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LETTERS TO THE JOURNAL

Sir,

A rare and unusual congenital arteriovenous communication of the retina

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Congenital arteriovenous communications or malformations are rare retinal vascular anomalies. We report an unusual juxtapapillary arteriovenous communication of the retina presenting as an incidental finding in an otherwise healthy individual.

Case report

A 41-year-old Caucasian male was referred with a retinal vascular lesion in one eye following a routine visit to his optometrist. He was asymptomatic with no past ocular history or family history of glaucoma. Five years back a benign lump had been removed from his tongue. His medical history was otherwise unremarkable.

Ocular examination revealed a best-corrected visual acuity of 6/5 in both eyes. The anterior segments and intraocular pressures were normal. Fundus biomicroscopy revealed an unusual juxtapapillary vascular lesion in the left eye (Figure 1a). The right fundus was normal.

A fundus fluorescein angiogram demonstrated anomalous vessels of arteriovenous nature. They were nasal to the optic disc and leaked as the angiogram progressed. No other abnormalities were detected (Figure 1b–d).

No vascular malformations were detected in the conjunctiva, mouth, or tongue. The histology of the lesion that had been removed from the patient's tongue was reviewed. This confirmed the diagnosis of a granular cell myoblastoma, ruling out the possibility of a second arteriovenous malformation.

Full blood count, ESR, blood glucose, and serum lipids were normal. Computerised tomography of the brain and orbits showed no abnormality. Mitochondrial DNA analysis for Leber's hereditary optic neuropathy (LHON 11 778, 3460, and 14 484) and sequence analysis of exons 1–3 of the Von Hippel–Lindau gene were negative.

The patient has been symptomatically and clinically stable for 1 year and remains under long-term review.

Comment

The patient presents with an unusual unilateral retinal vascular anomaly as an incidental clinical finding. The presentation, clinical and angiographic appearance is consistent with the diagnosis of a congenital arteriovenous communication group 1.

Congenital arteriovenous malformations or communications were classified into three groups on the basis of severity.[1,2] Group 1 consists of an anastamosis between a small arteriole and a small venule with the interposition of an abnormal capillary or arteriolar plexus. Group 2 demonstrates direct arteriovenous communications between a branch retinal artery and vein. Group 3 consists of diffuse marked dilatation of the vascular tree with many large calibre anastamosing channels.

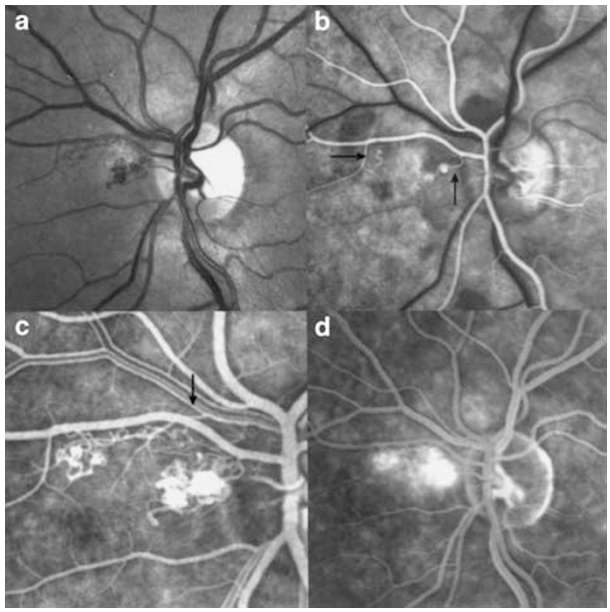

Figure 1 (a) Red free photograph of the left fundus showing the juxtapapillary arteriovenous vascular malformation. (b) Early arterial phase of the fundus fluorescein angiogram of the left eye showing the multiple arteriolar connections with the vascular malformation (arrows). (c) Early venous phase of the fundus fluorescein angiogram of the left eye showing the vascular pattern of the lesion with drainage into a retinal vein (arrow). (d) Late phase of the fundus fluorescein angiogram of the left eye revealing leakage.

Congenital arteriovenous malformations usually present on routine examination, with unilateral involvement in single or multiple sites of the same fundus.[1,2] They have a predilection for the papillomacular bundle and the superotemporal quadrant. They have rarely been reported nasal to the optic disc.[1,2] Capillary bed leakage within the area of arteriovenous malformation is uncommon but has been described.[1] The arteriovenous malformation in this case was located nasal to the disc and demonstrated leakage on angiography.

Ocular complications of arteriovenous malformations include intraretinal haemorrhage, and exudation, aneurysm formation, vascular occlusion, neovascular glaucoma and vitreous haemorrhage.[3] The association of retinal arteriovenous malformations with systemic arteriovenous malformations is well documented.[1] Sites of involvement include the central nervous system, orbit, eyelid, oro-naso-pharyngeal area, skin, lung, and spine.[1]

Group 1 arteriovenous malformations are usually isolated findings detected on routine examination, non progressive, and associated with good visual acuity.[1] Photocoagulation treatment may be indicated if exudative maculopathy develops.[3] The differential diagnosis of congenital arteriovenous malformations includes peripapillary telangiectasia, capillary and cavernous haemangiomas, and retinovascular disease.[1]

Peripapillary telangiectatic microangiopathy is well described in LHON.[4] The microangiopathy precedes the optic neuropathy, and the telangiectatic vessels do not demonstrate leakage on angiography.[5] In nearly all families, LHON is associated with one of three mitochondrial DNA (mtDNA) mutations, at base pairs 11 778, 3460, or 14 484.[6] Mitochondrial DNA analysis in this patient was negative.

Retinal capillary haemangiomas may be sporadic or occur as a part of Von Hippel–Lindau's disease.[7] Clinically, these often present with exudative features and show arteriovenous shunting.[8] These features were absent in this patient and genetic testing was negative.

Cavernous haemangioma is a rare unilateral congenital anomaly located between two retinal veins and presenting with the typical appearance of a cluster of grapes.[9] Arteriovenous malformations should also be distinguished from acquired optic disc venous collaterals secondary to retinal venous occlusion or optic disc meningioma.[3]

The clinical and fluorescein angiogram findings, normal CT scan of the brain and orbits, and negative genetic screening all point in favour of a congenital arteriovenous malformation. The atypical features in this case are the location of the arteriovenous malformation, which was nasal to the disc, and the leakage of dye on angiography.

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Sir,

A new cause for retinal haemorrhage and disc oedema in child abuse

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Systemic hypertension in very young children is uncommon. The conditions associated with acute or intermittent hypertension include renal vascular or parenchymal disease, drugs or poisons, central or autonomic nervous syndromes, thrombosis of the aorta, and chronic lung disease.[1] Acute ocular injuries from child abuse include retinal haemorrhages, periorbital haematoma, retinal detachment, lens subluxation, subconjunctival haemorrhage, and trauma to eyelids.[2] We report the case of a child with retinal haemorrhages and show how serial fundus photography helped us to look into causes of retinal haemorrhages in child abuse other than shaking injury.

Case report

A $3^{1/2}$ year-old boy was transferred to our paediatric unit from a district general hospital (DGH) following an