

1A

Acute Painful Ptosis, Complete Ophthalmoplegia with a Red Eye

CASE NO. 1A

A 55-year-old white female presented in the emergency room (ER) with a chief complaint of the onset of a headache during the previous four days. The headache resolved the following day, but she began having swelling of her right eye (OD). This worsened over the following few days and she developed diplopia. She related no eye pain but did complain that the right side of her face was numb.

On examination, her visual acuity was 20/100 OD and 20/20 OS. The pupillary examination showed a dilated and fixed pupil OD; the pupil OS was normally reactive to light. Ocular motility showed complete ophthalmoplegia OD and was full OS (Fig. 1A.1). External examination showed complete ptosis and marked proptosis OD (Fig. 1A.2). Hertel measurement showed 7 mm of proptosis OD. There was a prominent orbital bruit over the right eye. Slit lamp examination showed dilatation of the conjunctiva and episcleral vessels with diffuse injection. There was no chemosis. The cornea was clear, the anterior chamber was quiet, and the lens was normal. Examination OS was unremarkable. Intraocular pressure was 26 mm Hg OD and 18 mm Hg OS. Ophthalmoscopic examination was normal in each eye.

Discussion

Dr. Brazis' comments: The above clinical scenario is most consistent with a carotid-cavernous sinus fistula (CCF) which is an abnormal



Fig. 1A.1. Ocular motility photography showing the almost complete ophthalmoplegia in the right eye.



Fig. 1A.2. External photography showing the complete right ptosis.

communication between the cavernous sinus and the carotid arterial system. CCFs can be classified by etiology (e.g. traumatic versus spontaneous), by velocity of blood flow (e.g. high versus low flow), or by anatomy (e.g. direct versus dural; internal carotid artery versus external carotid artery supply versus both). In this patient, I would be

concerned about a direct high flow connection between the cavernous segment of the internal carotid artery and the cavernous sinus. These fistulas are of a high flow type, and because they are often caused by a single tear in the arterial wall they are thus called direct CCFs.

Direct CCFs represent 70% to 90% of all CCFs in most large clinical series. They occur in both men and women and may occur at any age. A direct CCF results from a single tear in the wall of the cavernous segment of the internal carotid artery. This produces a direct connection between the artery and one or more of the venous channels within the cavernous sinus. Direct CCFs most often are the result of head trauma, especially motor vehicle accidents, fights, and falls. The injury may be extremely severe or quite trivial. A substantial minority of direct CCFs are caused by rupture of a pre-existing aneurysm of the cavernous segment of the internal carotid artery. Direct CCFs may also be the iatrogenic, occurring after various diagnostic and therapeutic procedures (e.g. carotid endarterectomy, cranial and percutaneous retro-Gasserian procedure for the trigeminal neuralgia, transphenoidal surgery, or maxillofacial surgeries).

Although direct CCFs are usually not thought to be life-threatening, there are numerous reports of patients experiencing significant and even fatal epistaxis, subarachnoid hemorrhage, or intracerebral hemorrhage from rupture of the fistula. The ocular manifestations of a direct CCF are usually ipsilateral to the side of the fistula but may be bilateral or even contralateral. The lateralization of ocular manifestations depends on the venous drainage of the cavernous sinuses, including the connections between the two sinuses through the intercavernous sinuses and the basilar sinus, the presence or absence of cortical venous drainage, and the presence or absence of thrombosis within the sinus or a superior ophthalmic vein on one or both sides.

Proptosis is a common sign, occurring in almost all patients. In most cases proptosis develops rapidly on the side of the fistula and becomes pronounced within a few days. In the early stages of a direct CCF, the eyelids may become moderately or severely swollen. When the superior ophthalmic vein is enlarged, the medial portion

of the upper eyelid may be ptotic or swollen. Conjunctival chemosis to some degree occurs in most patients. In more severe cases, the inferior palpebral conjunctiva may prolapse through the interpalpebral fissure and rarely may cause conjunctival necrosis or superinfection. As arterial blood is forced anteriorly into the orbital veins, the conjunctival and episcleral veins becomes dilated, tortuous, and filled with arterial blood. This “arterialization” of the conjunctival vessels is the hallmark of a CCF. Although it may initially be mistaken for conjunctivitis or episcleritis, the dilation and tortuosity of the affected vessels is usually quite distinctive, running to the limbus and often arching back in a loop. Ocular pulsations are caused by transmission of the pulse waves from the internal carotid or ophthalmic artery to the ophthalmic veins. Abnormal ocular pulsations may be visible on applanation or other tonometry and may sometimes produce pulsating exophthalmos that may be palpable.

