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Optic nerve and chiasmal disease

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Abstract This review of optic nerve and chiasmal disease briefly outlines the clinical assessment and the use of diagnostic testing in the topical diagnosis of lesions of the anterior visual pathways. The commoner pathological entities including inflammatory, vascular, heredofamilial and compressive lesions are then summarised with specific reference to important points in the diagnosis and management. Specific disorders described include optic neuritis, papilloedema, ischaemic optic neuropathy, giant cell arteritis, Leber's hereditary optic neuropathy and pituitary tumours.

Clinical assessment

Symptoms: mode of onset of visual loss

The mode of onset of visual loss helps with the differential diagnosis: *transient monocular visual loss* lasting for seconds or minutes suggests papilloedema or embolic amaurosis fugax, respectively; *sudden, irreversible monocular blindness* points to a retinal vascular occlusion, intraocular haemorrhage, retinal detachment or ischaemic optic neuropathy [ION]; *monocular visual loss evolving over one or more days* suggests inflammatory optic neurits; and *visual loss evolving over weeks or months indicates* an extrinsic compressive lesion such as a pituitary tumour. Pain on eye movements is often reported in optic neuritis.

Physical signs: acuity loss,
visual field defects and pupillary changes

Because of the retinotopic organisation of the anterior visual pathways (Fig. 1), structural lesions affecting retinofugal axons passing from ganglion cells in the inner retina to the lateral geniculate ganglion are associated not only with symptomatic acuity loss but also with characteristic field defects. By contrast, diseases of media of the eye typically cause loss of acuity without a field defect. Ophthalmoscopically visible lesions of retina cause focal scotomas, and optic nerve diseases cause a scotoma which is usually central but may also be altitudinal. Optic disc swelling (papilloedema) initially cause an enlarged blind spot with preserved acuity while chiasmal lesions causes a bitemporal hemianopia, which is usually asymmetric with acuity loss in one or both eyes as well. Pure optic tract lesions may cause non-congruous homonymous field loss with preserved acuity. The relative afferent pupil defect is almost invariably present in unilateral optic neuropathies, as is subjective loss of colour appreciation.

Critical clinical skills in assessing optic nerve and chiasmal disease

With the following clinical examination techniques it should normally be possible to separate cases of visual loss arising from the ocular media and outer retina from

those due to inner retina (ganglion cells), optic nerve and chiasmal dysfunction:

– Snellen acuity chart and pin-hole [1]
– Confrontation field testing with red targets
– Quantitative perimetric visual fields (and artefacts!)
– Qualitative colour vision testing
– Pupillary light reflexes [2]
– Dilated and undilated fundoscopy

Special investigations

Fluorescein angiography

Early capillary dilation on the disc surface and late hyperfluorescence are hallmarks of pathological optic disc swelling, whilst autofluourescence is seen in pseudopapilloedema due to optic disc drusen. Retinal inflammatory and degenerative lesions which may be associated with neurological disease (e.g. periphlebitis in neurosarcoidosis) can also be shown [3].

Clinical electrophysiology

Visual evoked potentials

The visual evoked potentials (VER) reflect the integrity of the central visual field throughout the entire afferent visual system but is non-specific in localising a lesion to the retina, optic nerve or cortex. A flash generated VER is stimulated by luminance change while a pattern VER is stimulated by contrast change. The complex wave form is described in terms of amplitude and latency according to characteristics of the P_1 wave which is a large positive deflection occurring at around 100 ms. Amplitude varies widely amongst normal subjects but latency is constant.

Pathological conditions of the anterior visual pathways yield relatively non-specific abnormalities of the VER, but the test is nonetheless useful especially when carried out in conjunction with flash- and pattern-generated electroretinography (ERG; see below). Latency delay with relative amplitude preservation is found in demyelination, even long after the acute episode and following visual recovery and also in optic nerve compression. Amplitude reduction is a feature of ION and other conditions in which axonal loss is prominent compared to conduction block.

Additional specificity can be achieved by half field testing, whereby differences in amplitude and latency recorded from each hemisphere after uniocular stimulation can be used to assess abnormalities of the optic chiasm. Further refinements in VER techniques which are currently undergoing clinical evaluation include differential testing of the parvo- and magnocellular systems by use of test targets which selectively stimulate chromatic, temporal and spatial functions, and also the discrimination between striate, extrastriate and subcortical responses [4, 5].

