Chapter 7

The optic chiasm

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INTRODUCTION

The optic chiasm is formed when the optic nerves come together in order to allow for the crossing of fibers from the nasal retina to the optic tract on the other side. This enables visual inputs from the nasal half of the contralateral eye and the temporal half of the ipsilateral eye to be processed by the occipital lobe, vital for visual processing.

ANATOMY

The optic chiasm lies above the sphenoid bone, over the diaphragma sellae (Fig. 7.1); anatomical variations are common and can influence the presenting visual symptoms and signs in chiasmal disorders. In the majority of cases (79% of a series of 125 autopsy cases) the chiasm is situated overlying the diaphragma sellae (Schaeffer, 1924), prefixed when lying above the tuberculum sellae or within the sulcus chiasmatis (17% of cases), and post-fixed when it lies above or even behind the dorsum sellae (4%) (Bergland et al., 1968).

The intracranial optic nerves rise from the canal to the chiasm at an angle of 15–45°. The pia mater is contiguous with that of the nerves and that of the anterior tracts. The chiasm lies posterosuperiorly within the wall of the third ventricle and anteriorly with the cerebrospinal fluid within the chiasmatic cistern (Fig. 7.1). Above lies the hypothalamus and below the pituitary gland. The infundibulum of the pituitary lies immediately posteriorly and the mamillary bodies behind this, medial to the two optic tracts.

The internal carotid arteries lie to either side and the anterior communicating artery lies directly above; aneurysms of the anterior communicating artery or first segment of anterior cerebral artery may therefore compress the chiasm.

It is well known that fibers from the nasal retina cross within the chiasm; the ratio of crossed to uncrossed fibers within the chiasm is 53:47 (Kupfer et al., 1967). This crossing is of necessity very precise; only fibers from the temporal retina pass to the ipsilateral lateral geniculate nucleus, and only those from the nasal retina pass contralaterally. Those that cross do so as soon as they enter the chiasm and maintain their rostral to caudal position within the chiasm. Those that are uncrossed continue in the lateral chiasm into the ipsilateral tract. Fibers from the macula on each side, which account for the majority of fibers within the chiasm (Hoyt and Luis, 1969), are found more centrally and caudally than other fibers.

Rarely the chiasm may fail to develop (achiasmia). Patients studied have associated midline developmental disorders but intact fields; the visual-evoked potential characteristically is monocular (Sami et al., 2005).

BLOOD SUPPLY

The blood supply is variable, but in general comes from feeder vessels arising from branches of the anterior communicating artery, anterior cerebral, posterior communicating, posterior cerebral and basilar arteries (Wollschlaeger et al., 1971). The dorsal parts are supplied by branches of the internal carotid arteries and the more ventral parts from the posterior circulation. There is considerable collateral supply, with the result that infarction of the chiasm is very rare indeed.

VISUAL FIELD DEFECTS IN CHIASMAL DISORDERS

A careful study of the bilateral visual field defect acquired in chiasmal lesions can provide important
information on the nature and in particular the site of the causative lesion. Traditionally field defects have been divided anatomically into the anterior angle, the body, the posterior angle, and the lateral aspect of the chiasm (Fig. 7.2).

**The anterior angle**

Lesions that involve the anterior angle of the chiasm, the point at which the optic nerve passes into and forms the chiasm, may show a junctional scotoma (Traquair, 1949) since there is at this point separation of the crossing and uncrossed fibers. If the lesion is not large this will result in an ipsilateral monocular central field defect (from the optic nerve) and a small contralateral upper temporal homonymous field defect from involvement of the crossing fibers anteriorly. If the nerve itself is not affected as well, the defect will be monocular and temporal, since only the crossing fibers are involved; if only the macular fibers are involved then the defect is a midline-obeying temporal scotoma. This defect may be very small and can be

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**Fig. 7.1.** T1-weighted (A) axial and (B) coronal magnetic resonance images showing the position and immediate relationship of the chiasm to adjacent anatomical structures.

**Fig. 7.2.** Visual field defects seen in chiasmal disorders. (A) Ipsilateral central field defect and contralateral junctional scotoma in a lesion involving the anterior angle of the chiasm; (B) also at the anterior angle, when only the crossing macular fibers are affected; (C) bitemporal hemianopia when the body of the chiasm is affected; (D, E) bitemporal upper and lower quadrantanopia when the lesion compresses the chiasm from below and above respectively; (F) bitemporal hemianopic scotomas when a lesion involves the posterior angle of the chiasm; (G) a noncongruous hemianopia occurs when the tract is affected.
missed with kinetic perimetry; it is, however, usually picked up using static perimetry.