Exposure keratopathy is the most frequent corneal sign encountered in patients with a direct CCF. The keratopathy may be aggravated by a concomitant trigeminal neuropathy caused by injury or by the effects of the fistula on the trigeminal nerve within the cavernous sinus. In some patients with a direct CCF, the initial symptom may be a pulsatile tinnitus or subjective bruit which may or may not be associated with an audible bruit. Diplopia occurs in 60% to 70% of patients with direct CCF. The diplopia may be caused by dysfunction of one or more of the ocular motor nerves, the extraocular muscles, or both, and the degree of motility limitation varies from mild to complete ophthalmoplegia. Visual loss associated with a direct CCF may be immediate or delayed. Immediate visual loss is usually caused by ocular or optic nerve damage at the time of the head injury. Delayed visual loss is usually caused by retinal dysfunction, but it may be related to vitreous hemorrhage, central retinal vein occlusion, angle closure glaucoma from anterior rotation of the ciliary body, anterior ischemic optic neuropathy, or even corneal ulceration. Dilation of the retinal veins may be seen and can be asymptomatic but can produce venous stasis retinopathy or even frank central retinal vein occlusion with macular edema and diffuse retinal hemorrhages.

Occasionally, patients complain of facial pain in the distribution of the first and rarely the second division of the trigeminal nerve. Some patients will have decreased corneal sensation, decreased facial sensation, or both, perhaps related to ischemia or compression of the ophthalmic or maxillary divisions of the trigeminal nerve within the cavernous sinus. Glaucoma may develop in up to 30% to 50% of patients, and most commonly the rise in intraocular pressure is due to increased episcleral venous pressure or orbital congestion. Neovascular glaucoma may also occur associated with chronic retinal hypoxia and retinal neovascularization.

Finally, as in this patient, a direct CCF should be suspected in any patient who suddenly develops chemosis, proptosis, and a red eye. If there is no history of trauma, one should consider the possibility of a ruptured cavernous aneurysm.

Dr. Lee's comments: Dr. Brazis has provided a nice summary of direct and indirect CCFs. The indirect CCF may spontaneously involute and in the absence of vision threatening proptosis, exposure keratopathy, debilitating ophthalmoplegia, visual loss, or severe pain, many patients can be observed for spontaneous resolution from the natural history of thrombosis in these lesions. Non-catheter based neuroimaging (e.g. MRI and MRA or CTA) might be sufficient to confirm the diagnosis but most patients require catheter angiography. We have also used orbital ultrasound to follow the patient for spontaneous thrombosis and determine if clinical worsening might be paradoxically related to radiographic thrombosis and not an increase in the flow in the CCF. Halback *et al.* reviewed the angiographic and clinical data from 155 patients with CCF. The features associated with an increased risk of morbidity and mortality included the following:

- presence of a pseudoaneurysm
- large varix of the cavernous sinus
- venous drainage to cortical veins, and
- thrombosis of venous outflow pathways distant from the fistula.

The clinical signs and symptoms of a potentially “hazardous” CCFs included:

- increased intracranial pressure
- rapidly progressive proptosis
- diminished visual acuity
- hemorrhage and
- transient ischemic attacks.

The cortical venous drainage from the carotid cavernous fistula is due to occlusion or absence of the normal venous outflow pathways and can produce increased intracranial pressure or intraparenchymal hemorrhage. Cavernous sinus varix and extension into the subarachnoid space (with associated risk of potentially fatal subarachnoid hemorrhage) are also considered to be angiographic risk factors for morbidity and mortality. Standard catheter angiography in these patients might be diagnostic, prognostic, and in some cases therapeutic (i.e. angiographically induced thrombosis of the CCF). Direct CCF, on the other hand, typically require catheter angiography as the clinical signs and symptoms are more acute and severe, and as Dr. Brazis mentioned, the catheter angiogram can be diagnostic and in the same or sequential sittings be coupled with endovascular treatment of the CCF.

CT scanning, CT angiography (CTA), MR imaging, and MR angiography (MRA) can be used to confirm the diagnosis, showing enlargement of the extraocular muscles, dilation of one or both superior ophthalmic veins, enlargement of the affected cavernous sinus, and abnormal intracranial vessels (e.g. enlarged or increased flow voids on MRA or MRI in cavernous sinus). The ultimate diagnostic test, however, is catheter angiography.

Contrast brain MRI in this patient demonstrated right proptosis, enlarged flow void inside the right cavernous sinus, and an enlarged superior ophthalmic vein (Figs. 1A.3 and 1A.4). Catheter angiography revealed an internal carotid artery aneurysm (Fig. 1A.5) as the cause of the CCF.

The optimum treatment of a direct CCF is closure of the abnormal arteriovenous communication with preservation of the internal



Fig. 1A.3. Brain MRI axial T1 without contrast demonstrates right proptosis (*small arrow*) and enlarged flow void inside the right cavernous sinus (*large arrow*).



Fig. 1A.4. Brain MRI Axial T2 demonstrates the enlargement of the right superior ophthalmic vein (*arrow*).

carotid artery patency. Endovascular closure of direct CCF is most often accompanied by embolization using a variety of agents, primarily coils and detachable balloons. Complications most often result from balloon occlusion of a direct CCF when there is planned or



Fig. 1A.5. Right carotid angiogram demonstrates an aneurysm in the internal carotid artery (*arrow*).

unplanned occlusion of the internal carotid artery. These complications, primarily those related to reduction or interruption of the blood supply to the ipsilateral eye and cerebral hemisphere, include stroke and even death. Once the fistula is successfully closed, most of the ocular symptoms and signs resolve or will at least improve and do not recur. Of all the symptoms and signs of a direct CCF, visual loss is least likely to improve after successful treatment even when the internal carotid artery remains patent.