Electroretinography

Simulation of the eye with either a flash or a pattern reversal system results in a recordable potential from the retina which has two prinicpal components: the a-wave which reflects the activity of the photoreceptors, and the b-wave which arises from the inner nuclear layer and the Muller cells. Disorders predominantly involving cones can be distinguished from those involving rods by performing the test in light (photopic) and dark (scotopic) conditions. In primary retinal diseases, such as diabetic retinopathy, pigmentary retinopathies and dystrophies, and in toxic degenerations, both the a-wave and the b-wave are abnormal. Exceptions to this are seen in central retinal artery occlusion where in the acute phase the photoceptors which derive their blood supply from the choriocapillaris are preserved, and therefore the a-wave persists while the b-wave is extinguished. Superimposed on the b-wave in a normal retina are a series of rhythmic oscillations referred to as oscillatory potentials, which may be of diagnostic importance when lost in the early stages of both diabetic retinopathy and in pigmentary retinopathies. When visual loss results from diseases which involve the ganglion cells or optic nerve, such as Tay-Sachs disease, glaucoma and optic neuritis, the ERG in response to flashes of light is usually normal. Only in advanced cases of retinal ganglion cell loss does the flash ERG become abnormal, as a result of secondary transynaptic degeneration of outer retinal elements (bipolar cells and photoreceptors).

In practice, the clinician must distinguish between visual acuity loss of retinal and optic nerve origin. Sometimes a pattern ERG (PERG) is useful. Although the origins of the PERG are not entirely clear, it is accepted that the response does arise from retinal structures proximal to the photoreceptors, and it is this anatomical localisation which, together with fact that pattern reversal stimuli test the central retina only, gives the test its potential power. Unlike the flash ERG, the late components of the PERG are abnormal in optic nerve and retinal ganglion cell layer disorders. Therefore the PERG may be of value in discriminating subtle cases of central visual loss of central retinal origin, when early and late components are abnormal from visual loss of optic nerve origin when the late component alone is abnormal. By using specialised light projection systems it is possible to test cone function by projecting a spot stimulus to the fovea rather than across the entire field (ganzfield), and foveal ERGs may develop as an alternative to the PERG [6, 7].

Ultrasound, Doppler and colour Doppler flow mapping (colour Doppler imaging)

Real-time B-mode ultrasound can show calcified buried optic disc drusen causing pseudopapilloedema, and optic nerve sheath distension in chronic papilloedema can readily be demonstrated. Disc swelling due to local scleral or optic nerve sheath swelling can also be shown (scleritis and perineuritis), and also optic nerve calcification in cases in which the distinction between meningioma and glioma is difficult.

In addition to carotid vascular studies, Doppler ultrasound has important applications within the orbit. The demonstration of reversed blood flow with an arterial wave form in the superior opthalmic vein using colour Doppler flow mapping is of diagnostic importance in patients with carotid-cavernous and dural fistulas, and is a feature which can be used to monitor clinical progress. In addition, reduced ophthalmic artery blood flow may be shown in cases in which the chronic ocular ischaemia complicates occlusive carotid disease [8–12].

Neuro-imaging

Some features of optic nerve disease on magnetic resonance imaging (MRI) are the following:

Intrinsic lesions
– Optic neuritis: increased signal on fat-suppression short

T1 inversion recovery sequences
– Radionecrosis: late gadolinium enhancement
– Neurosarcoid: meningeal enhancement associated with optic nerve signal
– Optic glioma: demonstration of perineural arachnoid gliomatosis

Extrinsic lesions
– Perioptic meningioma: gadolinium enhancement and demonstration of intracranial extension in primary optic nerve sheath meningioma

The power of the magnetic resonance imaging (MRI) in the diagnosis of visual failure is widely recognised and need not be detailed here. However, occasions when computed tomography (CT) remains useful or preferable should be noted. This relates especially to the orbit, where unless surface coils and fat suppression techniques are available, MRI is frequently inferior to CT. Specific advantages of CT include the demonstration of bone lesions (fractures, neoplastic erosion), acute haemorrhage (blood invisible on MRI initially), and the demonstration of dystrophic calcification in optic nerve head drusen and optic nerve sheath meningioma [13–17].

Common pathological conditions of the optic nerve and chiasm

Pathophysiology of optic nerve disease and the swollen optic disc

Pathophysiology of optic nerve disease

The common feature of optic disc swelling regardless of the cause is stasis of both fast and slow retrograde axoplasmic transport. This results in axonal distension which is ophthalmoscopically visible at the level of the prelaminar optic nerve as a swollen disc. As a result of axonal swelling, secondary vascular changes follow which include capillary dilation, microaneurysms, haemorrhages and cotton wool spots on the disc and neighbouring retina and dilation of the central retinal vein [18].

In optic atrophy the reduction in the volume of the capillary bed within the neuroretinal tissue of the nerve head accounts for ophthalmoscopic pallor, together with loss of tissue volume and astrocytic proliferation [19].

Differential diagnosis of the swollen optic disc

Current convention reserves the term papilloedema for the syndrome of pathological disc swelling due to raised intracranial pressure. The remainder of cases are designated either as disc swelling arising from a local (i.e. in the eye or orbit) lesion or as pseudopapilloedema when the appearance arises as a congenital anomaly. Disc oedema due to local optic nerve disease is normally associated with early loss of acuity and colour vision, together with central, arcuate or altitudinal field defects. Involvement is usually unilateral, and consequently there is a relative afferent pupil defect.

