European Journal of Ophthalmology / Vol. 13 no. 3, 2003 / pp. 324-327

SHORT COMMUNICATION

Case report

Transient third nerve palsy in a young patient with intracranial arteriovenous malformation

G. WU, A. AGRAWAL, F.D. GHANCHI

Department of Ophthalmology, Bradford Royal Infirmary, Bradford - UK

PURPOSE. *To describe a patient with transient third nerve palsy as the possible presenting sign of intracranial arteriovenous malformation.*

METHOD. *Case report.*

RESULT. *A 24-year-old female presented to ophthalmic casualty with sudden onset binocular diplopia and was diagnosed to have right sided partial third nerve palsy. Within 30 hours the third nerve palsy had recovered completely. A MRI scan and subsequent carotid angiogram revealed a large, high flow, trans-cortical Spetzler-Martin grade 4 arteriovenous malformation. The feeder vessel of the AVM originated from the right middle cerebral artery. Superficial venous drainage was via the superficial middle cerebral vein to the right transverse sinus. The deep venous drainage was via thalamostriate veins into markedly dilated internal cerebral vein and vein of Galen (Great cerebral vein). Venous reflux was noted around the midbrain from the vein of Galen.*

CONCLUSIONS. *Transient third nerve palsy may rarely occur secondary to intracranial arteriovenous malformation. Ophthalmologists should consider neuroimaging in the investigations for transient cases of III nerve palsy in young patients. (Eur J Ophthalmol 2003; 13: 324-7)*

KEY WORDS. *Arteriovenous malformation, Third nerve palsy, Pupil*

Accepted: October 16, 2002

INTRODUCTION

Intracranial arteriovenous malformations (AVM) can produce signs and symptoms in the ocular motor and visual sensory system (1). We report a case of transient third nerve palsy as the possible presenting sign of AVM in a 24-year-old patient. This case highlights the importance of full evaluation of transient neurological deficit in young patients.

Case report

A 24-year-old caucasian female presented to the ophthalmic casualty with sudden onset of binocular diplopia and drooping of her right upper-lid for the preceding 4 hours. There was no history of headache or retro-orbital pain. Her past medical history was only remarkable for a few episodes of fainting over the preceding three months. She was not on any regular medications. On examination, her visual acuity was 6/6 unaided in both eyes. She had anisocoria, which was more marked in bright illumination. Pupils: OD 7mm, OS 3 mm (bright illumination) and OD 9mm, OS 8mm (dim illumination). The right pupil had sluggish reaction to light and accommodation. There was right upper-lid ptosis of 4 mm. She had right hypotropia in primary position with limitation of up-gaze. Other ocular movements were full. Fundus examination was normal. Rest of the neurological examination was normal. A clinical diagnosis of right partial third nerve palsy was made. She was admitted for neurological observations and neuro-imaging. Subsequently she developed a mild intermittent right-sided dull headache,

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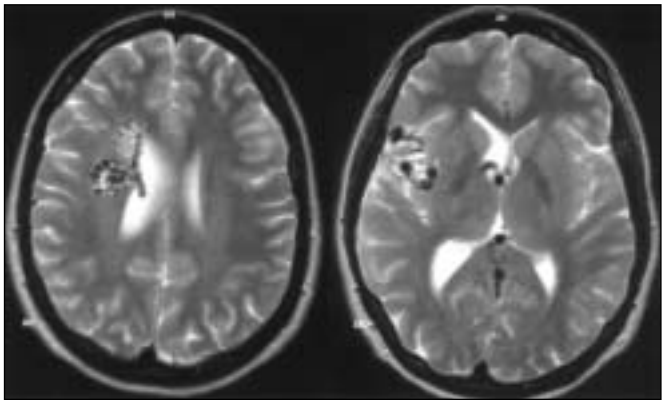

Fig. 1 - *MRI scan of the brain demonstrating the trans-cortical AVM. The epicentre of the lesion is in the deep white matter of the centrum semiovale of the right cerebral hemisphere. The lesion extends medially into a subependymal location in the right lateral ventricle with a nubbin extending into the ventricular lumen. Empty spaces in the lesion suggestive of high flow. Note the markedly dilated internal cerebral vein.*

which subsided after 45 minutes. Thirty hours after presentation, all her neurological symptoms and signs had resolved spontaneously. The extra-ocular movements were full, both pupils were equal in size with normal reactions and there was no ptosis.

