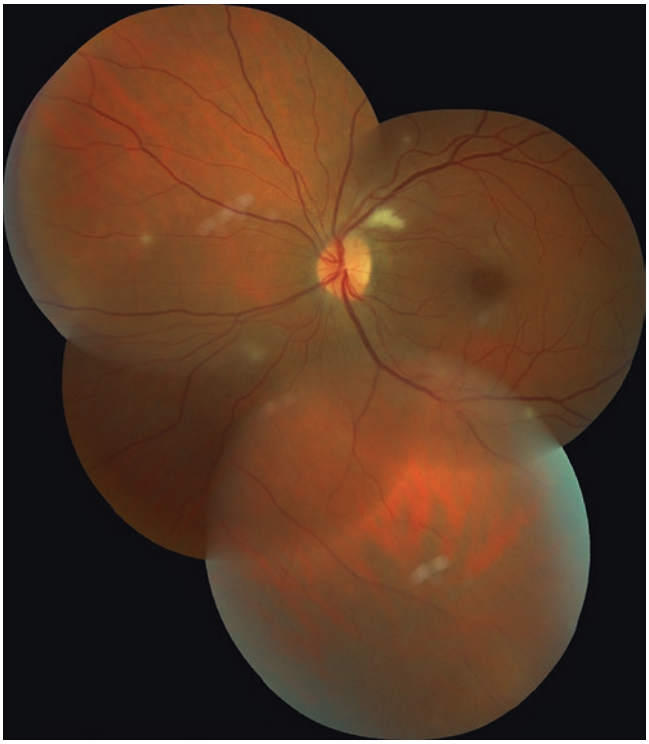

Fig. 15.4 Myelinated retinal nerve fiber layers in different points of the retina, including superior to the optic disc. Color fundus photography

Even though most MRNF cases are congenital, some conditions have been associated with acquired and progressive myelination: Goltz-Gorlin (multiple basal cell nevus) syndrome (De Jong et al. 1985), blunt trauma (Aaby and Kushner 1985), optic nerve sheath fenestration for chronic papilledema (Ali et al. 1994), optic nerve drusen (Parulekar and Elston 2002), family history of optic nerve hypoplasia (unaffected child) (Jean-Louis et al. 2000), Arnold-Chiari malformation associated with hydrocephalus (Baarsma 1980), and Von Recklinghausen's disease (Kushner 1979).

Management

MRNF are typically benign but to rule out potentially risky conditions, it is important to perform a complete blood count, ophthalmoscopy, and fluorescein angiography to differentiate from an embolic phenomenon. A visual field testing is mandatory in presence of visual defects. It is important to recognize the benign nature of MRNF to avoid additional diagnostic evaluations (Kodama et al. 1990; Shelton et al. 2013).