The appearance of “pseudopapilloedema” mimicking true papilloedema of raised intracranial pressure may cause difficulties. All too frequently the patient with headaches is wrongly thought to have raised intracranial pressure from the fundoscopic finding of elevated discs. A systematic approach is essential to resolve this difficulty [20]:

– Does the headache have the features of raised pressure (more severe on awakening, exacerbated by coughing)?
– What is the approximate refraction of the eye? Hypermetropes often have small, crowded elevated discs which are a normal variant. Similarly, myopes frequently have dysverted or tilted discs with an elevated indistinct margin supra-nasally.
– What can be can be learnt on careful fundoscopy? In true papilloedema secondary vascular changes are usually prominent with pre-papillary capillary dilation, haemorrhages and exudates as well as axonal swelling. Spontaneous pulsation of the central retinal vein is not

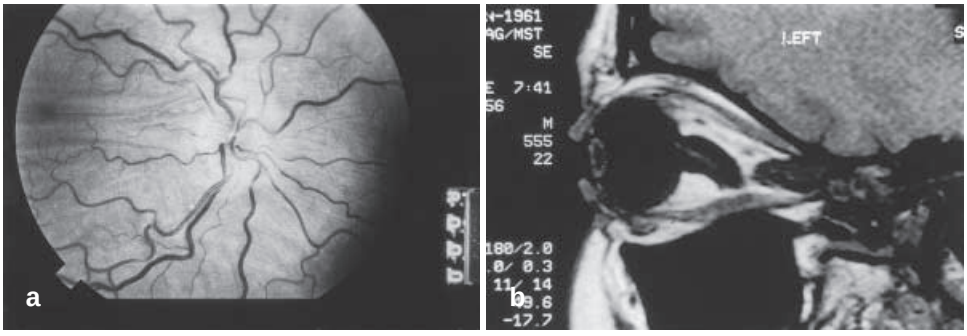

Fig. 1 **a** Red-free fundus photograph showing chronic optic disc swelling and associated choroidal folds extending under the retina in the horizontal plane. **b** Sagittal MRI of left orbit of the same patient showing a massively dilated optic nerve sheath. This patient had long-standing idiopathic intracranial hypertension and associated severe visual field loss

always observed in the normal subject, but when seen, raised intracranial pressure is highly unlikely. Superficial disc drusen appear as refractile bodies on the disc surface causing abnormal elevation, and hamartomas produce a mass effect without these vascular changes. Buried disc drusen result in an indistinct disc margin often with an anomalous retinal vascular pattern with trifurcations of the first-order vessels at the disc margin. Sometimes further investigations are required. Intrapapillary calcification of drusen can be seen on CT and ultrasound. Fundus fluorescein angiography shows an absence of any vascular dilation or late leakage from the the prepapillary capillaries: typically the disc shows late staining with fluorescein but without leakage beyond the disc margins. Buried disc drusen show autofluorescence when illuminated with blue light (Fig. 1).

Visual loss secondary to chronic raised intracranial pressure

More than 80% of patients with idiopathic intracranial hypertension have been shown eventually to develop some degree of visual loss, and up to 10% have significant field defects and acuity loss rather than simply obscurations and blindspot enlargement, and careful follow-up is required. Medical measures to control headache and visual symptoms in chronic raised intracranial pressure may be both ineffective and poorly tolerated. These include weight loss, carbonic anhydrase inhibitors, corticosteroids and serial lumbar punctures. Surgical CSF diversion procedures may be required and include lumbo-peritoneal shunting and optic nerve sheath decompression (fenestration) [21–23].

Heredofamilial optic neuropathies

Leber's hereditary optic neuropathy (LHON) is readily distinguished from other hereditary optic neuropathies by a characteristic clinical presentation: sudden central visual loss in the second to third decade of life and usually in males. Weeks to months later the second eye is involved, although bilateral simultaneous visual loss may occur.

Table 1 Primary mt-DNA mutations associated with LHON

Primary mutation	Frequency in northern Europe	Recovery rate
11778	50–60%	2–17%
3460	15–30%	20–40%
14484	10%	37–50%

Colour vision is severely affected, and perimetry shows either a central or caecocentral scotoma. Unlike in optic neuritis, there is no pain on eye movements, and recovery is both exceptional and delayed by months or even years when it occurs.

LHON is due to one of a number of point mutations in mitochondrial DNA, most commonly occurring at the 11778 location with the substitution of adenine for guanine. This mutation has not been found in control subjects. Other pathogenic mutations arise at sites 3460 and 14484. This information carries important prognostic implications. The visual recovery rate may only be 4% in patients with the 11778 mutation, but up to 37% of those with a defect at site 14484 may recover. (Table 1) Female involvement in these mutations appears to be rare. In Europe the 11778 and 3460 mutations account for at least 90% of families with this disease as defined by a subacaute optic neuropathy with a positive maternal family history. Epigenetic factors are also important and tobacco and alcohol have been reported to play a role in the phenotypic expression of the 3460 and 14484 mutations. This would explain the association between LHON and heavy tobacco consumption, perhaps in nutritionally deficient patients [24].