A MRI scan showed a right-sided deep trans-cortical arteriovenous malformation (AVM) with medial extension in a subependymal location in the right lateral ventricle (Fig. 1). A carotid angiogram confirmed a high flow, Spetzler-Martin grade 4 AVM (2), the nidus of which measured 4 cm in maximum diameter. The feeder vessel of the AVM originated from the right middle cerebral artery. There was no contribution to the lesion from the posterior circulation. The AVM had both superficial and deep venous drainage (Fig. 2). Superficial venous drainage was via the superficial middle cerebral vein to the right transverse sinus. The deep venous drainage was via thalamostriate veins into internal cerebral vein and vein of Galen (Great cerebral vein). The thalamostriate veins showed focal stenotic lesions while the internal cerebral vein and vein of Galen were markedly dilated. Venous reflux was noted around the midbrain from the vein of Galen. There was no evidence of intracranial haemorrhage in any of the scans (Fig. 2).

The patient was subsequently transferred to a neuro-surgical unit. A lumbar puncture showed no evidence of intracranial bleed or raised intracranial pressure. Since her symptoms were mild and the AVM was large and deep, no further intervention was contemplated by neurosurgeons. She remains under observation, with regular reviews at the AVM clinic in a neurosurgical unit.

DISCUSSION

AVMs are congenital complex tangles of abnormal arteries and veins linked by one or more fistulae, which lack capillary beds, allowing significant shunting of blood (1, 3). AVMs are uncommon with a prevalence of 0.1 %, affecting both sexes equally; 85% -90% of the AVM are supratentorial, mostly located superficially and primarily supplied by the carotid circulation. The remainder are located in the posterior fossa and are supplied by vertebrobasilar system (1). Intracranial haemorrhage is the most common presentation (30 to 82 %) followed by seizures, headaches, focal transient or progressive neurological deficits and syncope (1, 3-5).

The clinical features of AVM depend on its space occupying effect, or complications such as subarachnoid or intracerebral haemorrhage and raised intracranial pressure (1, 3). Large AVM may shunt blood away from otherwise normal brain tissue and produce symptoms due to a steal phenomenon (1, 3, 5, 6). Anomalies of venous drainage such as kinking, stenosis, thrombosis or venous aneurysms are often associated with AVM (3). Venous hypertension in high flow AVM, mass effect of dilated veins or focal venous stagnation with passive congestion of the brain parenchyma are other proposed mechanisms of neurological deficits (3, 7-9).

AVMs presenting with brief focal neurological signs may cause diagnostic confusion. In view of our patient's young age and pupillary involvement, neuroimaging was requested. The AVM in our patient was however located away from the midbrain and the course of the third nerve. Other conditions such as vasospasm of migraine attack may cause transient 3rd nerve palsy. However ophthalmoplegic migraine generally affects a younger age group (<10 years) and the ophthalmoplegia occurs at the height of the headache or follows it (10). Our patient had no previous history of migraine. Demylinating disorder can have a similar presentation but there was no evidence of demyelination on MRI scan in our patient. Rapidly expanding aneurysms can also rarely cause transient third nerve