Wilbrand’s knee is said to be an extension of the crossing fibers from retinal ganglion cells located nasal and inferior to the fovea into the ipsilateral distal optic nerve; the contralateral field defect arises with involvement of these fibers. Horton (1977), however, has stated that Wilbrand identified an artifactual state in the examination of enucleated eyes. Nonetheless, the importance of the identification of a small asymptomatic contralateral defect in the presence of a symptomatic monocular defect greatly encourages the examining physician to proceed with haste to imaging studies rather than diagnose an inflammatory optic neuropathy incorrectly.

The body of the chiasm

Lesions that affect the body of the chiasm produce the typical bitemporal hemianopia (Fig. 7.3). This may affect the whole hemifield, upper or lower quadrants, and peripheral or central areas. The macula is usually, but not always, split. In general, central visual acuity is unaffected. Clearly it is possible that the field defects are not the same in each eye, since of course the causative lesion may exert a greater effect on one side than the other. Complete defects tend only to occur in the case of trauma. When the peripheral fields are affected, the defect progresses in a clockwise direction in the right and counterclockwise in the left (Cushing and Walker, 1915). Lesions compressing from above cause the lower

Fig. 7.3. (A, B) Goldmann field showing bitemporal hemianopia caused by a lesion involving the body of the chiasm (shown in Fig. 7.15).
field to be affected first and the defects are less congruous; those from below affect the upper fields first and tend to be more congruous in appearance.

The posterior angle of the chiasm

Lesions within the posterior aspect of the body of the chiasm produce bitemporal hemianopic scotomas (Larmande and Larmande, 1977). These may be confused with centrocecal scotomas (Fig. 7.4), but the former will not be associated with significant reduction in visual acuity, whereas the latter will. More posteriorly placed lesions will also involve the optic tract, with the result that the field defect will be predominantly one of a homonymous hemifield defect.

The lateral aspect of the chiasm

Involvement of this area by compression leads to a homonymous hemifield defect on the contralateral side.

VISUAL SYMPTOMS

When lesions damage chiasmal fibers there is a progressive loss of central visual acuity and a noticeable dimming of the temporal visual fields.

Patients may also notice a disturbance of depth perception at fixation which is due to crossing of the two blind hemifields after the point of fixation in convergence (Fig. 7.5). Hence an object that is seen at a distance to be behind another will disappear when the eyes focus on the object in front.

“Hemifield slide” is a phenomenon in which patients complain of difficulty reading and notice a doubling, loss, or vertical deviation of words on a horizontal line (see Chapter 9). Normally there is an overlap between the temporal field on one side and the nasal field on the other to allow fusion, which helps to stabilize ocular alignment, with the result that the image is single and clear. Patients with minor phoric deviations of the eyes

Fig. 7.4. Goldmann field showing a field defect associated with a lesion involving the posterior angle of the chiasm (shown in Fig. 7.10).

Fig. 7.5. Illustration of the mechanism of postfixational blindness in bitemporal hemianopia. Unshaded area is area of binocular vision; light shading is area of monocular vision; dark shading is area of blindness.
NEURO-OPHTHALMIC SIGNS

When a chiasmal lesion leads to optic nerve fiber atrophy, “band” or “bow-tie” atrophy is observed, in which atrophy is more evident in the nasal and temporal sides of the disc and relatively spared in the superior and inferior sectors (Unsold and Hoyt, 1980) (Fig. 7.6). This is due to involvement of only those fibers arising nasal to the fovea (associated therefore with the bitemporal hemianopia) being affected and passing into the nerve from these sections of the disc.

Tumors of the diencephalon and sellar region may be associated with seesaw nystagmus, a rhythmic synchronous alternating rotation of the eyes, in which one eye elevates and intorts whilst the other simultaneously depresses and extorts. In the case of parasellar lesions the brainstem fibers associated with the interstitial nucleus of Cajal are involved, and so the lesion must also affect these fibers in order to cause the syndrome.