An effective treatment for LHON remains to be found: the use of vitamin B_{12} is unproved and probably ineffective, but it may be valuable to counsel individuals at risk (after first eye involvement, or unaffected maternal relatives) against tobacco and alcohol use and on the benefits of a balanced diet.

Inflammatory and ischaemic disorders

Typical optic neuritis

In a typical episode of optic neuritis there is an acute or subacute onset of visual loss progressing over hours or a few days with function reaching its nadir by 1 week. Unilateral involvement is usual (the less common bilateral disease is a characteristic of the post-viral form especially in children). Pain on eye movements and mild tenderness of the globe are often present at or shortly before the onset of visual symptoms. Visual loss may range from field and contrast defects with maintained acuity to profound acuity loss with no perception of light. Formal perimetry may show a variety of defects ranging from generalised depression to a central scotoma or altitudinal and nerve fibre layer defects.

Usually vision begins to recover after 2–3 weeks and stabilises by 5–6 weeks. Many patients experience virtually complete restoration of acuity, but persistent subtle residual defects of colour vision, contrast sensitivity, depth perception and cortical evoked potential latency changes are nearly always demonstrable. In some individuals acuity only makes a partial recovery: rarely there is no recovery whatsoever after initial loss. In retrobulbar neuritis, the fundus is by definition normal at the time of onset but subsequently disc pallor may be marked. The final degree of optic atrophy is not closely correlated with the level of visual recovery [25, 26].

Most patients with multiple sclerosis (MS) develop optic neuritis at some stage, and 40–70% of patients with clinically isolated optic neuritis go on to develop multiple sclerosis. Subclinical abnormalities of contrast sensitivity and evoked potentials in the fellow eye at the time of acute symptoms imply disseminated white matter lesions in space and time and a diagnosis of MS. Recurrent attacks in the same eye occur in 20–30% of cases, but the predictive significance of this for subsequent MS is uncertain. In our hospital it is current practice to treat with high-dose steroids only those patients with acute optic neuritis who: (a) have bilateral acute involvement, (b) have poor vision in the fellow eye due to some other disease process, or (c) have a severe visual deficit and severe pain. Patients with optic neuritis require informed counselling usually from a neurologist, remembering that MRI abnormalities in the brain at the time of visual symptoms may be associated with increased risk of MS in the future, *but* this cannot be correlated with clinical effects or eventual disease burden, and the likelihood of developing MS is not correlated with the likelihood of serious neurological disability.

Atypical optic neuritis

In patients who fall outside the usual age range for optic neuritis, who are systemically ill, whose vision does not start to recover within the normal time frame or who have a history of paranasal sinus disease, a careful search for underlying causes is mandatory. This includes autoimmune, infective and granulomatous disease and extrinsic compression of the optic nerve from tumours, aneurysms and paranasal sinus lesions [28].

Ischaemic optic neuropathy

Anterior ION due to acute infarction of the optic nerve head is a common cause of visual loss in patients past middle age. The blood supply of the optic nerve head is primarily derived from choroidal and posterior ciliary branches of the ophthalmic artery. The posterior ciliary arteries arise independently from the ophthalmic artery and

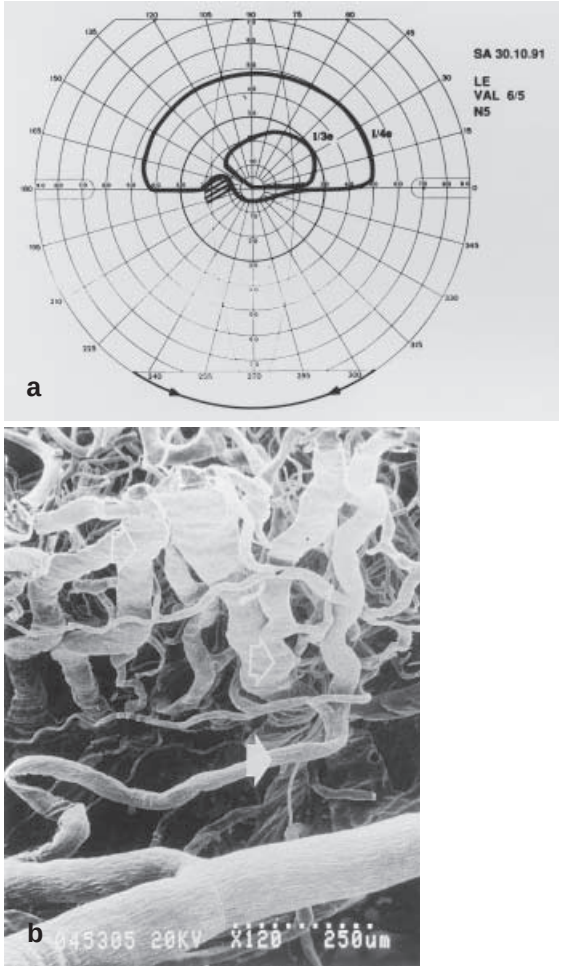

Fig. 2 **a** Dense lower altitudinal field loss (altitudinal scotoma) in a patient with ischaemic optic neuropathy. **b** Methylmethacrylate vascular cast study showing a slender branch of a posterior ciliary artery (*arrows*) contributing to the blood supply of the optic nerve head (*above*). Atherosclerotic occlusion of these vessels is thought to underlie the pathogenesis of ischaemic optic neuropathy

may number from two to four. Usually they form medial and lateral groups, and because they are end arteries a watershed zone is formed between territory perfused by the medial group and territory perfused by the lateral group. When the watershed zone traverses the optic nerve head, this structure is particularly vulnerable to haemodynamic disturbance resulting in infarction and a permanent visual deficit [29–31].