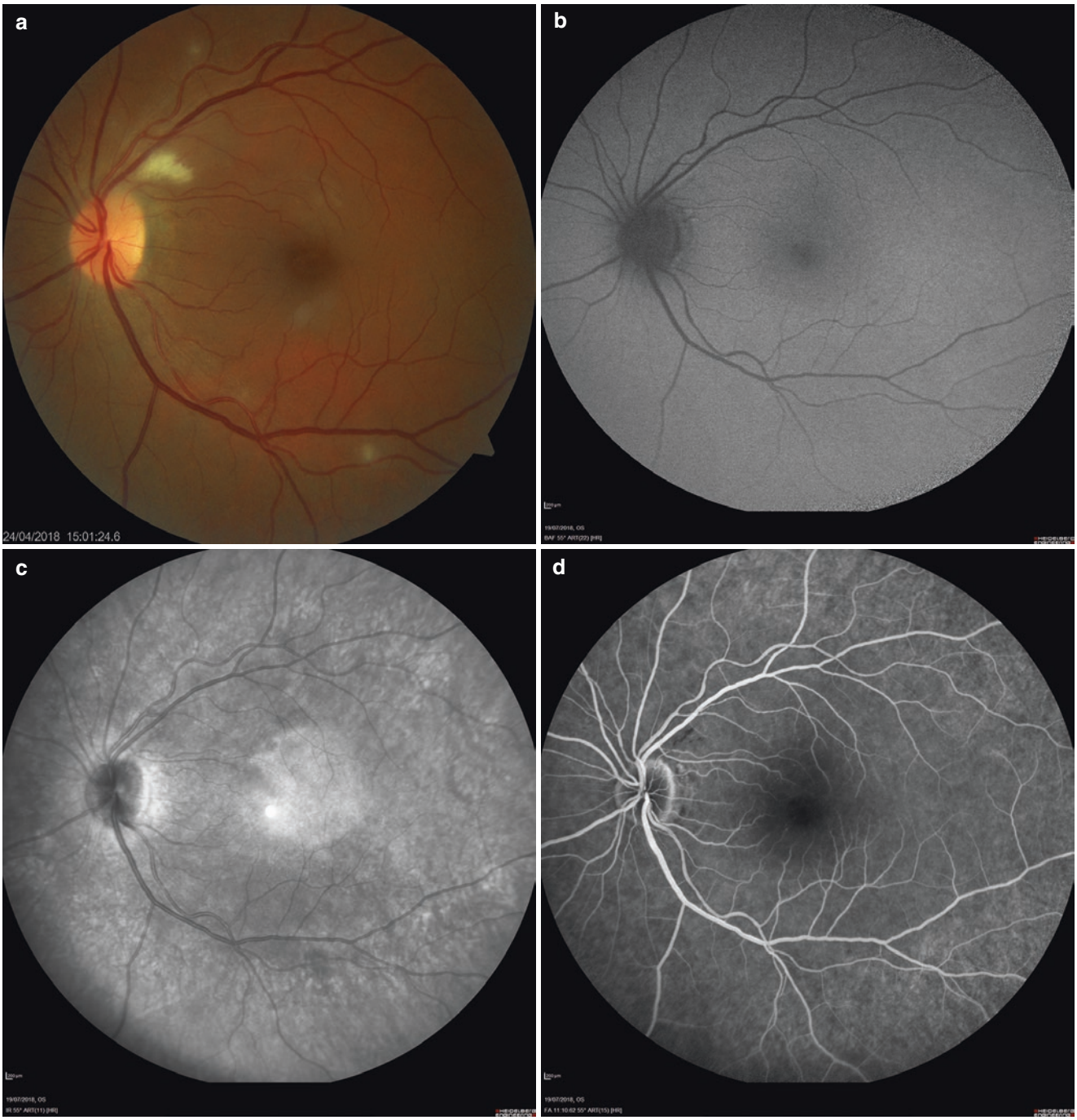

Fig. 15.5 (**a**) Myelinated retinal nerve fiber layers—color fundus photograph. (**b**) Same patient in fundus autofluorescence photogram exam. (**c**) Same patient in infrared fundus photograph exam. (**d**) Same patient in fluorescein angiography fundus photograph

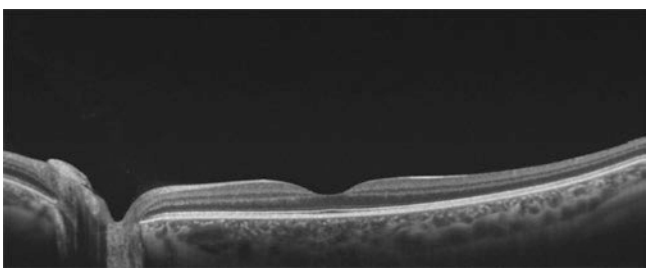

Fig. 15.6 Myelinated retinal nerve fiber layers. Optical coherence tomography shows thickened and hyper-reflective MRNF

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Congenital Optic Disc PIT

16

Mayara Fernanda Pacovska and Eduardo Tomazoni

Introduction

The first description of congenital optic disc PIT (ODP) was in 1882 by Wiethe who notes a dark depression in both optic discs of a 62-year-old woman (Wiethe 1882). There is no difference between gender, 85% of the patients have unilateral commitment, and the estimated incidence is 1 in 11,000 eyes (Brown et al. 1980; Kelkar et al. 2012). The main complication is maculopathy which occurs in 25–75% of patients presenting with poor visual acuity and macular detachment detected with fundus exam and OCT (Georgalas et al. 2010). In those cases, the treatment is with pars plana vitrectomy associated with laser photocoagulation in the margin of OPD, which generally is the first approach (Karanjia et al. 2018).

Pathophysiology

The development of congenital ODP is mainly sporadic, with some few cases reported autosomal dominant fashion (Stefko et al. 1997). In cases of maculopathy associated with ODP, the origin of the fluid that is detaching the retina is unknown. The likely sources are blood vessels in the area of liquid fluid (Gordon and Chatfield 1969), cerebrospinal fluid (Irvine et al. 1986), choriocapillaris vessels through Bruch's membrane below the macula (Wise et al. 1971), and liquefied vitreous (Brockhurst 1975; Sugar 1962). The most acceptable theory is the vitreous as source of the subretinal fluid, with access to subretinal space by the OPD defect (Karanjia et al. 2018; Lincoff and Kreissig 1998).

