4 Clinical and Opthalmological Evaluation

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4.1 Introduction

The clinical evaluation of patients with suspected or diagnosed orbital or cranio-orbital mass lesions is based on several important steps; these include the careful anamnesis and clinical history, the inspection of the eye and adnexa to evidence proptosis, eyelid and eye surface abnormalities, the functional evaluation of the visual acuity, visual field, ocular motility, and eye fundus. The optical coherence tomography (OCT) and ophthalmic ultrasonography are important complementary studies to the diagnosis. All these diagnostic steps are discussed in this chapter.

4.2 History

4.2.1 Family and Patient History

The remote anamnesis should investigate possible congenital diseases potentially associated with orbital mass lesions, such as neurofibromatosis [1, 2], Von Hippel-Lindau disease [3], familial cavernous malformations [4, 5]. Previous surgical procedures for oncological diseases, such as breast or prostatic cancer, must suggest the possibility of an orbital metastasis [6, 7]. Previous nasal and maxillary surgery may presage a mucocele [8] or a carcinoma with secondary invasion of the orbit. Several patients with ophthalmological problems underwent a neurosurgical procedure for skull base tumors, mainly of the suprasellar and parasellar regions; in such cases, a late orbital involvement due to tumor progression or recurrence must be suspected [9, 10]. In patients with previous diagnosis of orbital pathology and previous orbital surgery, clinical presentation, surgical description and histological diagnosis must carefully be reviewed.

4.2.2 First Complaints and Symptoms

Most patients with cranio-orbital mass lesions seek ocular consultation for proptosis, eventually associated with pressure sensation and orbital pain. Decrease of vision and diplopia may occur later [9, 10].

The pressure sensation is probably due to increasing orbital pressure secondary to slow expanding tumors or cysts. It has also been observed in patients with decompensation of the

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orbital apparatus after a long-standing inflammatory tumor [11].

Pain is a frequent symptom of orbital lesions. It may be continuous or episodic, sometimes nocturnal, and is often severe. The referred site of pain may be related to the location of the mass lesion. Pain located to the deep orbit may be referred to an orbital apex lesion, mainly if associated with ocular motility disturbance. If the pain is more intense in the infero-medial orbital compartment, it should suggest a malignant tumor of the ethmoid or maxillary sinus. If pain is referred to the lateral orbital compartment, a malignant neoplasm or an inflammatory process of the lacrimal gland may be present.

Headache is a less frequent symptom which may accompany inflammatory and vasculitic diseases, cranio-orbital meningiomas and secondary orbital invasion by malignant tumors. The head noise may be referred in patients with orbital vascular malformations or less frequently with intracranial vascular lesions contiguous to the orbit. It is more frequently of pulsating type and synchronous with the heartbeat.

4.2.3 Chronology of the Evolution Before Diagnosis

The length of the clinical history, the type of onset, and the rate of progression of the symptoms are very important for the diagnosis, although they are often difficult to define.

The period of onset is mostly referred for more relevant symptoms, such as visual impairment and diplopia; on the other hand, dating the onset of the proptosis is more difficult. In many cases, prior photographs may show that slight proptosis was already present many years before [11]. Lesions with a long interval time from the presentation are usually benign.

The onset of symptoms may be slow or acute. A slow clinical onset should suggest a benign mass lesion; an acute onset of symptoms is more typical for malignant and inflammatory mass lesions [12, 13]. However, this is not always true. A hemorrhage within an asymptomatic benign cranio-orbital cyst may cause acute symptoms; otherwise, a malignant mass lesion may initially present with slow symptoms. Thus, the rate of symptom progression is more relevant.

An indolent course with isolated proptosis for several years suggests a cyst or a benign tumor [14]. In this context, sudden increase of the proptosis should suggest malignant degeneration. The rapid evolution of symptoms after the onset may be referred to as malignant tumor, but it is also more typical of an inflammatory lesion.

4.3 Inspection of the Eye and Adnexa

Inspection of the eye and adnexal structures should include the assessment of the proptosis, the state of the eyelids, and the surface of the eye.