Anterior ION occurs in two forms: non-arteritic and arteritic, the latter usually due to giant cell arteritis. In a typical attack of non-arteritc anterior ION there is a painless acute onset of loss of acuity and field (usually in an altitudinal pattern) in a patient aged between 50 and 70 years (Fig. 2). The onset is usually acute, although some patients experience progressive step-wise visual loss over the period of 1–2 weeks. The lack of pain and the age at onset help to distinguish ION from inflammatory optic neuritis. A relative afferent pupil defect indicates that the visual loss is of neuroretinal origin and fundoscopy reveals pallid optic disc swelling which may be sectoral rather than general, often with one or more splinter haemorrhages at the disc margin.

In addition to the risk factors for vascular disease, there is evidence of an anatomical predisposition as low hypermetropic eyes with small optic discs with low cup-disc ratios ("the disc at risk") are especially vulnerable [12]. The fellow eye is eventually involved in up to 40% of cases.

Arteritic ION is often due to giant cell arteritis. Systemic symptoms of anorexia, malaise, proximal arthralgia and myalgia (polymyalgia rheumatica), together with headache, scalp tenderness and jaw claudication usually precede visual loss. The visual deficit is often severe, with pallid swelling of the entire optic disc and an additional retinal cherry red spot when ophthalmic artery disease results in a combined posterior ciliary and central retinal artery occlusion.

Management steps in acute ION include increasing optic nerve head perfusion pressure by reducing the intraocular pressure with oral or topical carbonic anhydrase inhibitors and identification and treatment of systemic vascular risk factors (carotid disease, hypertension, diabetes, pro-thrombotic states, smoking) to reduce the risk of subsequent stroke or fellow eye involvement. High-dose corticosteroids and temporal artery biopsy are used in suspected giant cell arteritis. Normally this comprises 80 mg prednisolone daily plus 200 mg intravenous hydrocortisone immediately. When the patient presents with second eye involvement, or the erythrocyte sedimentation rate does not fall rapidly after commencing oral steroids, a 3-day course of pulse intravenous methylprednisolone 10 mg/kg is given. A delay of up to 48 h does not affect the biopsy result after the patient has started corticosteroids. Anti-platelet agents or low-dose heparin may also be of value in cases of progressive visual failure [32].

There is probably no role for optic nerve sheath decompression in ION, in spite of earlier hopes [33].

Tumours and the optic nerve

Primary optic nerve tumours

Meningiomas. Optic nerve function may be directly impaired by meningiomas arising primarily from the arachnoid cells of the optic nerve sheath, usually in the orbital portions of the nerve. More commonly, however, optic nerve sheath meningiomas arise secondarily from the sphenoid wing, tuberculum sellae or olfactory groove and subsequently invade the optic canal and orbit. The treatment of orbital meningiomas remains controversial. In sphenoid wing tumours which principally have an intracranial component surgical resection is the treatment of choice. By contrast, in primary optic nerve sheath meningiomas can often be managed conservatively [34].

Optic and opto-chiasmal glioma. Primary glial tumours of the anterior visual pathways (optic glioma) are seen in two fundamentally different forms: benign gliomas of childhood and rare malignant glioblastomas of adulthood. These are discussed as separate entities, even though they may both present with visual loss and orbital mass effects. Involvement may be of one optic nerve, intraorbital or intracranial, of both optic nerves, of the optic chiasm and of the hypothalamus or thalamus. Malignant optic gliomas of adulthood do not arise from indolent childhood tumours [35]. A careful search for the cutaneous stigmata of neurofibromatosis 1 is important. Chiasmal optic gliomas of childhood are commoner than those confined to one optic nerve. Cases present with visual loss, visual field defects and nystagmus. The visual loss is generally bilateral and field analysis may show either central scotomas or temporal hemianopic defects.

The optic nerve in systemic cancer

Infiltration of the meninges by systemic cancer may sometimes result in painless progressive visual loss due to retrobulbar optic neuropathy. Adenocarcinoma of the breast and lung are the common causes of this syndrome, which may be diagnosed by demonstrating meningeal contrast enhancement on MRI together with cytological findings of neoplastic cells in the CSF [36].

