Cerebellar arteriovenous malformation presenting with bilateral proptosis mimicking caroticocavernous fistula

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INTRODUCTION

Caroticocavernous fistula (CCF) is a clinical syndrome consisting of pulsating exophthalmos, chemosis, bruits accompanied by orbital–frontal headaches and extraocular palsy. Associated features may also include loss of visual acuity and fifth cranial nerve involvement. The majority of cases are the result of trauma to the internal carotid artery resulting in direct communication between the high-pressure lumen of the artery and the low-pressure cavernous sinus (Jones and Ross, 1989). Less commonly, it results from a weakened atherosclerotic internal carotid arterial wall, dural arteriovenous fistulae in the cavernous sinuses or connective tissue disease involving the carotid artery (Forlodou et al, 1996). The authors describe a patient with cerebellar hemispheric arteriovenous malformation (AVM) who manifested the classic clinical signs of CCF.

DISCUSSION

There are two possible pathophysiological mechanisms that could explain this patient's findings: the first that the remote facial trauma in the patient's history resulted in a direct CCF, and the second that the right cerebellar hemispheric AVM caused major rerouting of venous blood. The former possibility is excluded by the normal carotid angiography. The second postulated mechanism, while quite unusual, is significantly more plausible. A posteriorly located AVM can rarely mimic the orbital and ocular signs of CCF

CASE REPORT

A 21-year-old man presented with a 3-year history of bulging and redness of his right eye associated with double vision. He had a history of facial trauma 5 years previously. These symptoms, which had progressed over the year preceding the presentation, were accompanied by a bilateral pounding headache and gait unsteadiness with veering to the right. On examination, he had bilateral pulsating exophthalmos that was more marked on the right with a loudly audible orbital bruit. Mild soft tissue swelling of the right eyelid along with dilated veins in the right conjunctival fornices was observed. His visual acuity was 20/20 and there was no papilloedema. Eye movement examination showed restricted up-gaze, in conjunction with skew deviation, the right eye being hypertropic. Although impaired convergence was observed, there was no convergence retraction nystagmus. Right lateral rectus palsy and right sensorineural hearing loss were noted. Coordination testing revealed right finger to nose and heel to shin ataxia associated with a slightly wide based gait.

The magnetic resonance images showed several findings of interest. There was a massive right cerebellar arteriovenous malformation, with a large tangle of vessels extending into the quadrigeminal plate and perimesencephalic cistern with mass effect on the right aspect of the midbrain (*Figure 1*). There was bilateral proptosis, significantly more pronounced on the right, with enlargement of the left lateral rectus muscle and bulging of the right cavernous sinus. Surprisingly, no significant enlargement of the right superior ophthalmic vein was noted (*Figure 2*). Cerebral angiography showed a massive right cerebellar arteriovenous malformation, fed by the bilateral posterior inferior cerebellar arteries, anterior inferior cerebellar arteries and superior cerebellar arteries, as well as direct branches from the basilar artery and right posterior cerebral artery. Superficial venous drainage was observed directly to the right transverse and sigmoid sinuses, as well as to the deep venous system, with an enlarged straight sinus, vein of Galen and internal cerebral veins. There was also retrograde forward-directed venous flow filling the right cavernous sinus and the inferior petrosal sinus. Emanating from the right cavernous sinus (*Figure 3*), there were two anterior directed veins: a smaller superior one, representing a cortical vein and a larger inferior vein, representing the right superior ophthalmic vein, which was only mildly dilated. The inferior ophthalmic vein was not visualized. No fistulous communication between the carotid artery and cavernous sinus was seen.

Figure 1. Axial T1-weighted magnetic resonance image demonstrating a large right cerebellar hemispheric arteriovenous malformations with mass effect on the right aspect of the midbrain.