DISEASES THAT MAY AFFECT THE CHIASM

Inflammatory diseases

Granulomatous infiltration

Sarcoidosis

Sarcoidosis is an autoinflammatory disorder of uncertain etiology in which granulomatous inflammation develops leading to tissue destruction and fibrosis. It is thought that some 10% of cases arise within or also involve the central nervous system. The neurological disorder is a meningeal-based inflammatory infiltration (Kidd and Beynon, 2003). The anterior visual pathway may be involved at any location; granulomas may arise within the disc, the optic nerve itself is involved most commonly, in which an intrinsic lesion may develop leading to the clinical syndrome of an optic neuritis (Frohman et al., 2003; Koszman et al., 2008; Figs 7.7 and 7.8), or an optic perineuritis may arise. Compression by an inflammatory mass at the orbital apex and, much less commonly, at the cavernous sinus (Zarei et al., 2002) may occur and be associated with ophtalmoparesis and trigeminal neuropathy. The tracts may also be affected (Frohman et al., 2003).

Involvement of the chiasm comes about by the development of an inflammatory mass within the pituitary gland (Guoth et al., 1998). This arose in 1 in 14 patients in one series (Chen and McLeod, 1989) and 4 in 100 in my own personal series. Patients present with symptoms of hypopituitarism and with visual field defects in keeping with a chiasmal problem. Pain is...
uncommon and the syndrome appears to be indistinguishable from pituitary adenoma both on clinical and on radiological grounds (see Fig. 7.7). There are isolated reports of pituitary sarcoid arising within an adenoma (Rubin et al., 2001) and associated with a Rathke’s cleft cyst (Cannavo et al., 1997); whether or not these are related or coincidental is unclear.

It should be stressed that the majority of patients with sarcoid who present with hypopituitarism have involvement of the hypothalamus or the pituitary stalk rather than the anterior pituitary itself (see Fig. 7.8).

**ANCA-positive vasculitis**

ANCA-positive vasculitis is a granulomatous inflammatory disorder with features of arteriolar and venular perivasculitis which affects the lungs, skin, eyes, and kidneys. Neurological involvement arises as a hypertrophic pachymeningitis, as an isolated inflammatory mass, or as a central or peripheral manifestation of inflammatory perivasculitis (Seror et al., 2006).

In one series 3 out of 6 cases had pituitary involvement, the majority of cases presenting with diabetes insipidus and not with a mass lesion. A recent review
of the literature cited 22 cases published (Yong et al., 2008), of which 80% showed pituitary enlargement, although this was sufficient to cause visual impairment in only 1 case. The pituitary stalk was thickened and the high magnetic resonance signal usually seen on T1-weighted images was absent in all those with diabetes insipidus. Another case was fulminating and treatment was not associated with visual improvement (McIntyre and Perros, 2007).

LYMPHOCYTIC INFILTRATION

Idiopathic optic chiasmitis

This uncommon condition may be diagnosed when other infective and inflammatory causes discussed above have been ruled out. A recent series from Indiana (Kawasaki and Purvin, 2009) showed 20 patients, 60% of whom were female; 40% presented with monocular visual loss but with evidence of chiasmal field defects, the remainder showing bitemporal hemianopia. Pain was not noted in this series.

Magnetic resonance imaging (MRI) revealed chiasmal swelling with or without enhancement in 12 of 15 cases (Fig. 7.9), and white-matter lesions were seen elsewhere in 6 of 15 cases. In follow-up, 6 of 15 had developed multiple sclerosis, including 3 in whom imaging of the brain initially was considered to be normal. Visual outcome following treatment was a normal acuity in 3 of 17, the remainder having improved to a degree with residual visual impairment, which was modest in the majority of cases.

It seems to this author that idiopathic chiasmitis is not really a separate disease entity, merely the description of a clinical syndrome that appears idiopathic at outset but that may evolve into a more widespread inflammatory neurological disorder over time.

An optic chiasmitis has also occasionally been seen in other systemic inflammatory diseases, for example systemic lupus erythematosus (Frohman et al., 2001).

Multiple sclerosis

The chiasm may be involved in multiple sclerosis; early reports (Traquair, 1925; Sacks and Melen, 1975; Spector et al., 1980; Beck et al., 1983; Newman et al., 1991) highlighted the clinical syndrome of a subacute unilateral or bilateral visual loss typical of an optic neuritis in which chiasmal-type field defects were seen. MRI shows swelling of the chiasm and often enhancement.