4.3.1 Assessment of Proptosis

Proptosis, or forward protrusion of the eye globe, is an almost constant sign of orbital mass lesions [11, 15, 16]. It may be evidenced with a front view (Fig. 4.1), but the patient observation from above the eyebrow allows to better define the different globe position between the affected and the normal side.

The degree of globe displacement as compared to the contralateral is quantified in millimeters with the Hertel exophthalmometer, eventually with Krahn modification. The normal values derived with the Krahn device ranges from 14 to 21 mm in adults. Values <14 mm are considered

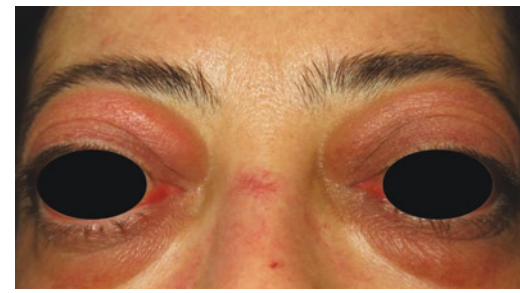

Fig. 4.1 Bilateral proptosis. (Image courtesy of Prof. Diego Strianese, Oculoplastic Unit University of Naples Federico II)

diagnostic for enophthalmos. Values >21 mm, although sometimes found in individuals as an ereditary tendency, may be diagnostic of proptosis, mainly when the difference between the two eyes is >2 mm.

Proptosis is associated with displacement of the eye in about of 80% of orbital mass lesions [11]. The type of displacement may be correlated with the tumor location. Superolateral and lacrimal gland mass lesions cause infero-medial displacement; medial and inferior lesions mainly cause eye displacement superiorly and laterally. Apical or intracranial lesions present with axial proptosis. The presence of eye displacement without proptosis is a rare event which may be observed for tumors arising from the eyelid and invading the orbit.

Although proptosis from orbital mass lesions is almost always unilateral, bilateral proptosis may be observed for several malignant lymphomas, metastatic carcinomas, or dural arteriovenous malformations; more often an unilateral orbital tumor may extend to the contralateral eye during its course.

Palpation of the proptotic eye may elicite tenderness, as for intraorbital mucoceles and cystic lesions. Pulsation synchrone with the arterial pulse is typical of the carotid-cavernous fistula. However, the pulsation may rarely be observed for some high vascular tumors and cavernous malformations.

4.3.2 Eyelids

Several eyelid abnormalities may be observed in patients with cranio-orbital mass lesions, including retraction, edema, rubor, ecchymosis.

The most accurate means to detect eyelid retraction is measurement of the distance between the margin of the upper eyelid and the superior orbital fold. When eyelid retraction occurs, the superior palpebral fold deepens, and the interval between palpebral fold and lid margin is shortened. Thus, the fold-lid margin ratio is a more reliable marker of eyelid retraction.

Orbit neoplasms with rapid growth may cause severe proptosis so that the eye is pushed out of a protective cover of the eyelid, thus mimicking an eyelid retraction. The true eyelid retraction with increased fold-lid margin ratio may occur for some slowly growing neoplasms.

Edema of the eyelid without erythema or other inflammatory signs may be associated with vascular and neurogenic tumors, probably secondary to long-standing venous stasis. Edema with inflammatory signs, such as hyperemia and/or tenderness is commonly associated with thyroid eye disease and orbital inflammatory syndromes, both infectious and non-infectious. Mild edema of the superior eyelid may occur for tumors located in the superior areas of the orbit. Edema of the nasal portion of the upper eyelid suggests a mucocele of the frontal sinus. Some degree of eyelid edema is also associated with orbital carcinomas. Finally, bilateral edema of all four eyelids is a frequent finding of orbital lymphomas.

Eyelid hyperemia and rubor may accompany fast-growing neoplasms and acute inflammatory mass lesions located in the anterior orbital areas, such as rhabdomyosarcoma and leukemia [12].

Ecchymosis of the eyelid may be associated with hematoma, metastatic neuroblastoma, but also to amyloidosis, plasmacytoma, and leukemia infiltrates.