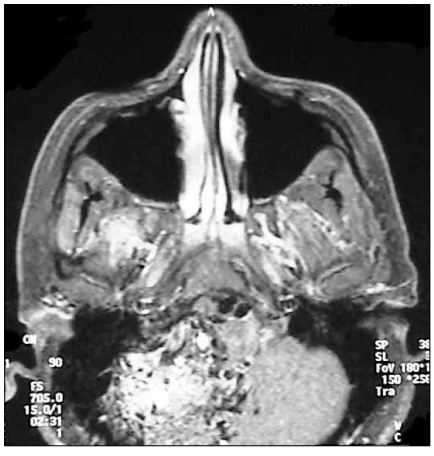
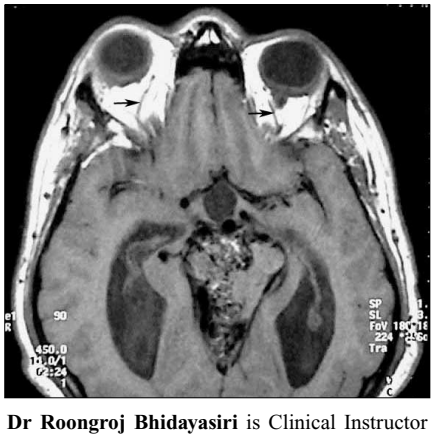


Figure 2. Axial T1-weighted magnetic resonance image demonstrating bilateral superior ophthalmic veins, which were mildly dilated (arrow). Bilateral proptosis was noted, more marked on the right.



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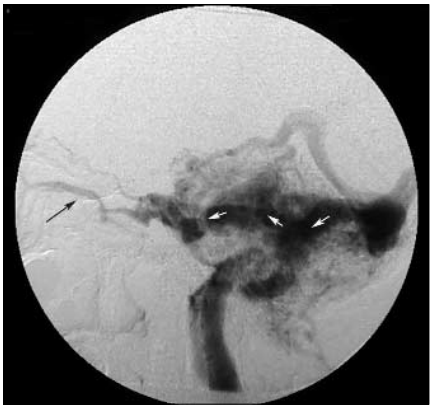

Figure 3. Right vertebral artery injection, lateral view. This showed massive right cerebellar arteriovenous malformations with retrograde forward-directed venous flow filling the right cavernous sinus (white arrows). Right superior ophthalmic vein was seen (black arrow).

through retrograde drainage of venous blood into the orbital venous system.

Under normal conditions, cerebellar venous blood drains into the great cerebral vein, straight sinus, transverse sinus and petrosal sinuses. However, in this case, blood shunted through the AVM flowing anteriorly in a retrograde direction through the large perimesencephalic and periclival veins towards the cavernous sinus, resulting in increased orbital venous pressure. In addition, a high-pressure head of arterial blood in the right cavernous sinus was most likely transmitted across the intercavernous sinus to the left cavernous sinus and then the left superior ophthalmic vein, resulting in contralateral proptosis.

In previously published case reports of CCF and CCF mimics, marked dilatation of the superior ophthalmic vein seemed to be a constant finding because the abnormal increased blood flow from the fistula or AVMs entered the cavernous sinus, causing retrograde flow and increased orbital venous pressure through the trabeculae, which was then transmitted to the superior ophthalmic veins (Buchanan et al, 1982; Kurosu et al, 1998; Volpe et al, 2000; Anderson et al, 2001; Stiebel-Kalish et al, 2002). In contrast, this case presents an unusual finding of lack of a significantly dilated superior ophthalmic vein in the setting of clinically evident increased orbital venous pressure.

Although the angiogram did not provide any clues to explain this unusual phenomenon, it is conceivable that the venous hypertension noted in this case was a result of a wide and even distribution of pressure without dilating any specific vein to a great degree. This distribution of pressure could occur along the superior ophthalmic vein as well as along the inferior ophthalmic vein and the vortex veins. This notion also suggests that pressure lower than that which is necessary for venous dilatation may still be capable of producing symptoms and signs of increased orbital venous pressure.

This patient's findings of diplopia, restricted up-gaze, skew deviation and impaired convergence could be the result of the mass effect of the AVM on the dorsal midbrain, consistent with Parinaud's syndrome, while sensorineural hearing loss was probably secondary to mass effect on the seventh and eighth nerve complex at the right cerebellopontine angle.

CONCLUSIONS

This case demonstrates that CCF mimics may be caused by a variety of vascular lesions that cause a rise of pressure in the posterior dural sinuses. In addition, it reveals that the lone vein mechanism for this clinical picture as represented by a massively enlarged superior ophthalmic vein, although common, is evidently not a necessary angiographic finding. Lastly, this case report suggests that a classic presentation of CCF, when accompanied by non-classic findings such as cerebellar findings, should prompt a search for a CCF mimic. HM

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