I have seen a patient who presented with a bilateral Uhthoff phenomenon with normal vision in between; the field defects were seen to enlarge with exercise. The patient went on to develop a slowly progressive form of multiple sclerosis with visual loss, ataxia, and spasticity.

Lymphocytic hypophysitis

This uncommon condition is more common in women (by a factor of 5:1) and often occurs in the late stages of pregnancy or the early postpartum period (Tubridy et al., 2001; Kidd et al., 2003; de Bellis et al., 2008). Patients present with headache, then signs of an expanding sellar lesion

![Fig. 7.9. T1-weighted (A) sagittal magnetic resonance image (MRI) showing enlargement of the body of the chiasm in idiopathic chiasmitis; (B) coronal MRI showing considerable enhancement of the same lesion. (Courtesy of Drs Aki Kawasaki and Valerie Purvin.)](image)
with visual impairment and endocrine hypofunction (Fig. 7.10). Occasionally hyperprolactinemia is seen. The adenohypophysis is affected more often than the neurohypophysis, although isolated diabetes insipidus may be seen. Many have other organ-specific autoimmune diseases. Hypopituitarism arises as a consequence of immune-mediated attack on the pituitary cells, rather than simply a compressive effect. MRI shows a symmetrical intensely enhancing tissue mass within the pituitary and there may be a dural tail (Sato et al., 1998). The pathology of the lesion is one of intense infiltration by lymphocytes and plasma cells, often with lymphoid follicles, associated with evidence for necrosis of adjacent pituitary tissue. Antipituitary antibodies may be detected (Bensing et al., 2007), although the assays appear not yet to have clinical utility (de Bellis et al., 2008).

Treatment is often unnecessary if a hypophysectomy has been undertaken, and follow-up suggests a low risk of recurrence (presumably because all pituitary tissue has been removed). Those who are diagnosed clinically by supposition, or following pituitary biopsy, appear to respond well to treatment with corticosteroids. The role of immunosuppression is not known; there are case reports of a response to radiotherapy in patients who fail to improve with steroids (Selch et al., 2003).

**Histocytic infiltration**

**Xanthomatous hypophysitis**

This is a pathological description for another inflammatory disorder of the pituitary in which cystic enlargement of the gland is seen, containing yellow fluid. The pathological features are of a lymphocytic and histiocytic infiltration of the gland. It is likely to be an incomplete form of the condition known as Erdheim-Chester disease, in which a histiocytic infiltration of the bones, lungs, and numerous other tissues may arise (Egan et al., 1999). Neurological complications occur most commonly due to involvement of the sellar region and orbit, but other areas may also be involved (Allen et al., 2004). This condition responds poorly to steroids and immune suppression, and the fatality rate when the lungs and kidneys are involved is said to be around 50% (Mills et al. 2008). Interferon-alpha may be helpful (Haroche et al., 2006).

Rosai-Dorfman disease is a rare histiocytic disorder of children and adults which presents with massive lymphadenopathy and infiltration of the nasopharynx, respiratory pathways, endocrine glands, bones, and skin. It may also involve the central nervous system (Kidd et al., 2006) in 4% of cases, in which a meningoeal-based inflammatory mass or multiple masses may arise which simulate meningiomas on imaging. Orbital and cavernous sinus masses may arise, and 3 cases of pituitary involvement by mass lesions of the parasellar region have been published (Kelly et al., 1999; Woodcock et al., 1999; Kidd et al., 2006) (Fig. 7.11).

**Mucoceles of the paranasal air sinuses**

One report deals with a chiasmal visual field defect in a patient with a mucocele of the sphenoid sinus (Goodwin and Glaser, 1987).

**Infections**

**Tuberculosis (TB)**

It is well known that TB may infiltrate the sellar region. Pituitary hypofunction and visual field defects may arise as a result of the development of tuberculoma within the pituitary, giving rise to a mass lesion with chiasmal compression (Domínguez et al., 2002) (Fig. 7.12) or to optochiasmal arachnoiditis, in which a basal tuberculous meningitis encroaches upon that region and induces visual loss, frequently in association with diabetes insipidus (Akhadder et al., 2001).

Patients may present with or without evidence for TB elsewhere and may present subacutely or as an emergency (Sharma et al., 2003). Tuberculoma mass
lesions are indistinguishable from other pituitary mass lesions (Akhadder et al., 2001), although it has been suggested that thickening of the pituitary stalk is more common in TB.

