Sir,

Compressive optic neuropathy in fungal hypertrophic cranial pachymeningitis

We report a rare case of compressive optic neuropathy secondary to hypertrophic cranial pachymeningitis. Fungal aetiology is rarely reported in the literature and can be life threatening.

Case report

A 73-year-old male presented with a 3-week history of left-sided visual loss, associated with headache, scalp tenderness, shoulder pain, and general fatigue. Of note in his medical history was type II diabetes mellitus and pulmonary asbestosis. There was no history of immunocompromise. On examination, his visual acuity was R 6/18 and L NPL, with a left afferent pupillary defect and left optic disc pallor, but a normal appearance of the right optic disc. The vision in his right eye was known to be reduced secondary to diabetic macular ischaemia. Neurological examination was otherwise unremarkable, as was his right visual field on Goldmann perimetry.

Initial blood tests revealed a white cell count of 12.7, ESR 61, and CRP 48. Temporal arteritis was initially suspected, but was ruled out with a negative temporal artery biopsy and lack of response to corticosteroid treatment.

MRI scanning showed an opacified left frontal sinus and large meningeal deposits with abnormal dural thickening and enhancement with gadolinium over both the sphenoid wings. There was compression of the left optic nerve at the anterior clinoid (see Figure 1). Lumbar puncture showed raised protein (567 mg/dl) without inflammatory cells. His left frontal sinus was drained, and culture grew *Aspergillus flavus*. A meningeal biopsy established the diagnosis as hypertrophic cranial pachymeningitis (HCP) caused by this organism. It was felt that the origin of his systemic fungal infection was pulmonary, as asbestosis is known to predispose to *Aspergillus* infection. He was commenced on systemic antifungal treatment, but his general medical condition rapidly deteriorated. He died 3 months later, secondary to respiratory failure and secondary overwhelming bacterial septicaemia.

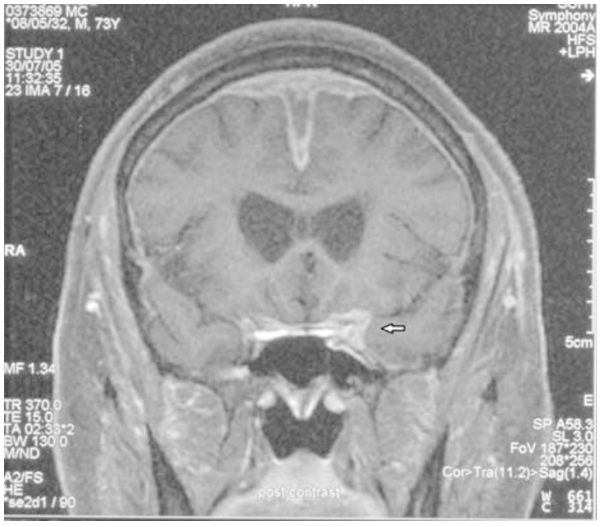

Figure 1 T1-weighted post-contrast MR image showing extensive thickening and enhancing dura over the left sphenoid wing (arrowed).

Comment

HCP is a rare disorder characterized by diffuse or focal thickening and inflammation of the dura, and leads to all three meningeal layers becoming fused by dense fibrotic membranes.[1] Although several inflammatory and infectious causal agents have been identified, many cases are idiopathic. Patients usually present with multiple progressive cranial neuropathies, and the optic nerves and orbital apex are occasionally involved.[2] Blindness in HCP may be caused by constriction of the optic nerve by inflammatory thickening in the optic canal[3] or direct inflammatory cell invasion of the optic nerve.[4]

CSF analysis in HCP varies according to the aetiology, and may be normal even in infective cases.[5] Gadolinium-enhanced MR imaging is essential for identifying meningeal inflammation and also for locating suitable biopsy sites.

Optic nerve involvement in HCP usually confers a poor visual prognosis. Infective fungal cases are not well reported in the literature, but there is a significant mortality risk, and early diagnosis and treatment may be life saving. One report has shown some visual improvement and thinning of gadolinium-enhanced lesions on MRI, after systemic antifungal treatment.[6] In non-infectious cases, some improvement in visual acuity has been shown with high dose corticosteroid treatment.[2,7]

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Sir,

Evolution and management of diabetic tractional papillopathy: an optical coherence tomographic study

Vitreopapillary traction (VPT) has been reported in both young and old diabetic patients, with background/proliferative retinopathy.[1–4] Vision may be adversely affected; recovery follows spontaneous/surgical relief of VPT.[3,4] However, the need for vitrectomy has been questioned, as VPT has not been conclusively shown to be the sole cause of visual decline.[5] We demonstrate VPT as the sole cause of visual loss in a diabetic patient, further evidenced by full visual recovery after vitrectomy.

Case report

A 40-year-old diabetic hypertensive man presented with recent-onset blurred vision OD; best-corrected visual acuity (BCVA) was 6/12 OD and 6/6 OS. On examination, anterior segment was unremarkable OU. Fundus OD showed retinal haemorrhages, macular oedema, and a large neovascular frond on the optic disc, probably representing enlarged telangiectatic vessels, similar to those seen in diabetic papillopathy (Figure 1a). Left fundus showed mild non-proliferative diabetic retinopathy. Fluorescein angiography (FA) revealed no leakage from the disc vessels, and no capillary dropout OD (Figure 1b and c). Optical coherence tomography (OCT) revealed VPT with adjacent vitreomacular traction (VMT), causing nasal macular thickening (Figure 1d). After 3 months, BCVA dropped to 6/36, with pre-retinal haemorrhages, disc oedema, and elevated disc vessels OD (Figure 1e). OCT demonstrated increased VPT, but reduced macular oedema owing to release of VMT (Figure 1f). A relative afferent pupillary defect was noted. Automated perimetry showed generalized depression, and reduced foveal thresholds (22 dB) OD. FA remained essentially unchanged. With patient's informed consent and approval of the Institutional Review Board, pars plana vitrectomy was performed, and VPT was removed. Postoperatively, BCVA improved to 6/6 over a month. The disc vessels underwent fibrosis; disc oedema reduced substantially, clinically, and tomographically (Figure 1g and h). The pupillary response and foveal perimetric thresholds also normalized (38 dB); the generalized field depression persisted. The functional and anatomic recovery was sustained 12 months post-vitrectomy.

Comment

Kroll *et al*[4] proposed that VPT caused mechanical/vascular damage to the papillomacular bundle, with surgically reversible functional impairment. However, they obtained only modest improvement in vision and visually evoked potential, probably due to surgical delay. McLeod[5] highlighted the disparity between their visual and electrophysiological outcome; and disputed VPT as the sole cause for visual loss, without concrete documentation of vitreomacular status. Karatas *et al*[1] used OCT to demonstrate VPT; but proposed VPT as a cause of macular oedema, rather than a direct mediator of visual deterioration. Further, their series was retrospective; and did not report any surgical results to substantiate their hypothesis. In contrast, we demonstrate progressive VPT—without contribution from VMT/macular oedema—as the sole cause of visual decline. Though spontaneous posterior vitreous detachment (PVD) is reported to improve vision;[3] in view of the patient's age, relentless visual deterioration over 3 months, and reported consequences of delayed surgery, further observation appeared less appealing than a simple surgery. McLeod[5] cautioned that surgery may itself damage the nerve fibres; we avoided this complication by gentle and gradual PVD induction. VPT should be remembered as a rare cause of unexplained visual deterioration in diabetic patients, especially because it can be reversed by early vitrectomy.