Clinical Findings

More than 90% of the patients with ODP present with a depression with ovoid aspect and gray-white appearance (Fig. 16.1), in others may be yellowish or black. Localized temporal to the disc in 72% with size varying from 0.1 to 0.7 disc diameters (Brown et al. 1980; David and Reed 1999). Visual acuity generally is normal in cases without complications (Brodsky 1994). If the ODP is in the periphery of the disc, peripapillary atrophy and other pigmentary changes in EPR are expected (Brown et al. 1980; Karanjia et al. 2018). The ODP aspects do not change during life, although retinal findings may be progressive, and 25–75% of patients develop secondary maculopathy commonly during the third decade (Georgalas et al. 2010; Karanjia et al. 2018). In these cases, macular findings are macular detachment, retinoschi-

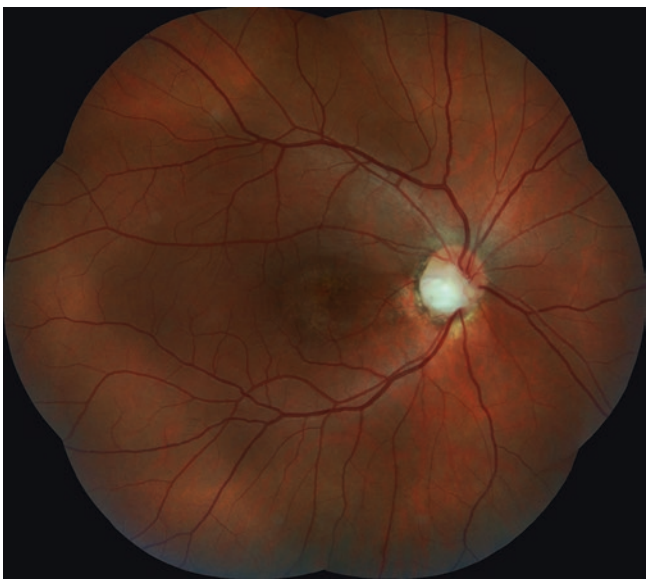

Fig. 16.1 Fundus image of patient with optic disc pit in temporal side, with an ovoid white appearance and border of the PIT surrounded by atrophy and pigmentary changes. Special thanks to Eduardo Büchele Rodrigues, from Visum Retina

M. F. Pacovska
Department of Ophthalmology, Governador Celso Ramos Hospital, Florianópolis, SC, Brazil

E. Tomazoni (✉)
Ophthalmology, Retina Fellowship, Visum Retina, Florianópolis, Santa Catarina, Brazil

E. B. Rodrigues et al. (eds.), *Trauma and Miscellaneous Disorders in Retina*, Retina Atlas, https://doi.org/10.1007/978-981-13-8550-6_16

sis, cystic degeneration, and degenerative pigment changes (Georgalas et al. 2010). These patients present with decreased visual acuity to 0.1 or worse (Bonnet 1991; Sobol et al. 1990).

Ocular Trauma-Associated Maculopathy

The relation between ocular trauma and development of serous retinal detachment was related by some authors. They hypothesized that the cause of macular detachment is the anomalous vitreous radial traction to the macular region, mainly caused by persistence of Cloquet's canal or due to primary vitreous attachment (Fig. 16.2a–c; Billi et al. 1996; Colyer et al. 2007; Meyer and Rodrigues 2004).

Visual Field

The arcuate scotomas found in this exam probably reflect the absence of nerve fiber layer caused by ODP (Karanjia et al. 2018), although defects can appear randomly without correlation between the PIT location and the corresponding defect (Brown et al. 1980). Central scotomas are found in complicated cases with central serous retinopathy (Bonnet 1991; Karanjia et al. 2018; Kranenburg 1960).

Optical Coherence Tomography (OCT)

After fundus exam, OCT exam is the best way to evaluate ODP complications. In non-complicated cases, a reduction in the thickness of the nerve fiber layer corresponding to the

