Bilateral proptosis, dilatation of conjunctival veins, and papilloedema: a neuro-ophthalmological syndrome caused by arteriovenous malformation of the torcular Herophili

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SUMMARY A patient with an occipital dural arteriovenous malformation (AVM) developed signs of a carotid-cavernous sinus fistula and raised intracranial pressure. Bilateral transverse sinus occlusion associated with the AVM produced these signs by rerouting intracranial venous drainage anteriorly through the cavernous sinuses and superior ophthalmic veins. Angiography and computerised tomographic reformation techniques were used to define these extraordinary cranio-orbital venous pathways.

Orbital and ocular signs of a carotid-cavernous sinus fistula can be produced by any posteriorly located arteriovenous malformation (AVM) draining anteriorly into the orbital venous system.^{1–9} Ocular signs of raised intracranial pressure (ICP) can be produced by any intracranial AVM that causes a rise of pressure in the posterior dural sinuses.¹⁰

We report major alterations in cerebral, dural, and orbital venous systems, confirmed by angiography and computerised tomography (CT), in a patient whose AVM at the torcular Herophili (confluence of superior sagittal, straight, and transverse sinuses) caused a clinical syndrome of bilateral proptosis, dilated conjunctival veins, and papilloedema.

Case report

This 25-year-old woman had a 4-year history of raised ICP that was caused by a dural AVM of the torcular Herophili. In August 1979 she noted proptosis of the

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Fig. 1 Top: Face photograph showing bilateral asymmetrical proptosis (exophthalmometer readings right 25 mm, left 22 mm) and enlarged left angular vein (arrow). Bottom: Right eye showing dilated conjunctival veins.

Fig. 2 Top: Common carotid angiogram showing the torcular AVM (arrow), with feeder vessels from the occipital artery, posterior branches of the middle meningeal artery, and meningeal branches of the internal carotid artery. Bottom: Venous drainage of the malformation through the superficial middle cerebral vein (above), and deep cerebral veins (below), into the cavernous sinus (CS) and superior ophthalmic vein (SOV) (small arrows indicate direction of flow).

right eye, redness of both eyes, and increasing headaches. Her visual acuity was 6/6 bilaterally, and she had asymmetrical proptosis that was bilateral, but more marked on the right (Fig. 1, top), dilated conjunctival vessels in both eyes (Fig. 1, bottom), and a prominent angular vein on the right. Intraocular pressures were normal. She had chronic papilloedema bilaterally and a partial homonymous hemianopia of the left visual field.

Selective carotid and vertebral angiography showed an occipital dural AVM with feeding vessels from the right and left occipital arteries, the posterior branches of the right and left meningeal arteries, and tentorial branches of the right and left internal carotid arteries (Fig. 2, top). Both transverse venous sinuses were totally occluded. Venous drainage from the AVM and from the brain flowed in a retrograde direction (Fig. 2, bottom) through: (1) the straight

sinus, the vein of Galen, and deep cerebral veins; (2) the superior sagittal sinus and the superficial middle cerebral veins; (3) the meningeal veins.

These venous pathways and all others from within the skull entered the cavernous sinuses and the superior ophthalmic veins. The latter veins were markedly dilated (Fig. 3, top).

Axial CT scans showed portions of the occipital AVM, as well as the dilated basal veins and the anterior extensions of these veins toward the anterior cavernous sinuses.

By means of a research 'trace programme' (General Electrical Medical Systems, Milwaukee, Wisconsin, USA) vertical reformations along the wandering path of the dilated and contrast-enhanced veins in the basal cisterns permitted accurate identification of the point at which the enlarged basal veins penetrated the dura around the cavernous sinus, the superior orbital

Fig. 3 Top: Axial computerised tomography scan showing trace programme along a contrast-enhanced basal vein. Bottom: Lateral view of vertical reformation of same vein.

Fig. 4 Top: Postcontrast axial computerised tomography scan showing markedly dilated superior ophthalmic veins. Bottom: Postcontrast axial scan indicating enlarged optic nerve sheaths.

fissure, and entered the orbit (Fig. 4). In addition the CT scans of the orbit showed enlargement of the optic nerves secondary to dilatation of the vaginal sheath spaces (Fig. 3, bottom).

Radical resection of the dural AVM relieved the elevated pressure in the retrograde venous outflow channels at the base of the brain in the cavernous sinuses and in the orbital venous system. The patient's proptosis resolved, and the conjunctival redness and papilloedema disappeared.

Discussion

When posteriorly located dural AVMs cause bilateral transverse sinus occlusion,¹¹ a major rerouting of venous blood from the head must occur. Blood shunted through the AVM flows anteriorly in a retrograde direction through the basal, cortical, and dural veins toward the cavernous sinus. As exemplified by the CT and angiographic findings in the patient we describe, the basal veins become grotesquely enlarged, their calibre exceeding that of the carotid arteries. Before they pierce the dura of the anterior cavernous sinus they occupy a position first adjacent and just lateral to the optic nerves and chiasm, and then lateral and inferior to the anterior clinoid processes.

In the absence of adequate alternative venous outflow channels from the cavernous sinus, blood from the basal veins, together with blood from the sphenoparietal and cavernous sinuses, flows in a retrograde manner into the orbital venous bed. As a result orbital veins dilate, and the patient develops bilateral proptosis and dilated conjunctival vessels, thus simulating the features of a carotid-cavernous sinus fistula. These signs, together with evidence of raised ICP, including papilloedema, constitute an unusual neuro-ophthalmic syndrome suggesting a dural AVM with major sinus occlusion.

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