In optochiasmatic arachnoiditis imaging does not show a mass lesion but perichiasmal enhancement (Silverman et al., 1995). The visual prognosis is often good (Hughes et al., 2008), although not always (Sharma et al., 2003).

Optochiasmatic arachnoiditis may occur in other infective meningeal processes such as syphilis, Pseudomonas, staphylococcus, and streptococcus infections.

**Pituitary Abscess**

One series noted that patients with pituitary abscesses present with headache and visual loss mimicking a pituitary mass rather than signs of fever or meningitis; most cases were only diagnosed at surgery when a pus-filled mass was opened (Vates et al., 2001). Treatment with antibiotics after surgery was helpful but the mortality rate was 8% (Vates et al., 2001). Of 25 cases reported within the previous 5 years around half grew no organisms, whilst the majority of the remainder were pyogenic organisms, fungi, or parasites (Dalan and Leow, 2008).

**Cryptococcus**

Optic neuropathy is common in cryptococcal meningitis; a pathological study showed that the nerves and chiasm were damaged as a result of direct infiltration by the organism from the adjacent meninges (Cohen and Glasgow, 1993). There was no evidence for an associated vasculitis.

The prognosis for visual recovery even with prompt treatment is very poor.

**Cysticercosis**

One series of 23 patients from the USA showed 4 cases with chiasmal involvement; in 2 cases there was compression from the third ventricle on to the chiasm (see below) and in the other 2 cases there were intrasellar cysts causing compression from below (Chang and Keane, 2001).

**Viruses**

Isolated chiasmitis related to viral infections seems rare, but has been reported with varicella-zoster virus and Epstein–Barr virus (Purvin et al., 1988; Greven et al., 2001). It occurs more commonly alongside a severe syndrome in which the uveal tract, retina, and optic nerve and chiasm may all simultaneously be affected (Brazis and Miller, 2005).
Tumors and cysts

PITUITARY ADENOMA

These constitute 10% of all intracranial tumors; the prevalence increases with age. Adenomas arise from tissues within the anterior pituitary; functioning or secreting adenomas secrete prolactin, somatotrophin, adrenocorticotropic hormone, and thyroid-stimulating hormone. Most are microadenomas (measuring less than 10 mm in diameter), of which the majority will present with the effects of hormone hypersecretion and other incidental findings. Nonsecreting adenomas are more likely to be large; they account for only 10% of cases and present with visual loss and headache (Wilson, 1992) (Fig. 7.13). Some 10–20% of pituitary adenomas are identified incidentally on MRI brain scans (Aron and Howlett, 2000).

Prolactinomas account for 40% of pituitary tumors and present with galactorrhea and amenorrhea; the majority (90%) are microadenomas (Schlechte, 2003), but macroadenomas can enlarge appreciably in pregnancy (Kupersmith et al., 1994). The majority are treated successfully with dopamine agonists, and surgery even for large tumors is not required (Schlechte, 2003).

Fig. 7.13. Pituitary adenoma: (A) T1-weighted coronal magnetic resonance image (MRI) showing a large chromophobe adenoma with cystic change causing chiasmal compression. (B) T1-weighted sagittal MRI showing a large, predominantly cystic adenoma. (C) Thyroid-stimulating hormone-producing adenoma. (D) Pituitary apoplexy: chromophobe adenoma presenting with headache, visual loss, and a complete right-sided ophthalmoplegia.
It should be remembered that nonfunctioning macroadenomas may induce a rise in prolactin levels by the mechanism of compression on the pituitary stalk, leading to a loss of dopaminergic inhibition, and so to an increase in the prolactin level.

Corticotrope- and somatotrope-secreting tumors produce Cushing’s disease and acromegaly/giantism respectively. The former are more likely to present with endocrine abnormalities but growth hormone-producing tumors may enlarge unnoticed until visual symptoms arise (Rivoal et al., 2000). The majority of cases are due to pituitary tumors, but adrenocorticotrophic hormone and growth hormone may be produced ectopically by lung carcinomas or by carcinoid tumors respectively. Cushing’s disease requires surgery and, since there is a high rate of tumor recurrence, postoperative radiotherapy. Treatment of growth hormone-producing tumors is with somatostatin analogs.

