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Sir,

Chiasmal cavernous angioma. a rare cause of progressive visual loss

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Intracranial cavernous angiomas are not uncommon.[1,2] Only 5% are supratentorial, most of which are intraparenchymal and are located in the cortical and subcortical ganglia. Infratentorial angiomas may be located within the brainstem, cerebellum, third or fourth ventricles or the posterior fossa.

Angiomas can very rarely develop within the substance of the optic nerve, optic tract and optic chiasm, where they produce acute or progressive loss of vision in one or both eyes. We report a case of chiasmal angioma presenting with progressive visual loss.

Case report

A 42-year-old man presented with a history of blurred vision in the right eye for 4 days. He had an unaided visual acuity of 6/4 in both eyes. Pupillary reactions and fundus examination were normal. He returned a week later with a history of decrease in colour vision in his right eye. Again, apart from mild colour desaturation, there was nil of note. In particular, he had no afferent pupillary defect or optic disc swelling. There was no pain on ocular movements. As he demonstrated Uhtoff's phenomenon, he was thought to have mild optic neuritis.

Visual fields demonstrated central loss in the right eye. Electrodiagnostic tests demonstrated a complete loss of pattern visual evoked potential (VEP) with preserved flash VEP response from the right eye. His right visual acuity subsequently dropped to 1/60 and he developed a right afferent pupillary defect and optic disc pallor. Magnetic resonance imaging (MRI) scan demonstrated a lesion at the right optic chiasm, which was thought to be a meningioma or a craniopharyngioma (Figure 1). Magnetic resonance angiography (MRA) was reported to be normal. The patient was referred to the neurosurgeons. He

underwent right pterional craniotomy and exploration of the right optic nerve.

There was a vascular lesion within the optic nerve and encroaching onto the chiasm and opposite optic nerve (Figure 2). The tumour was completely resected with preservation of optic nerve on the right side. Postoperatively the patient developed diabetes insipidus for which he is being treated medically by the endocrinologists. He retained normal vision on the opposite side with an improvement in right eye vision from perception of light to counting fingers. The tumour was histologically proven to be a cavernous angioma (Figure 3).

Comment

Cavernous angiomas of the intracranial optic pathways are extremely rare with only a few reported cases in the literature.[2–6] They are quite common in the orbit with intraconal lesions outnumbering extraconal ones.[1]

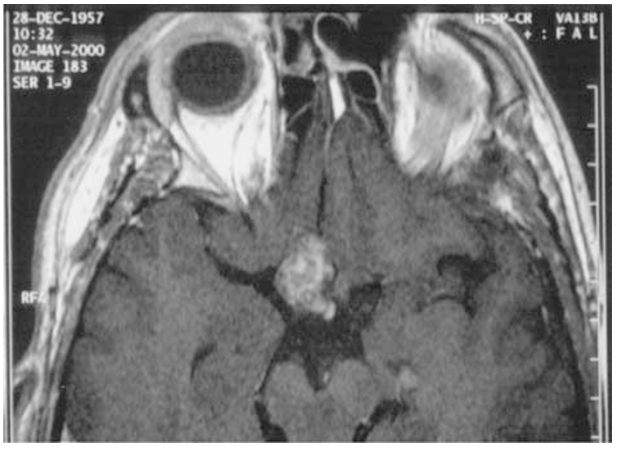

Figure 1 MRI scan showing the lesion in the region of the optic chiasm.

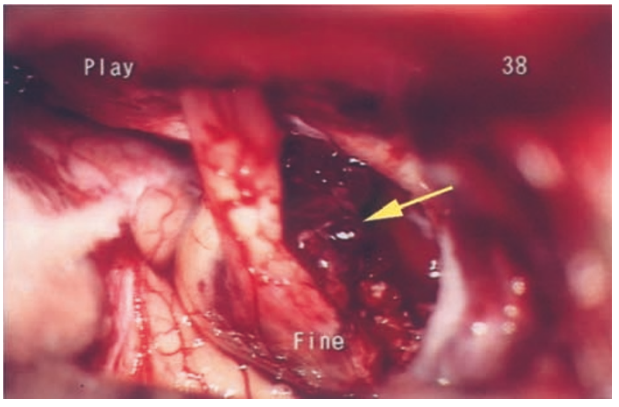

Figure 2 Peroperative photograph with arrow showing a fleshy, vascular lesion within the optic nerve.