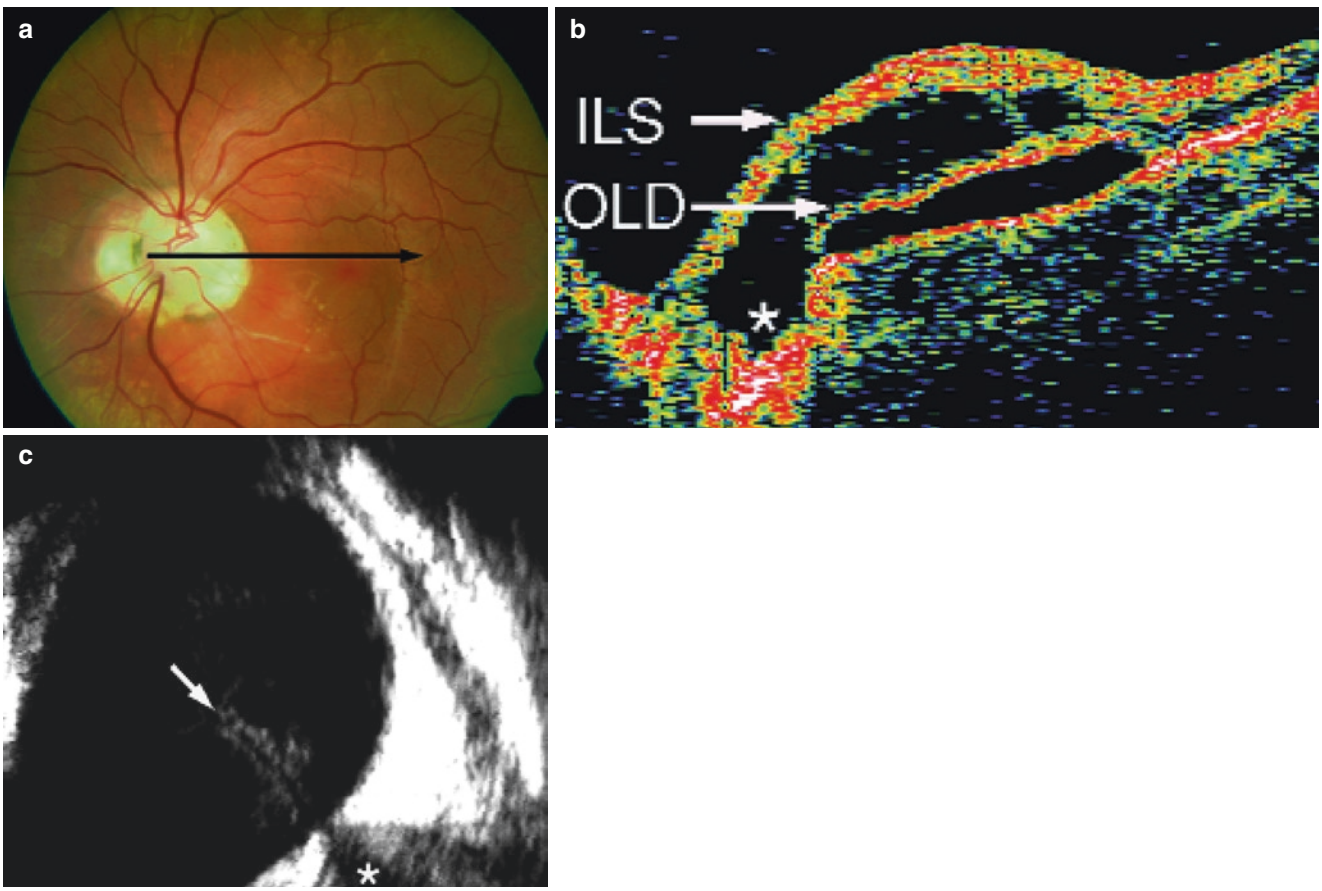

Fig. 16.2 (**a**) Fundus image of a 16-year-old victim of blunt ocular trauma for the previous 4 weeks. (a) ODP and serous retinal detachment from the optic nerve to the macula; the ODP is inferotemporal and is surrounded by hyperpigmented margin. (**b**) Horizontal OCT of the same patient presenting with a thick hyperreflective orange to reddish color (nerve fiber detachment) from the mid of optic nerve across papillomacular region to the margin of the fovea; ODP is localized in the temporal edge of the optic nerve as a deeper hyperreflective reddish lesion; the second hyperreflective band is the outer layer detached from the retinal pigment epithelium; large schisis-like cavity is seen as non-reflective space between the inner layer and outer layer segment; the retinal pigment epithelium and choriocapillaris complex presents as another hyperreflective band; the fluid under the outer layer detachment can leak through a hole in the outer layer into the subretinal space. (**c**) B-scan ultrasound with optic nerve presenting with retrobulbare hyporeflective band (white star), and intravitreal tubular hyperreflective structure compatible with the Cloquet's canal extending perpendicular from the rim of the optic nerve into the vitreous cavity (white arrow). Special thanks to Carsten Helmut Meyer