4.3.3 Surface of the Eye

Several changes of the eye surface may be observed in patients with orbital mass lesions. Dilated, tortuous, reddish-purple vessels are diagnostic of vascular malformations or shunts. Episodes of subconjunctival hemorrhage may be a presenting sign of orbital mass lesions, mainly cavernous hemangiomas and lymphangiomas.

The epibulbar discoloration, defined salmon-hued lesion, is an objective sign typical of orbital lymphoma [13]. The painless and slowly growing mass of salmon color appears along the surface of the eye in a superior location. It results from forward extension of the orbital mass.

Epibulbar melanotic spots are signs of an intraocular melanoma or primary melanoma of the conjunctive.

4.3.4 External Mass

An external mass in the anterior orbit can sometimes be present. The mass must be evaluated for mobility, consistency (solid versus cystic), relationships with the orbital bones and underlying soft tissues. Mass lesions which may more frequently present with an external palpable mass include lacrimal gland tumors, dermoid cyst, and mucocele.

4.4 Functional Evaluation of the Eye

Determining the ocular function is a fundamental step in the diagnosis of patients with orbital mass lesions. In fact, the preservation of the visual function is the most important aim of surgery of orbital tumors.

The ocular function may be affected by an orbital mass in different ways: dislocation of the eye and optic nerve, anatomic contact between tumor and eye, infiltration of the nerves and muscles, damage to the blood supply to the eye.

4.4.1 Visual Acuity

The visual acuity should be measured at distance in Slenner or LogMAR notations. Near vision may also be measured but it does not replace the determination of distance acuity.

In many cases of orbital mass lesions, there is no correlation between visual acuity and entity of proptosis. Even gliomas arising from the optic nerve may remain asymptomatic or may present with proptosis and normal vision for a long time [17, 18].

4.4.2 Visual Field

The perimetry, or visual field analysis, is realized by quantitative static methods, such as Humphrey's 24-2 or 30-2, but may also be performed by other means, such as Goldman perimetry and Octopus perimetry.

The role of the perimetry is to record the degree of pretreatment visual loss, with the aim to decide the surgical resection. Besides, the preoperative visual field is important to define the functional outcome of the treatment [11].

4.4.3 Ocular Motility

The impairment of the ocular motility is an important clinical finding of patients with craniorbital mass lesions. It occurs with different mechanisms and often depends on the intraorbital location and size of the lesion itself. A mass lesion confined to one sector or quadrant of the orbit may cause diplopia only when the ocular rotation occurs into the area occupied by the mass. However, very large tumors may be associated with diplopia in all directions.

Apical mass lesions cause diplopia early in the eye movement, because of the early impairment of the oculomotor nerves at the superior orbital fissure.

Apical and medial orbital masses may sometimes cause transient diplopia and obfuscation of the vision during the forced eye excursion in the direction of the impaired extraocular muscles; this is due to transient compression to the vascular supply of the orbital portion of the optic nerve [11].

Besides the compression mechanism, diplopia from orbital mass lesions may be secondary to edema, inflammation, or infiltrations of the affected muscle. The muscle infiltration mainly occurs in orbital metastatic carcinomas and often evolves in muscular contracture and eye deviation.

Diplopia is usually the first symptom of schwannomas of the oculomotor nerves, in association with functional impairment of the involved nerve [19, 20]. However, nerve-related symptoms and signs may be absent [17].

4.4.4 Ophthalmoscopy

The ophthalmoscopic study is not highly relevant in the diagnosis of orbital mass lesions.

Indentations of the eye wall and choroidal folds are mainly observed in tumors of the anterior portion of the orbit.

Pallor of the optic disc and less frequently hyperemia, edema, or optic disc elevation are associated with optic nerve tumors (meningioma or glioma) or to retinoblastoma [11].

4.4.5 Intraocular Pressure

An increase in the intraocular pressure is associated with orbital vascular pathologies, including carotid-cavernous fistulas, arteriovenous malformations of the posterior orbit, dural vascular shunts. All these conditions cause rise of the orbital venous pressure. The increase of the intraocular pressure is resistant to medical therapy, but it is associated with scarce or no visual impairment.