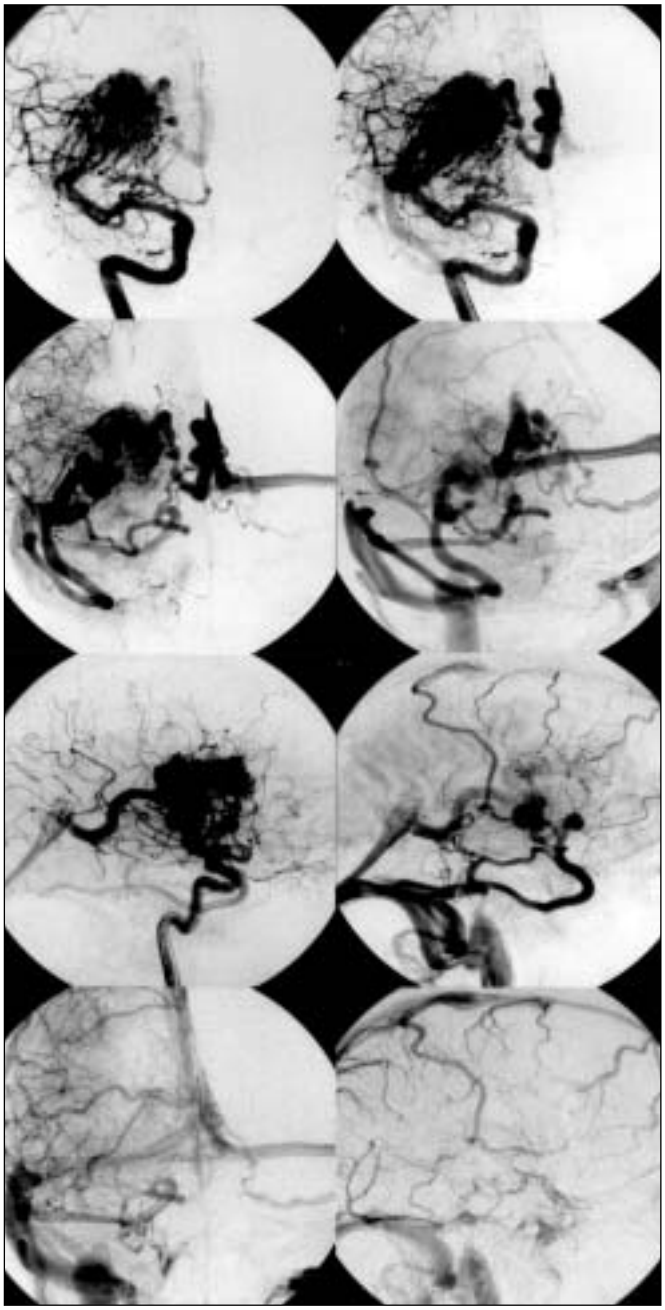

Fig. 2 - *Neuro carotid angiogram demonstrating a Spetzler-Martin grade 4 lesion. The arterial blood supply is from the right middle cerebral artery. Deep venous drainage is via a markedly dilated internal cerebral vein & vein of Galen. There is venous reflux involving the vein of Galen. The thalamostriate veins show focal stenotic lesion.*

palsy (11). In an older patient the differential diagnosis should also include vascular causes such as diabetes mellitus, hypertension or giant cell arteritis.

Although we can only speculate that the transient third nerve palsy was related to the arteriovenous malformation, this is the most likely possibility. Studies have shown that unruptured arteriovenous malformations frequently cause mass effect. The contributing factors include dilated tortuous draining veins, elevated draining vein pressure, the size of the AVM and any associated brain oedema (8, 9). High pressure in the venous drainage system may also produce venous ischemia with neurological signs and symptoms (12). In our patient dilated veins were noted around the lateral and posterior mid brain. There was also marked venous reflux from the vein of Galen into the tectal vein and via the collicular veins around the midbrain to the petrous vein. We postulate that in our patient, the signs & symptoms were related to a transient venous phenomenon with the underlying mechanism being the mass effect from the dilated veins around the midbrain at the height of cortical venous reflux. In addition, the arteriovenous malformation in our patient was high flow and was likely to be associated with high venous pressure, which in itself can produce cortical ischemia. Steal phenomenon is unlikely to explain the oculomotor nerve palsy in our patient because the feeder vessel of the AVM originated from the middle cerebral artery with no contribution from posterior circulation. However, past episodes of fainting in our patient may have been induced by cerebral ischemia due to steal phenomenon.

This case illustrates unusual neuro-vascular abnormality in a young patient with transient III nerve palsy. Ophthalmologists may be the first clinician to assess a patient with transient visual symptoms associated with headaches. The palsy in our patient may have been missed if she had presented a day later. A careful history evaluating the visual symptoms and a thorough neuro-ophthalmological examination to elicit any subtle resolving neurological signs in case of transient attacks, are important in identifying the underlying pathology. In summary transient third nerve palsy in young patients warrants full investigations including neuroimaging.

Reprint requests to:
Ashish Agrawal, Ms (Ophth), MrcOphth, FrcsEd
Specialist Registrar
5, Woodlea Park
Meanwood, UK
ashishtanu@supanet.com

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