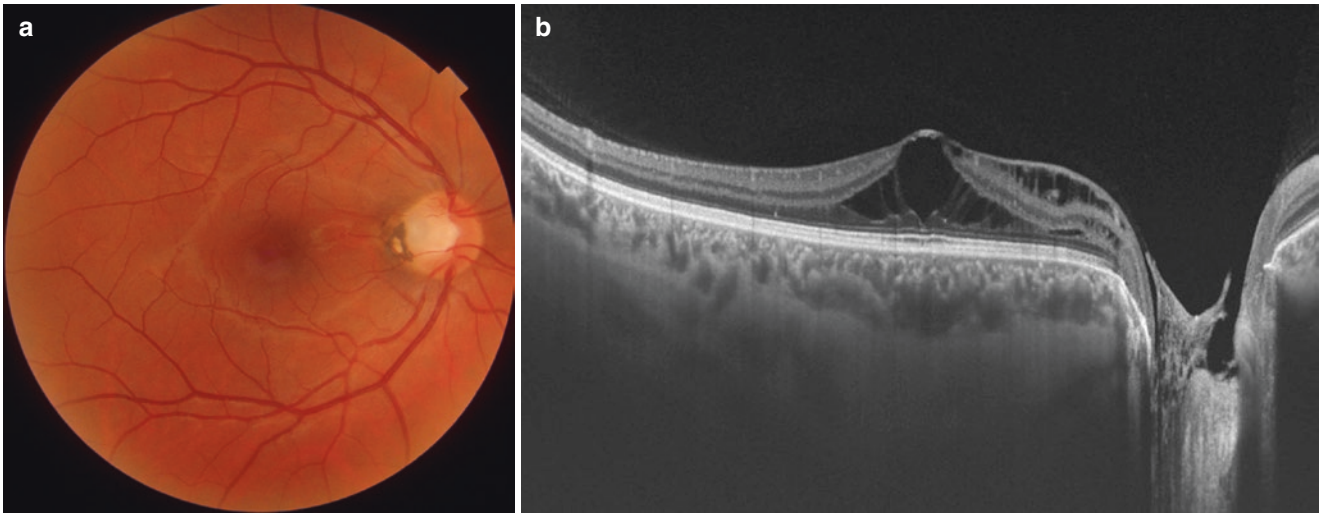

Fig. 16.3 (**a**) Fundus image of congenital optic disc pit with associated maculopathy. (**b**) OCT exam of the same patient with schisis and stria between inner and outer layers, retinal edema with fluid in the intraretinal layer, fluid in the nasal part of the optic nerve head and with membrane spanning over the optic disc pit. Special thanks to Gabriel Andrade, from BRAVS

optic disc PIT location can be found. Maculopathy associated with ODP is the main complication, and the findings include serous retinal detachment, retinoschisis and stria between inner and outer layers, retinal edema with fluid in the intraretinal and subretinal layers, fluid below the optic nerve head, and membrane spanning over the optic disc pit (Fig. 16.3a, b; Georgalas et al. 2010; Karanjia et al. 2018; Michalewski et al. 2014).

Treatment

Patients with congenital OPD must be accompanied due to the high risk of complication, especially secondary maculopathy. In these cases, the treatment remains a challenge. Isolated use of laser proves no benefit (Brockhurst 1975). Few surgeons test scleral buckling, also without success. The use of pars plana vitrectomy (PPV) associated with laser therapy is the treatment of choice by many surgeons with first therapy (Jain 2014). Other techniques such as use of isolated micropulse laser and platelet-rich plasma were also used (Valdés-Lara et al. 2018).

PPV and Laser Therapy

It is the main treatment for most of the surgeons in the first approach (Jain 2014). Laser photocoagulation in the margin of ODP after PPV acts by blocking the fluid to subretinal space and has been proved to be beneficial by avoiding the persistency or formation of maculopathy, even after 10 years of follow-up (Jain 2014).

Platelet-Rich Plasma Application with Gas Tamponade

Autologous serum with platelet-rich plasma (PRP) over the ODP in combination with PPV, internal limiting membrane (ILM) peeling, and gas tamponade was suggested by Moreira Neto and Moreira Junior (2013). They evaluated five patients submitted to these procedures without recurrence by at least 7 years of follow-up. They believe at the time of the surgery that PRP could act in benefit to reapply the macular region. After analyzing other studies (Banker et al. 1999; Ezra et al. 2004), the authors believe that the main cause of success in these patients was the other procedures (PPV, ILM peeling, and gas tamponade), and not by using the PRP.

Isolated Subthreshold Micropulse Macular Laser

Valdés-Lara et al. report a case of a patient with persist macular detachment after being submitted to PPV with gas tamponade and no laser treatment. After 4 months of follow-up of the patient, there was an improvement in the vision and reduction of central macular thickness (Valdés-Lara et al. 2018). The authors of this chapter did not find any other related publication of using micropulse laser in the treatment of secondary maculopathy in ODP.

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Melanocytoma of Optic Nerve

17

Paula M. Marinho, Eduardo Tomazoni,
and Eduardo Büchele Rodrigues

Introduction

Melanocytoma of optic nerve (MON) is a rare primarily benign lesion, but due to its malignant potential—estimated around 1–2% of cases (Shields et al. 2006), possible complications and the clinical similarities with other pigmented optic nerve lesions, such as malignant melanomas, one should be able to know and recognize clinical features, hence providing adequate follow-up and assistance.