Compressive optic neuropathies

Thyroid ophthalmopathy. Patients with thyroid ophthalmopathy (Graves' disease, dysthyroid eye disease, thyroid eye disease) show signs of orbital congestion, proptosis and ophthalmoplegia in varying degrees. The extraocular muscles and orbital fat are expanded by inflammatory infiltrate, mucopolysaccharide deposition and, later in the disease process, by fibrotic tissue. The optic nerve may

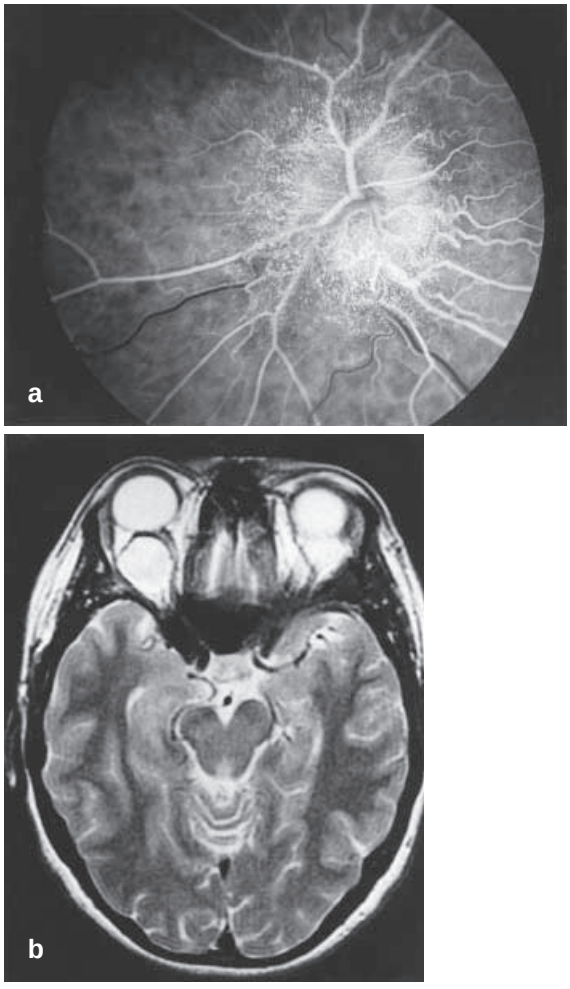

Fig. 3 **a** Arterial phase fundus fluorescein angiogram showing pre-retinal capillary dilation in pathological optic disc swelling; these vessels leak fluorescein dye profusely later in the study. **b** T2-weighted MRI of the right orbit of the same patient showing a large haemangioma displacing the optic nerve medially and causing the disc swelling

become compressed by swollen muscles and fat at the confined space of the orbital apex. Visual failure most commonly occurs in the inactive, fibrotic phase of the disease, often without conspicuous features of anterior orbital disease (proptosis, lid swelling). However, in some cases optic nerve compression forms part of the presenting, acute inflammatory phase, in which case chemosis and conjunctival injection particularly at the sites of insertion of the rectus muscles is conspicuous. Dyschromatopsia, choroidal folds and optic disc swelling support the diagnosis, but often fundoscopy is normal. Visual function is best monitored by serial measures of acuity and fields [37] (Fig. 3).

Toxic and nutritional optic neuropathies

Slowly progressive bilateral visual loss with central or centrocaecal scotomas and loss of colour vision always suggests the possibility of optic nerve failure due to a toxic cause or nutritional deficiency. A careful family history excludes heredofamilial disease. In whites the toxic-nutritional optic neuropathies are usually due to a multifactorial combination of dietary B_1 and B_2 vitamin complex deficiency, ethanol abuse and high levels of tobacco consumption. Less commonly, malabsorption and vitamin B_{12} deficiency states produce the same picture.

In other patient populations, additional associations exist. Social dislocation amongst refugee groups in Africa is associated with consumption of uncooked cassava, which contains high levels of cyanide. Adequate detoxification mechanisms may be defective when there is an additional vitamin B_{12} deficiency state. Human T-lymphotropic virus 1 retrovirus infection has been implicated in the syndrome of combined spastic paraparesis and optic neuropathy in African-Americans. A recent epidemic of optic neuropathy in Cuba has been extensively investigated from the point of view of both genetic and environmental factors. Multiple dietary deficiencies have been identified as the probable cause [38]. There is evidence of a possible synergistic effect between dietary deficiencies, mitochondrial DNA mutations and underlying adenosine triphsophate deficiency in the onset of optic neuropathy in some of these patients [39].