4.4.6 Auscultation

In patient with suspected orbital mass lesions, the discovery of the bruit is suggestive of a vascular malformation of the orbit or contiguous intracranial compartment. This finding is evident in arteriovenous fistulas but not in the venous malformations, and thus it results from the arterial flow. Although the bruit is usually evidenced in the affected orbit, it may also be present at the frontal and temporal region.

4.4.7 Optical Coherence Tomography (OCT)

OCT is a helpful ancillary test [21–23]. It is typically performed for quantitative measurement of retinal nerve fiber layer (RNFL) thickness, retinal ganglion cell (RGC) layer thickness, and qualitative and quantitative assessment of macular health. In the presence of optic nerve damage, the RNFL and RGC layers are thinned; the quantitative measurement is a reliable indicator of stability or progression of damage over time. In the presence of orbital diseases, the RGC layer thickness is a more reliable indicator than RNFL thickness because the RNFL thickness may be increased by venous congestion (Fig. 4.2). Thus, increasing RNFL thickness over time may be considered an objective measurement of increasing venous congestion and/or a sign of optic nerve head inflammation, infiltration, or ischemia. The OCT is more sensitive in the assessment of change over time than ophthalmoscopy, serial fundus photography, or magnetic resonance (MR).

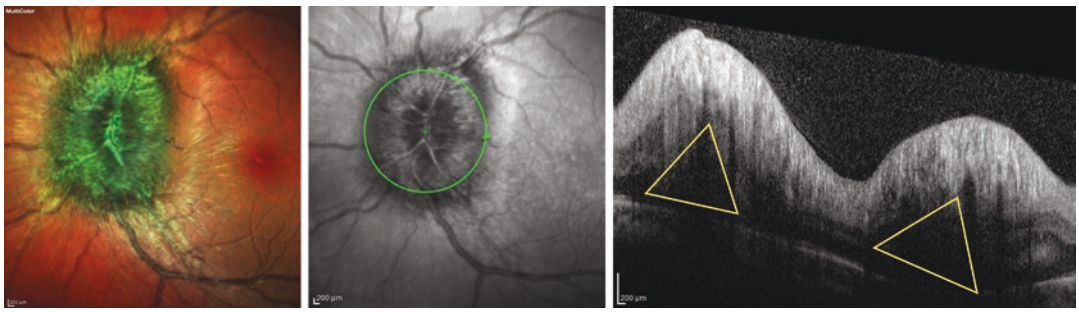

Fig. 4.2 Papilledema. The OCT scan of the optic nerve head shows the triangular subretinal hyporeflective space (in yellow) in papilledema. (Image courtesy of Dr. Gilda Cennamo, University of Naples Federico II)

4.5 Ophthalmic Ultrasonography

The ophthalmic ultrasonography, performed by a skilled echographer, is a very useful test [24]. It is more sensitive than computed tomography (CT) or MR in the assessment of intraocular, retinal, or subretinal diseases and is helpful to CT or MR in the characterization of orbital mass lesions (Figs. 4.3 and 4.4). If CT and MR have been used in the initial evaluation of an orbital abscess or mid- or anterior orbital inflammatory disease, such as posterior scleritis, serial B-mode ultrasound is adequate in monitoring the response to treatment, thus minimizing the need for several neuroradiological studies. If CT and MR have evidenced a mid or anterior orbital tumor or an infiltrative process, the echography is helpful in assessing the internal acoustic characteristics of the lesion.