Thyroid-secreting hormone-secreting tumors account for only 3% of cases and present either with thyrotoxicosis or with visual loss (25%) (Brucker-Davis et al., 1999). They are usually macroadenomas (see Fig. 7.13), and so treatment is surgical, although some respond to somatostatin analogs since their receptors are present.

A headache disorder is a prominent feature of some 34% of cases (Abe et al., 1998), and is more common in patients with hyperprolactinemia and acromegaly (Levy et al., 2002). In one recent series 76% had features of migraine headache, 27% primary stabbing headache, and 9% had features of cluster headache or short-lasting, unilateral, neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) (Levy et al., 2002). In one recent series 76% had features of migraine headache, 27% primary stabbing headache, and 9% had features of cluster headache or short-lasting, unilateral, neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) (Levy et al., 2005). The latter were more common in acromegaly and when the tumor involved the cavernous sinus. The pathogenesis of the headache disorder remains poorly understood; early assumptions that it reflected dural stretching or cavernous sinus involvement have been refuted (Levy et al., 2002). It was noted that 49% had a family history of migraine, implying that they were already predisposed to a primary headache disorder (Levy et al., 2005). Only half of the patients noted an improvement in headache with treatment, and use of cabergoline worsened the headache in 20% of those with prolactinoma (Levy et al., 2005).

**Treatment**

The risk of recurrence following transsphenoidal surgery in nonsecreting tumors is less than 10%, and higher following transcranial surgery (Couldwell, 2004).

With the exception of corticotrope adenomas, radiotherapy is usually withheld until there is evidence of tumor recurrence; hypopituitarism inevitably occurs, but late radiation-associated optic neuropathy is uncommon. The risk of oncogenesis leading to glioma, meningioma, or sarcoma is considered to be exceedingly rare (Jones, 1991).

**Visual prognosis following surgery**

This clearly depends on the size of the lesion, the duration of compression, patient age, and visual acuity at the time of surgery. In general an appreciable improvement occurs postoperatively, often within 24 hours (Powell, 1995). A slower improvement after 4 months extending to 3 years is less likely to proffer significant reduction in visual impairments. In prolactinoma visual improvement also starts quickly following initiation of dopamine agonist therapy; following radiotherapy recovery is more gradual.

**Craniopharyngioma**

These are benign lesions that nonetheless can cause prominent and irreversible pituitary and hypothalamic dysfunction. They arise from squamous epithelium at the junction of the infundibular stem and the anterior pituitary, and are thought to be remnants of Rathke’s pouch. There is a bimodal incidence, in childhood and again in middle to late life, with clinical features of a sellar or hypothalamic lesion. They may be situated above or below the chiasm, and rarely within it (Brodsky et al., 1988). The lesions are cystic (Fig. 7.14) and often contain viscous, oily (“engine oil”) fluid. Cholesterol clefts, calcium, and keratin may also be present.

In children these lesions present with hypothalamic dysfunction and hydrocephalus, in adults with features of chiasmal dysfunction, optic nerve or tract signs, and ophthalmoparesis.

The MR features vary with the composition of the lesion (Eldevik et al., 1996).

Surgery aims to provide debulking rather than removal, since surgery is often complicated by the development of hypothalamic dysfunction. Postoperative radiotherapy increases 10-year survival and may lead to prolonged remission, although recurrence is common (Manaka et al., 1985). Use of more modern radiotherapy techniques appears to improve survival further (Kobayashi et al., 2005).

The visual prognosis following treatment depends, as before, on the presenting acuity and the nature of the lesion, and the presence of disc atrophy or papilledema (McFadzean, 1989).

**Rathke’s cleft cyst**

These benign cysts form in embryogenesis if the lumen of Rathke’s pouch does not close, giving rise to a cyst situated between the anterior and intermediate lobes of the pituitary. Most are asymptomatic and do not...
enlarge. Patients with symptoms are predominantly female and present in middle life with visual loss due to chiasmal compression, headache, and hypopituitarism (Voelker et al., 1991). They are difficult to diagnose with certainty on MRI as there are usually features of craniopharyngioma or pituitary adenoma and the diagnosis is usually made at surgery. Treatment is with endoscopic transsphenoidal drainage and marsupialization. Visual improvement occurs postoperatively in 70% of cases (El Mahdy and Powell, 1998); those with poor preoperative acuities do less well.