Initial diagnostic usually happens on routine evaluations during ophthalmoscopic examination in asymptomatic patients (Zografos et al. 2004), where it is commonly seen as a dark pigmented—brown to black—lesion at the optic nerve (ON) with soft margins (Figs. 17.1 and 17.2). In some cases, the lesion can be elevated and not restricted to ON, involving the adjacent retina.

Additional exams such as retinography, autofluorescence, visual field, ultrasound, and optical coherence tomography should be performed when lesion is first found, providing adequate information for further analysis.

Histopathology

Melanocytomas are benign tumors that develop anywhere where melanocytes are present. Regarding MON, we can find those types of cell on the lamina cribrosa, and they are

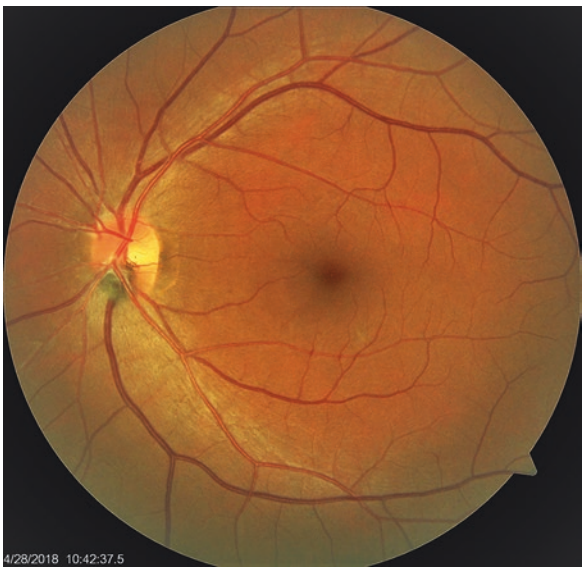

Fig. 17.1 Color fundus photograph shows pigmented lesion in inferior border of left optic nerve

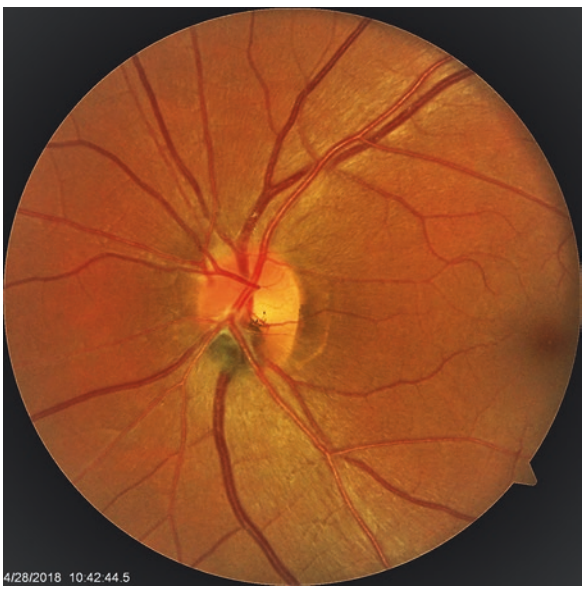

Fig. 17.2 Same patient with zoom in optic disc

P. M. Marinho
Department of Ophthalmology, São Paulo Federal University,
São Paulo, Brazil

E. Tomazoni (✉)
Ophthalmology, Retina Fellowship, Visum Retina,
Florianópolis, Santa Catarina, Brazil

E. B. Rodrigues
Department of Ophthalmology and Visual Sciences,
Federal University of São Paulo, São Paulo, Brazil

Department of Ophthalmology, Saint Louis University
School of Medicine, Saint Louis, MO, USA

E. B. Rodrigues et al. (eds.), *Trauma and Miscellaneous Disorders in Retina*,
Retina Atlas, https://doi.org/10.1007/978-981-13-8550-6_17

classified as a variety of magnocellular nevus (De Potter et al. 1996) composed histopathologically of intensely pigmented round to oval cells with copious quantities of cytoplasm and small, round, uniform, bland nuclei (Shields et al. 2006), better analyzed under bleached preparations.

Clinical Features

Studies and case reports have shown a discrete prevalence for female patients (2.1), with mean age of 46–50 years old. Most cases are asymptomatic with visual acuity of 0.8 or better, and lesions were found during routine ophthalmological evaluation.

MON usually presents itself as unilateral pigmented lesion with 1–2 mm width, but it can also outgrow the ON. When not restricted to the ON, it overtakes adjacent choroid and/or retina, the last one presenting a feathering appearance and an increased correlation with neovascular processes.

Some of the major studies in MON (Joffe et al. 1979; Shields et al. 2004) have shown that up to 15% of the tumors may grow, but it has no correlation with malignant changes.

Fundus Image and Fundus Autofluorescence

The aim in retinography is to document the findings and provide an easier and accurate way of reaccessing it in order to compare possible growth, whereas in fundus autofluorescence, we find a total absence of autofluorescence (Fig. 17.3).