Lymphoma, melanoma, and cysts have no or low internal reflectivity; other mass lesions have mid- (such as schwannoma) to high internal reflectivity (such as many carcinomas or cavern-

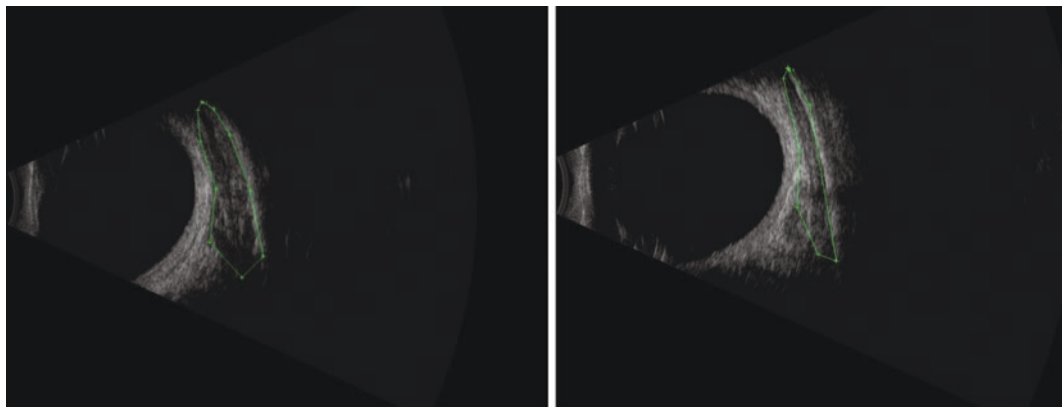

Fig. 4.3 The ultrasound b-scan picture is compatible with a lymphangioma-type pathology located in the superior-medial angle of the orbit. The lesion is bounded by green lines; and it shows a decreased volume after compression, this is a typical phenomenon of cystic tumors. (Image courtesy of Dr. Maria Angelica Breve, University of Naples Federico II)

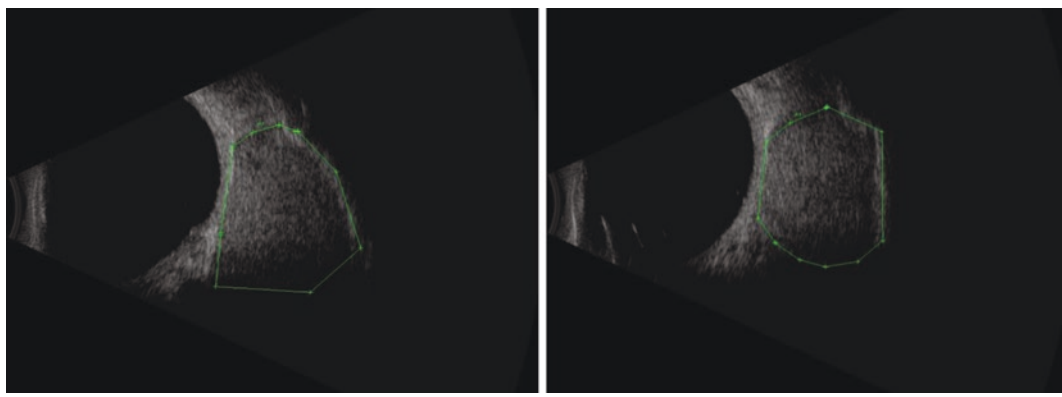

Fig. 4.4 The ultrasound b-scan picture is compatible with a solid round lesion located in the superior-medial angle of the orbit. The lesion is bounded by green lines; and it shows a fixed volume after compression, this is a typical phenomenon of solid tumors. (Image courtesy of Dr. Maria Angelica Breve, University of Naples Federico II)

ous hemangioma). Ultrasonography also allows real-time dynamic assessment of tumor adherence or infiltration to adjacent tissues [25–28].

The B-mode echography is also an useful tool for image-guided fine needle aspiration biopsy (FNAB) of orbital tumors. This scarcely invasive diagnostic technique is mostly performed for the identification of metastatic tumors, allowing definitive diagnosis without necessity of open biopsy. It also aids in the cytopathologic diagnosis of lacrimal gland mass inflammations, lymphoma, pleomorphic adenoma, or carcinoma.

Ultrasonography can be done quickly in the clinic at a low cost. It is the unique imaging technique able to reveal dynamic changes such as compressibility and internal reflectivity, as well as the cystic nature of lesions. Doppler imaging provides information regarding vascular flow characteristics within a lesion. This modality is typically applicable to anteriorly located mass lesions.

Thus, ultrasonography is an important complementary imaging technique to CT and MR for defining the orbital mass lesions.

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