**EPIDERMOID CYSTS**

When these cysts arise from the suprasellar region they present in the same way as a pituitary adenoma or craniopharyngioma, with visual loss and hypothalamic dysfunction (Mtanda et al., 1986). It is rarely possible to remove them in their entirety owing to their propensity to be adherent to the surrounding tissues, and recurrence therefore is common, although the rate of growth of these lesions is exceedingly slow. They are lined by stratified squamous epithelium; enlargement occurs due to progressive exfoliation from the epithelium of keratinous material, which is laid down in a lamellar pattern.

**DERMOID CYSTS**

These are also rare and similar to epidermoids except that the cyst is lined by pilosebaceous structures leading to the formation of hair. Whilst epidermoids are often laterally placed, dermoids tend to arise in midline structures.

Rupture of these cysts is associated with a chemical meningitis and seizures; those in the suprasellar region may therefore present with an apoplectic disorder similar to pituitary adenomas.

**RARE CHORISTOMAS IN THE SELlar REGION**

Occasionally cystic lesions also arise in the optic nerve, chiasm, or tract. The lesions are found to be composed of muscle and adipose tissue (Zimmerman et al., 1983).

**SUPRASELLAR ARACHNOID CYSTS**

These arise relatively frequently and present with signs of compression of the underlying structures, and occasionally upward compression leading to hypothalamic dysfunction and hydrocephalus. It is difficult with imaging to differentiate these from other cystic lesions such as Rathke’s cleft cyst (Rao et al., 2008), and the diagnosis is often only made at surgery; treatment is with incision and fenestration of the cyst with a low risk of recurrence.
CHORDOMA

During development of the axial skeleton the notochord is compressed and sequestration occurs but notochordal remnants may persist into life. Most are found in the sacrococcygeal region but some 25% arise in the base of the skull (Stippler et al., 2009). They arise most commonly from the clivus, where they present with diplopia due to sixth-nerve paresis (Fig. 7.15), but extension upwards may lead to optic tract or chiasmal involvement and hypothalamic dysfunction. Treatment is with surgery, and again recent advances in endoscopic techniques in skull base surgery have led to a higher chance of operative removal and a lower operative visual morbidity (Stippler et al., 2009). The prognosis depends on the histological appearances, but in general these are slow-growing lesions. There is evidence that postoperative radiotherapy is helpful, although high doses (70 Gy) are required (Debus et al., 2000). Newer techniques such as spot-scanning proton radiation therapy appear to be even better (Ares et al., 2009).

GERMINOMA

These uncommon tumors of early adult life arise most commonly in the pineal region, but may also be seen in the suprasellar region, where they may arise from the floor of the third ventricle or from the chiasm itself (Tageuchi et al., 1978). Germinomas are locally invasive and may metastasize (Nakajima et al., 2001).

Those in the sellar region present with visual failure and hypothalamic dysfunction, in particular, diabetes insipidus (Tageuchi et al., 1978). Since germinomas are highly radiation-sensitive a biopsy is undertaken rather than debulking, and a response to radiotherapy results in a good prognosis and survival rate (Isayama et al., 1980).

OTHER GERM CELL TUMORS

Teratoma may arise in the sellar region; some are benign and others are locally invasive. Imaging may show evidence of high-density tissues, for example bone and cartilage alongside adipose tissue. The prognosis is good in benign lesions that are removed in their entirety; others may respond to radiotherapy, but the survival rate diminishes with the mitotic activity of the tumor.

Embryonal cell carcinoma is a highly malignant tumor with a poor response to treatment and which may also present as a midline suprasellar lesion.

Yolk sac tumors also arise in the midline and often show combinations of the pathological features of choriocarcinoma and embryonal carcinoma. Alpha-fetoprotein level is usually raised and carcinoembryonic antigen and human chorionic gonadotropin (hCG) levels are normal, and treatment response may be monitored with this assay. Despite surgery, radiotherapy, and chemotherapy the prognosis remains poor.

Choriocarcinoma is very rare as an isolated intracranial lesion. It secretes beta-hCG and the prognosis, despite treatment, remains grave.

There is some evidence that these tumors are associated with chromosomal abnormalities (Sato et al., 2009).

GLIOMA

Gliomas may involve the chiasm either by arising within the chiasm itself or by infiltration from the hypothalamus. They show similar histological features to those of unilateral optic nerve gliomas, and may also arise in neurofibromatosis type 1. They may present at any age but are more common in children. The onset of visual loss is usually insidious, and may even be discovered at a routine check-up. The discs are usually pale at presentation, and the field defects bitemporal (Dutton, 1994). Headache may also be a feature.