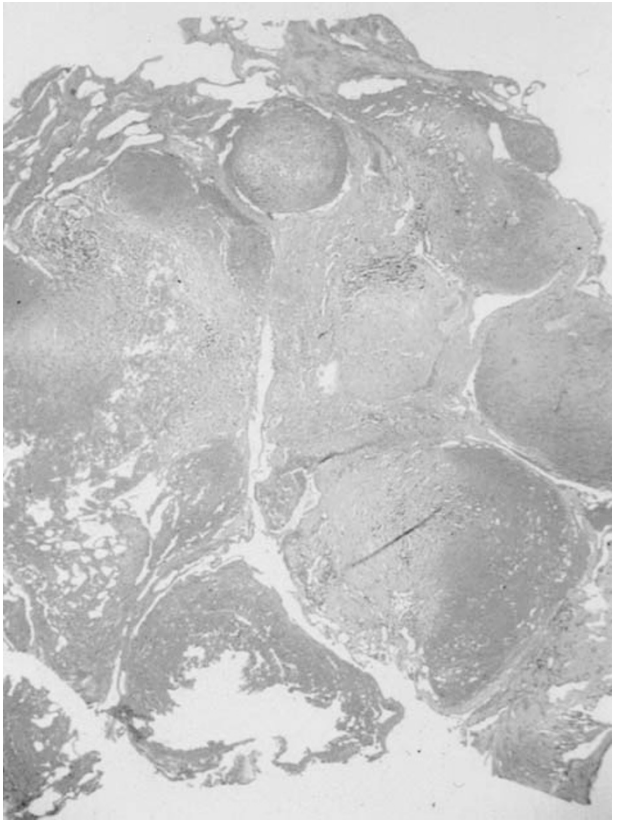

Figure 3 Microscopic section shows fragments of densely fibrotic nerve, fibrous tissue and blood clot. There is evidence of old haemorrhage. There are remnants of very abnormal blood vessel walls. The appearances are consistent with a cavernous angioma (×1, HE).

Rare cases have been reported within the optic canal. They may rarely present as a saccular 'grape-like' lesion of the optic disc and cause vitreous haemmorhage.

Various modes of presentation have been described with acute chiasmatic syndrome being the most common mode of presentation.[4] Patients have been reported to present with retroorbital pain, bitemporal headaches and field loss after subarachnoid haemorrhage and intracerebral haematoma.[2–4,6] Less commonly, cavernous angiomas of the optic nerve and chiasm present with slowly progressive visual loss similar to the present case.[5]

As in the present case, in many instances the diagnosis is not apparent until surgery is performed. MRI is a sensitive and specific modality for the diagnosis of cavernous angiomas, which must be suspected in the presence of an acute chiasmal syndrome. MRI in these cases shows an acute or subacute chiasmatic haemorrhage. A typical MRI pattern of cavernous haemangioma has been described with heterogenous alteration of foci of blood of

different ages, with a central focus of methemoglobin, a peripheral rim of hemosiderin, an adjacent focus of acute or subacute haemorrhage and minimal or no enhancement on gadolinium administration.[7] The patient in the present report had MRA on two occasions, both of which were reported to be normal. Angiography is typically silent with these lesions. This may be because the small caliber of feeding arteries, and their slow circulation, causes dilution of contrast medium, or extensive thrombosis or both.[1]

In other cases, MRI may suggest a meningioma or a venous anomaly as in the present case. Other possibilities include a haematoma, calcified glioma or a venous angioma.[1] Unlike similar lesions in the orbit, intracranial angiomas have no true capsule, although compression from an expanding lesion produces a gliotic pseudocapsule. Microscopically their walls are composed of collagen with tongues of fibrillary neuroglia which penetrate the lesion.[1]

Cavenous haemangiomas may have an autosomal dominant inheritance. Not all lesions require surgery. Those presenting with seizures and minimal or stable visual loss can be observed. Surgery is advocated for those with severe or progressive visual loss.

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Sir,

Central retinal vein occlusion associated with Sturge Weber syndrome

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Central retinal vein occlusion associated with Sturge Weber syndrome is rare, to our knowledge there are only two other reported cases in the literature.[1,2] We report such a case.

Case report

A 22-year-old male patient with left-sided Sturge Weber syndrome and well-controlled ipsilateral glaucoma, presented to the eye casualty department with a left central retinal vein occlusion (Figure 1). The visual acuity was 6/36 in the left eye and intraocular pressure 18 mmHg. The cornea was clear and there was no evidence of rubeosis iridis. A relative afferent pupilliary defect was noted prior to dilated fundoscopy, which revealed the typical features of a central retinal vein occlusion. Routine blood investigations, including a thrombophilia screen, were all normal, and antithrombin III and plasminogen levels were all within normal limits.

The patient suffering from Sturge Weber syndrome originally presented to the eye department at 9 years of age, with an elevated intraocular pressure of 34 mmHg in the left eye, the right was 20 mmHg. The visual acuity was 6/6 in each eye and the optic discs were healthy (Figure 2). The left corneal diameter was 2 mm greater than the right. A visual field test revealed a right homonymous hemianopia, which was attributed to Sturge Weber syndrome. He was known to be epileptic and controlled with sodium valproate and carbamazepine. There was no other significant medical history. A diagnosis of Sturge Weber

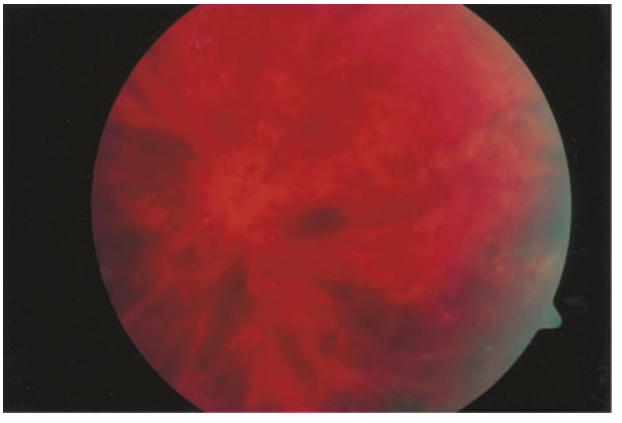

Figure 1 Central retinal vein occlusion in the left eye.