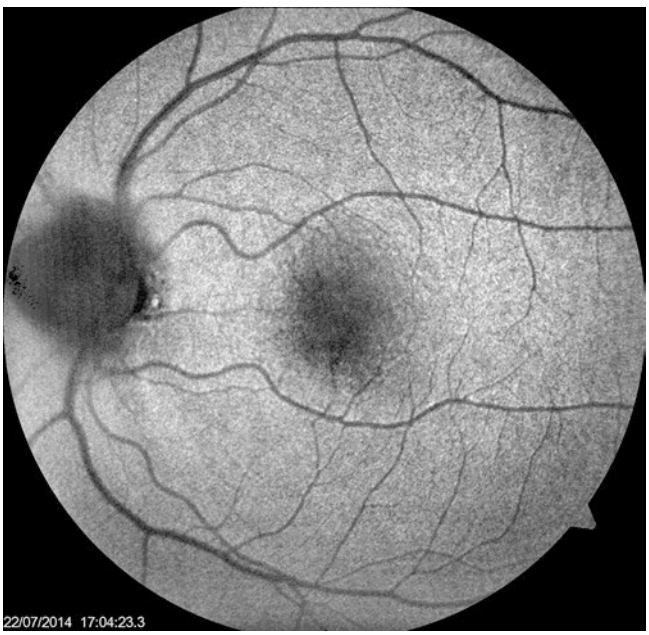

Fig. 17.3 Fundus autofluorescence exam shows absence of fluorescence in nasal border of left optic nerve

Fluorescein Angiography

MON presents a marked hypofluorescence in all stages of angiogram. Hyperfluorescent areas may be present due to secondary alterations, like edema of the optic nerve or adjacent subretinal fluid (Fig. 17.4).

Optical Coherence Tomography (OCT) and Optical Coherence Tomography Angiography (OCT-A)

The presence of a dome-shaped tumor configuration with severe posterior shadowing is the most usual finding in OCT, especially regarding spectral domain OCT. Swept source OCT has been used in order to obtain more detailed images, and initial data have described findings as a thick irregular hyperreflective layer continuous with the retinal outer layers composed of aggregated nodular dots and scattered hyperreflective dotting (Fig. 17.5). It has been considered that the hyperreflective dotting may be correlated with melanophages.

Few data are available regarding OCT-A, but its capacity to evaluate vascular structures may provide a new pathway regarding intratumoral alterations.

OCT should be considered as an important tool to access and monitor subtle changes with no or small clinical consequences, like nerve compression, retinal thickness, and choroidal changes, especially neovascularization.

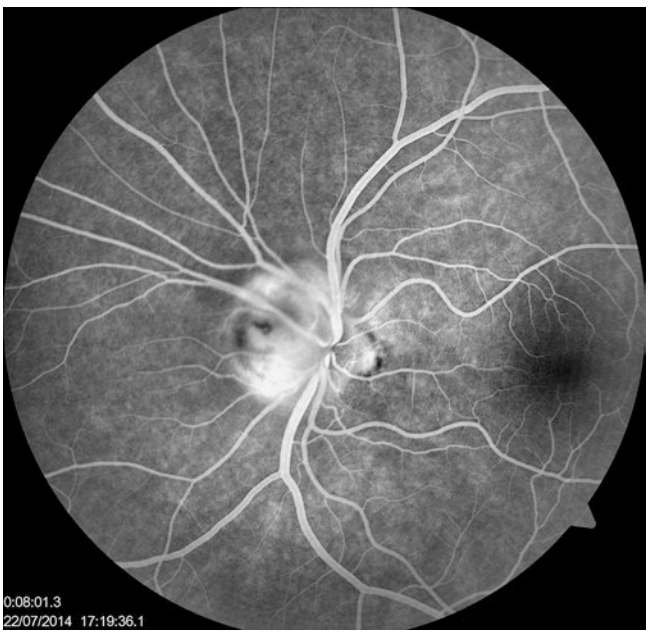

Fig. 17.4 Fundus fluorescein angiography shows left optic nerve with central areas of hypofluorescence in nasal border associated with secondary edema and fluid—hyperfluorescent areas in the periphery of the disc