Imaging shows enlargement of the chiasm, sometimes with cysts and rarely with calcification (Hoyt et al., 1987).

Treatment is more often hazardous than helpful; the long-term visual prognosis is overall fair (Cappelli et al., 1998), and attempted biopsy or removal is likely to damage visual and hypothalamic function permanently. Radiotherapy may be associated with late
adverse effects such as necrosis, new tumors, and the development of moyamoya disease (Lee, 2007). However, some are exophytic and therefore surgically accessible.

Chemotherapy with vincristine, temozolamide, and platinum agents has been shown to be helpful in children (Laithier et al., 2003).

Malignant optic nerve gliomas may also affect the chiasm (Fig. 7.16). These present with a much more rapidly evolving clinical syndrome and respond poorly to treatment (Rudd et al., 1985). Metastasis to other parts of the nervous system may occur (Murphy et al., 2003). The pathology of the lesion is very different to that of the benign chiasmal glioma, and is more similar to glioblastoma multiforme (Hamilton et al., 1973).

MENINGIOMA

These tumors are twice as common in women as in men and arise more frequently in the second half of life. They tend to be benign and slow-growing but some may infiltrate the underlying tissues. The chiasm may be affected by those that arise from the sphenoid wing, the clivus, and the olfactory groove (Cockerham et al., 2005), and primary optic nerve sheath meningiomas may grow backwards to involve the chiasm (Dutton, 1992; Miller, 2008). Tuberculum sellae or parasellar meningiomas are uncommon (Fig. 7.17), accounting for some 3% of intracranial meningiomas. They may arise at any point in relation to the chiasm, but are most common in the retrochiasmatic region. They present with progressive painless loss of vision leading to the same visual field defects noted above, although rarely, when the chiasm is prefixed, the tumor may compress the optic nerves medially, leading to compression by the internal carotid arteries laterally. When the chiasm is prefixed a tractopathy is more likely.

Treatment is with surgery, and not all can be removed in their entirety; as such it may be advisable simply to watch the tumor growth for a time, but worsening vision can result. Nonetheless, surgery carries with it significant morbidity, although this is diminishing with new endoscopic endonasal techniques (Gardner et al., 2008). An improvement in visual acuity of modest proportions occurs in around two-thirds (de Divitiis et al., 2008).

In a series from Johns Hopkins (Chicani and Miller, 2003) half of the patients followed had stable vision postoperatively whilst the others deteriorated. This was associated with radiographic evidence of tumor recurrence in 39% after a mean of 10.7 years.

METASTATIC TUMORS

Metastasis to the sellar region has been reported with most carcinomas and also lymphoma and leukaemia.

VASCULAR DISORDERS

CAVERNOMA

A recent series has found 40 cases published, in which the majority present acutely with visual loss due to bleeding. A total of 87% of those who underwent surgery showed an improvement in visual acuity postoperatively (Crocker et al., 2008).

Fig. 7.16. Malignant glioma of the chiasm: (A) coronal T1-weighted scan showing enlargement and enhancement of the chiasm; (B) axial scan of the same lesion.
ARTERIOVENOUS MALFORMATIONS

Arteriovenous malformations may rarely arise within the chiasm and provoke a variety of syndromes, including transient bilateral visual obscurations, and field defects due to bleeding (Sibony et al., 1982).

ISCHEMIA

The extensive blood supply to the chiasm has been noted above and for this reason ischemic infarction of the chiasm is exceedingly rare. Ahmadi and colleagues (1984) reported a patient with very severe occlusive internal carotid artery disease in whom the chiasmal syndrome was considered to be due to ischaemia.

Compression from hydrocephalus

A host of visual field defects has been seen in hydrocephalus, due to compression of the optic nerve, chiasm, and tract (Osher et al., 1978; Humphrey et al.
When the third ventricle presses downwards directly and symmetrically on to the chiasm a bitemporal hemianopia or upper quadrantinopia would be expected (Fig. 7.18).

**Traumatic chiasmal syndrome**

This usually develops in victims of motor accidents and is associated with skull and facial fractures. The field defect is a complete bitemporal hemianopia. In one series the prevalence of associated diabetes insipidus was 37% (Hassan et al., 2002).

**REFERENCES**


