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Sir,

Retinal and orbital arteriovenous malformations

Arteriovenous malformation of the retina is a rare clinical entity of congenital origin, that of the orbit even rarer. We report a case in which both exist without any other neurological or systemic involvement.

Case report

A 7-year-old Caucasian girl was seen by ophthalmologists when a routine optometry visit revealed gross reduction of vision in the left eye. Subsequent examination confirmed decreased visual acuity; in addition there was ipsilateral deficit in colour vision and left-sided relative afferent pupillary defect. Fundoscopy of the left eye revealed greatly enlarged and tortuous blood vessels (Fig. 1), of similar colour, making the arteriovenous distinction nearly impossible. These vessels completely obscured the view of the optic disc. There was, however, no evidence of retinal exudation or haemorrhage. The right eye was normal and the child was otherwise healthy.

The patient underwent a CT scan, which revealed dilated vessels in the left orbit consistent with the presence of an arteriovenous malformation. An MRI scan of the brain (Fig. 2) excluded intracranial extension of the arteriovenous malformation, and an abdominal ultrasound scan ruled out any visceral involvement.

Observation of the child was our preferred option; she has now been followed-up for 3 years with no change in her clinical condition.

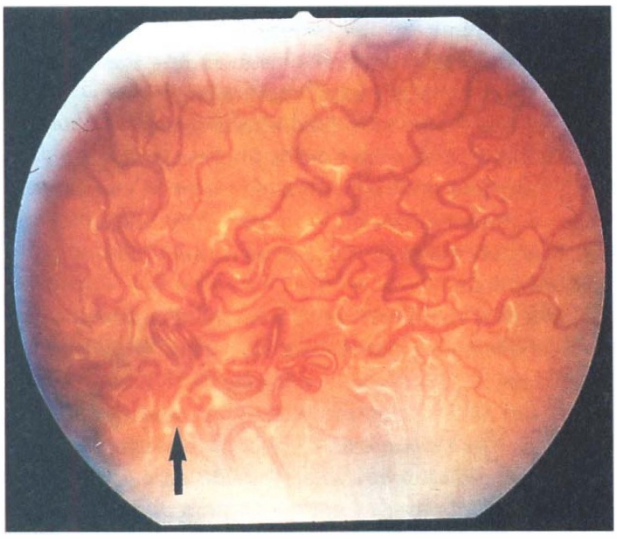

Fig. 1. *Left fundus showing enlarged and tortuous blood vessels, with no arteriovenous colour gradient. The optic disc is completely obscured (arrow at presumed site).*

Comment

Retinal arteriovenous malformation when associated with those present in the central nervous system, orbit, maxilla or mandible comprises a rare phacomatosis grouped in the syndrome of Wyburn-Mason,[1] as described by him in 1943. He was the first to recognise the relationship between the various components, although Bonnet, Dechaume and Blanc[2] described this rare congenital unilateral hemicephalic disorder even earlier, in 1937. Magnus[3] first described the retinal component, also known as racemose haemangioma, in 1874.

It appears that arteriovenous malformations of the retina, brain and face were reported infrequently until Wyburn-Mason investigated their association. In his original series he reported that one lesion may exist without the other, and this was also noted by Brown *et al.*[4] Although cases of retinal arteriovenous malformation occurred mainly with those in the midbrain in Wyburn-Mason's series, numerous other

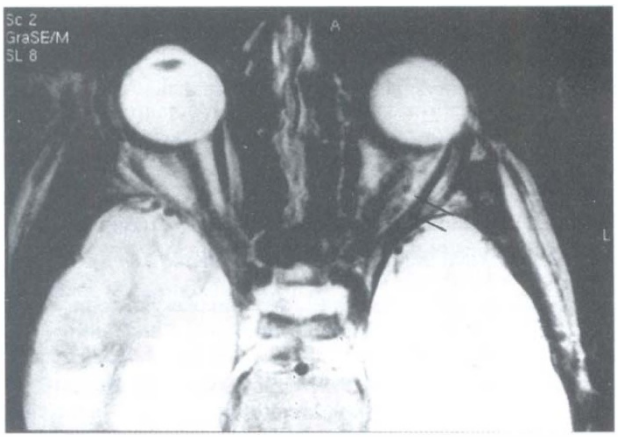

Fig. 2. *MRI scan showing dilated vessels (arrows) in the left orbit with no posterior extension through the optic foramen. The right orbit is normal.*

cases of ipsilateral cerebral hemispheric involvement have been reported, common sites including the basofrontal region, posterior fossa and meninges.

Archer *et al.*[5] studied retinal arteriovenous malformations *per se* with fundus fluorescein angiography and ophthalmodynamometry to elucidate structural abnormalities, and grouped them according to the nature and extent of the arteriovenous communications.

A review of the literature also reveals that retinal arteriovenous malformations are capable of growth,[6] haemorrhage,[5] sclerosis[7] and thrombosis.[8,9] These changes can affect vision profoundly by causing optic atrophy, as can a compressive effect of the lesion.

Orbital manifestations, when they do occur, include enlargement of the optic foramen and, occasionally, proptosis, which may or may not become pulsatile.

Dermatological lesions can occur in a minority of cases and take the form of ipsilateral skin naevi, in the distribution of the trigeminal nerve.

Conclusion

Our young patient has Wyburn-Mason syndrome by virtue of ipsilateral retinal and orbital arteriovenous malformations. To our knowledge, such an association without any lesion in the brain has not been reported previously.

Although retinal arteriovenous malformations can sometimes be confused with inherited retinal arteriolar tortuousity or even congenital retinal macrovessels, in our case, the typical age of presentation and the unilateral involvement suggest otherwise.

Intervention, when contemplated, may comprise embolisation (frequently used to treat central nervous system arteriovenous malformations), laser photocoagulation or proton beam radiation, the latter being described in maxillary and mandibular lesions.

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Sir,

Primary acute angle closure glaucoma associated with suprachoroidal fluid in three Chinese patients

Separation of the choroid from overlying sclera may occur with hypotony, inflammation, neoplasia, panretinal laser photocoagulation,[1] nanophthalmos and uveal effusion syndrome.[2] Clinically detectable choroidal detachments associated with profound reductions in intraocular pressure are described after trabeculectomy, bleb needling or even topical treatment.[3–5]

Acute primary angle closure glaucoma (APACG) demands prompt, aggressive lowering of raised intraocular pressure to break the attack and minimise optic nerve damage and ischaemic sequelae. We describe three patients with suprachoroidal fluid collections following such treatment. As part of a larger prospective study we examined all patients with APACG attending the Singapore National Eye Centre over 18 months. Ultrasound biomicroscopy (UBM)[6] examinations (Humphrey Instruments, San Leandro, CA) were performed within 24 h of presentation, before peripheral laser iridotomy but after medical treatment.

Case reports

All three patients presented with a history of periorbital pain and blurred vision of between 1 day's and 2 weeks' duration. They all had raised intraocular pressure with signs of APACG and closed anterior chamber angles on gonioscopy (Table 1). All were female. Treatment with intravenous and oral acetazolamide, mannitol, topical pilocarpine, timolol and steroids successfully broke the attacks.

UBM demonstrated suprachoroidal fluid collections in all three cases (Fig. 1), the maximum depths of which are shown in Table 1. Patient 1 had the largest collection, extending over the full 360° circumference of the globe and posteriorly beyond the equator, whereas those in patients 2 and 3 were shallower and did not reach to the equator. All uninvolved eyes had narrow but open angles and no abnormality on UBM. All three fluid collections resolved within 5 days.