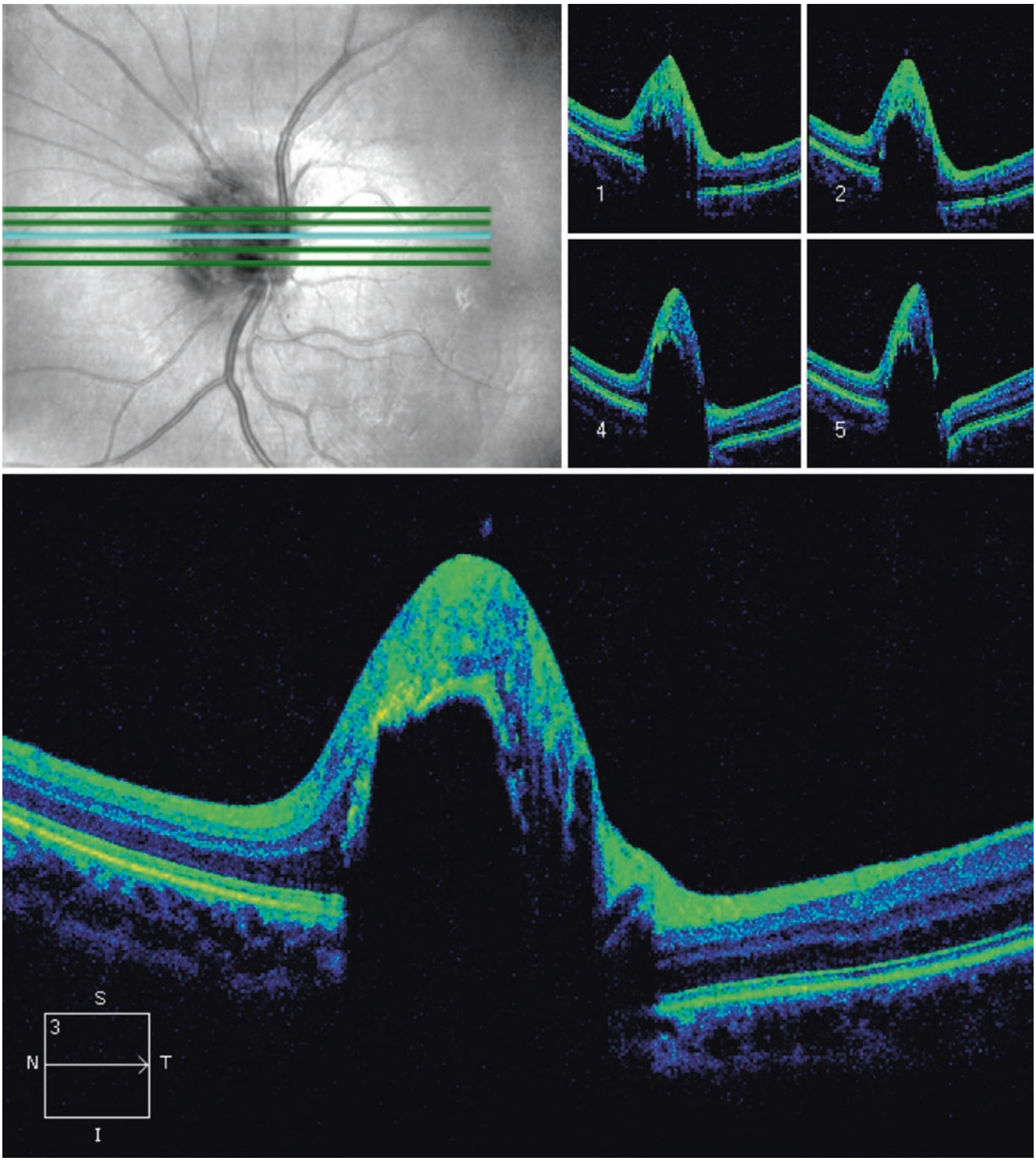

Fig. 17.5 OCT shows dome-shaped configuration with areas of hyporeflectivity and hyperreflectivity, with marked posterior shadowing

Ultrasonography

Although controversial, some authors do emphasize the use of ultrasonography to differentiate melanocytomas from melanomas, mainly when their elevation is over 0.5 mm. In MON. it is possible to see high internal reflectivity on A-scan and avascular behavior, opposing to the classical presentations of melanomas (Gologorsky et al. 2010; Lisker-Cervantes et al. 2017). In borderline presentations, we can use ultrasonography in a complementary manner.

Visual Field

Visual field alterations may occur due to lesion size or later complications, such as focal necrosis. When present, the most commonly diagnosed alteration is enlargement of the blind spot, but it may have polymorphic alterations such as quadrantopsy, fascicular defects, or tubular field.

Differential Diagnosis

Other pigmented lesions should be considered as a differential diagnosis when approaching MON, mainly choroidal melanoma, choroidal nevus, hyperplasia of the retinal pigmented epithelium, and metastatic melanoma.

Management

Since it is considered one of the most common pigmented lesions of the ON, patients with MON can be clinically observed yearly with adequate imaging assessment. As previously reported, small increase in size has no strict correlation with malignancy, but in case of significant growth or visual alterations ocular enucleation must be considered. Some authors (Shields et al. 2018) suggest the confirmation with fine needle biopsy prior to enucleation in these cases.

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