Applied physiology and clinical features of chiasmal disease

Bitemporal hemianopia is the clinical hallmark of chiasmal lesions. In some individuals the intracranial portion of the optic nerves is relatively long, and the chiasm is formed in the posterior cistern (post-fixed), with the consequence that expanding sellar lesions may cause optic nerve compression rather than chiasmal compression (Fig. 4). Conversely, other individuals may have a pre-fixed chiasm so that mass lesions of the pituitary fossa may impinge on the optic tract. As fibres from the optic nerve enter the chiasm, those serving the nasal and temporal hemiretinas become separated and the anterior nasal projecting fibres loop into the contralateral optic nerve as they cross the midline. Consequently lesions of the anterior chiasm result in a contralateral temporal hemianopic field defect with an ipsilateral central scotoma. Lesions at the posterior chiasmal notch may selectively involve only dorsal crossing fibres serving central vision resulting in hemianopic scotomata. Macular fibres are present throughout the chiasm, and because they serve the central 5° of field and form at least 30% of all the fibres, this has two simple clinical consequences: Firstly, chiasmal lesions usually cause defects in central vision (e.g. acuity or

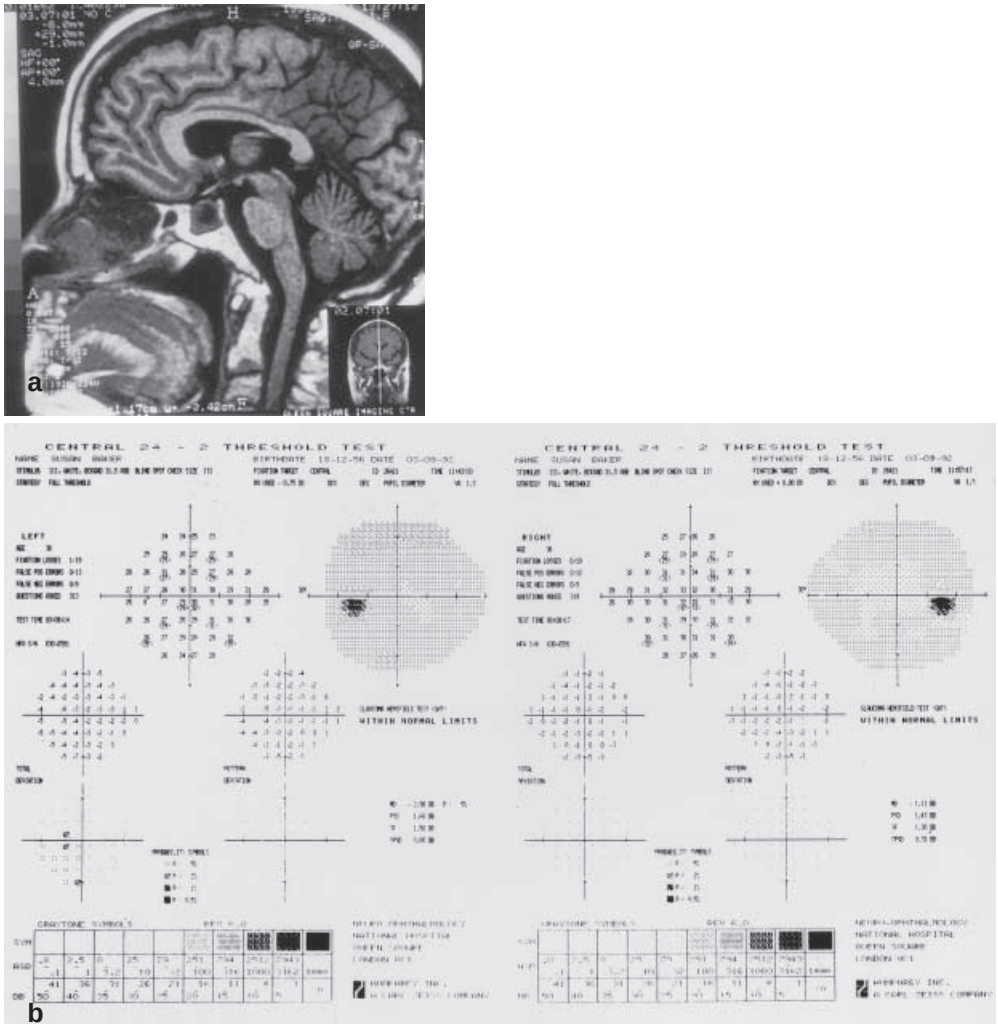

Fig. 4 **a** Sagittal T1-weighted MRI showing an intrasellar mass. The optic chiasm is clearly seen above the lesion separated by CSF. **b** Normal Humphrey visual field study from same patient; intrasellar masses do not cause visual field defects

colour tasks) in one or both eyes. Secondly, temporal defects respecting the vertical meridian are usually apparent when testing the central visual field if appropriate stimuli are used.

Patients with a bitemporal hemianopia may present with double vision in the presence of normal eye movements. When there are dense temporal field defects, binocular fusion is not supported by overlapping temporal hemifields in the visual cortex. As a result, latent heterophorias may readily break down, leading to variable vertical or horizontal diplopia. In other instances there may be difficulties in performing tasks requiring depth perception and judgement. This reflects post-fixational blindness so that when the subject fixates on a near target, objects beyond may project onto both nasal hemiretinas and become invisible. Faulty motion perception may arise as a result of asymmetric optic nerve conduction latencies (Pulfrich phenomenon).

Diplopia may also be paretic: sometimes this is due to a lateral rectus palsy complicating raised intracranial pressure as late presentation occurs with obstructive hydrocephalus. More commonly diplopia reflects parasellar involvement of one or more of the ocular motor nerves either by compression of the cavernous sinus or direct invasion [40].

