CASE REPORT

Vertebral–venous fistula: an unusual cause for ocular symptoms mimicking a carotid cavernous fistula

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SUMMARY

Vertebral–venous fistulas (VVF), or vertebral–vertebral arteriovenous fistulas, are an uncommon clinical entity. Typically, they present as a result of a direct vascular connection between an extracranial branch of the vertebral artery or its radicular components and the epidural venous plexus. These may manifest with signs and symptoms referable to cervical myelopathy secondary to compression or steal phenomenon. To our knowledge, this is the first case to identify a patient who presented with classic ocular symptoms attributable to a carotid cavernous fistula but secondary to a VVF. We present its treatment and clinical outcome. In addition, we present a brief literature review surrounding this uncommon disease.

BACKGROUND

Carotid cavernous fistulas (CCFs) are an aberrant connection that results from shunting between the carotid artery and the cavernous sinus. The contents of the cavernous sinus include the internal carotid artery, along with several cranial nerves, including the oculomotor nerve, trochlear nerve, V1 and V2 branches of the trigeminal nerve, and the abducens nerve.¹ As a result of the disruption of the normal hemodynamics with compression of the cavernous sinus contents, patients with a CCF commonly present with proptosis, chemosis, orbital bruits, ophthalmoplegia, and headaches.^{2 3}

In addition, patients not uncommonly complain of symptoms referable to retinal ischemia, such as diplopia and pain in the orbital region. Less often presentations can include intracerebral or subarachnoid hemorrhage.⁴ The severity of the clinical manifestations are dependent on the cumulative pressure within the cavernous sinus, and the subsequent ability to accommodate the increased venous pressure.²

In contrast, vertebral–venous fistulas (VVF) result from a high flow connection between the extracranial vertebral artery and the surrounding epidural vertebral venous plexus. Spetzler *et al* suggested that extradural arteriovenous fistulas present as symptoms associated with cord compression. This is secondary to venous congestion or a vascular steal phenomenon.⁵ As a result, patients manifest with progressive myelopathy.

Our patient is unique in that the extradural vertebral artery caused venous congestion that manifested as symptoms clinically referable to the ocular presentation of a CCF.

CASE PRESENTATION

An individual presented with progressively worsening right eye pain, decreased visual acuity, and orbital swelling, occurring acutely over several days. In

Figure 1 MR venography depicting venous congestion with filling of the superior ophthalmic vein.

Figure 2 Anteroposterior projection of the right vertebral artery (RVA) in mid-arterial phase DSA, showing an abnormal fistulous connection.

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Figure 3 Lateral projection of the left vertebral artery (LVA) in early arterial phase DSA, showing an abnormal fistulous connection at the level of C1, draining into the anterior condylar vein.

retrospect, the patient detailed a history of penetrating neck injury several years previously, without needing further investigation or treatment. The initial intraocular pressure was documented as above 30. On examination, ophthalmoplegia, chemosis, and proptosis were present. No ocular bruit was auscultated. MR venography showed suspicious venous congestion and retrograde drainage ([figure 1](#)).

Because of a previously documented anaphylactic reaction, the initial cerebral angiogram was performed using 30 mL of gadopentetate dimeglumine (Magnevist; Bayer). At the level of the C1 vertebrae, a fistula originating from the right vertebral artery segment with the anterior condylar confluence was found ([figures 2 and 3](#)). Although there was antegrade flow through the internal jugular vein, there was obvious reflux into the inferior petrosal sinus, cavernous sinus, and ultimately the superior ophthalmic vein ([figure 4](#)). In addition, there was physiologic

Figure 4 Lateral projection of the left vertebral artery (LVA) demonstrating the venous pathway showing retrograde drainage through to the superior ophthalmic vein.

Figure 5 Pretreatment microcatheter injection into the arterial feeder from the ascending pharyngeal artery with fistulous connection.

occlusion of the right vertebral artery proximal to the origin of the posterior inferior cerebellar artery, which was filled retrograde from the contralateral vertebral artery.

The patient tolerated endovascular occlusion with coils and glue. Within 12 h post-treatment, the patient's chemosis and ophthalmoplegia had significantly been reduced without cranial neuropathies. In addition, intraocular pressures were found to have normalized, and the patient was discharged to acute rehabilitation on postoperative day 3.

TREATMENT

A minor component of the fistula supplied by a branch of the right ascending cervical artery ([figure 5](#)) was first catheterized and occluded with 0.2 mL of 40% n-butyl cyanoacrylate ([figure 6](#)). Then, the main component of the fistula supplied by the

Figure 6 Post-treatment microcatheter injection depicting the n-butyl cyanoacrylate cast across the fistula. n-BCA, n-butyl cyanoacrylate.

Figure 7 Anteroposterior projection of the right vertebral artery (RVA) showing occlusion of the fistulous connection. n-BCA, n-butyl cyanoacrylate.

right vertebral artery was catheterized. In order to ensure a durable result, despite an already physiologically occluded proximal right vertebral artery, we proceeded to occlude the right vertebral artery proximal to the occlusion. The fistulous connection along the distal right vertebral artery was also occluded, with a combination of coils and 40% n-butyl cyanoacrylate (figure 6). A follow-up run confirmed obliteration of the fistulous connection (figures 7 and 8).

OUTCOME AND FOLLOW-UP

Resolution of the presenting clinical symptoms was found immediately after operation and at the 6 month follow-up examination.

DISCUSSION

In general, VVFs are uncommon. In a recent review, Briganti *et al* documented 20 previously reported cases, all with symptoms referable to cord compression. They reported that the most common etiology of VVF is from penetrating trauma, similar to this case.⁶ This may have resulted in delayed occlusion at the level of trauma with a fistulous connection occurring with the anterior condylar confluence, leading to retrograde venous

Figure 8 Lateral projection of the left vertebral artery (LVA) confirming occlusion of the vertebral venous fistula. n-BCA, n-butyl cyanoacrylate.

congestion in our patient. The late venous phase showed antero-grade occlusion of the left transverse sinus and caudally.

Currently, endovascular intervention is favored as firstline treatment of VVFs. A variety of devices, including detachable balloons, and transarterial or transvenous coils are used to occlude the fistula while attempting to maintain the patency of the vertebral artery.^{6–9} Our case report involved venous drainage into the anterior condylar confluence, which is the pathway for causing ocular symptoms, due to involvement of the inferior petrosal sinus.^{10 11} In an extensive review, Spittau *et al* elegantly depicted the pathophysiology and treatment of hypoglossal canal dural arteriovenous fistulas, which may present with ocular symptoms similar to our case. Due to the complexity of the arterial–venous connections, a transjugular approach was the most commonly used treatment method, reserving transarterial treatment for lesions without venous access.¹² In their meta-analysis, the ascending pharyngeal, middle meningeal, and occipital arteries were the primary arterial feeders.

Our case is unique in that the physiologically occluded vertebral artery was the primary source and allowed for successful transarterial treatment.

Learning point

- Use established therapeutic principles to diagnose and treat unusual pathological entities.

Contributors All aspects of manuscript preparation were performed by all of the authors.

Competing interests None declared.

Patient consent Not obtained. The patient was not able to physically sign the form. Telephone consent was obtained with a two physician witness.

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