Common pitfalls in visual field testing in suspected chiasmal disease

In the knowledge that the hallmark of chiasmal disease is a bitemporal hemianopia, the clinician may wrongly ascribe every case with bitemporal field loss to chiasmal disease. Bitemporal field defects are also found in:

- Massive papilloedema due to blindspot enlargement
- Centrocaecal scotomas
- Tilted, dysplastic optic discs
- Uncorrected refractive errors
- Baggy overhanging upper eyelids
- Bilateral nasal retinal disease (e.g. sector retinitis pigmentosa)

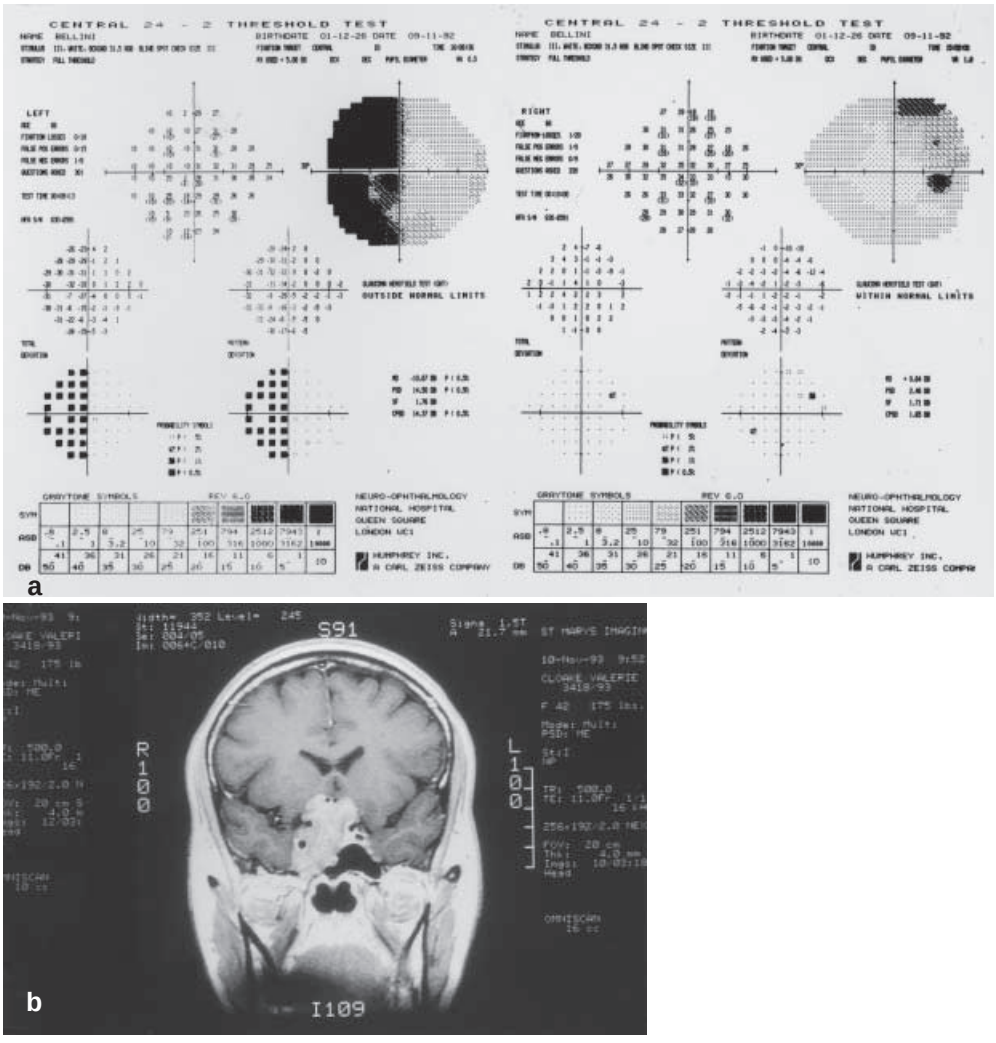

Fig. 5 **a** Humphrey visual field study showing a dense left temporal field defect and subtle changes in the right temporal hemifield. **b** Contrast enhance T1-weighted coronal MRI of same patient showing a mass arising from the pituitary fossa with significant suprasellar extension explaining the field loss

In practice, these errors are much more likely on confrontation field testing than on some form of quantitative or semiquantitative testing when close attention is given to ensuring accurate fixation. In this way it can be determined whether the defect respects the vertical meridian and thereby can correctly be attributed to a chiasmal rather than to a more anterior cause. However, automated perimetry may create its own problems, for example, fixation is checked automatically by retesting stability of the blindspot, and this cannot be carried out in a subject with dense temporal defects. As a rule, we have found the combination of confrontation testing with red-coloured targets, tangent screen testing to a red target together with perimetry using the manual Goldmann or the automated Humphrey machines to be much more reliable than any one technique on its own [41] (Fig. 5).

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