CASE REPORT

An unusual presentation of a dural arteriovenous fistula of the sphenoparietal sinus

Andrew Macdonald, Puneet Plaha, James Byrne

Department of Neurosurgery,
John Radcliffe Hospital,
Oxford, Oxfordshire, UK

Correspondence to

Andrew Macdonald,
Department of Neurosurgery,
John Radcliffe Hospital,
Headley Way, Oxford,
Oxfordshire OX3 9DU, UK;
andrewmacdonald@gmail.com

Received 6 January 2014
Revised 12 February 2014
Accepted 14 February 2014

SUMMARY

Dural arteriovenous fistulae (DAVFs) are a rare form of intracranial arteriovenous malformation. We present the case of a patient who presented in a previously undescribed manner with facial swelling and bruising initially misdiagnosed as cellulitis. He was found subsequently to have DAVFs of the sphenoparietal sinus that had hemorrhaged. The rarity of DAVFs at this location may account for this unique presentation. Successful treatment was achieved by transarterial embolization.

BACKGROUND

Dural arteriovenous fistulae (DAVFs) comprise an abnormal shunt between dural arteries and dural veins or sinuses. Presenting symptoms depend on the location of arteriovenous shunting and pattern of venous drainage. DAVFs are found most frequently at the transverse/sigmoid sinus (approximately 60%) followed by the cavernous sinus (10%).¹ Typically, patients present with pulsatile tinnitus and chemosis with exophthalmos respectively. Other presentations include headache, cranial nerve palsy or other neurological deficits which may be related to spontaneous intracranial hemorrhage.

DAVFs that drain into the sphenoparietal sinus are particularly rare and were first reported by Pakarinen in 1965.² We describe a previously unreported presentation of a patient with a sphenoparietal DAVF. This involved the rapid onset of facial swelling, bruising and tenderness in the absence of any abnormal neurological or ophthalmological signs. Its unique nature led to a delay in diagnosis.

CASE PRESENTATION

A 42-year-old man presented to his local emergency department (ED) with left-sided facial swelling, bruising and tenderness in the absence of significant headache. Three days previously he awoke in the morning with these symptoms. He strongly denied any history of trauma. He had no past medical history of note and his only relevant family history was the death of his mother due to aneurysmal subarachnoid hemorrhage at the age of 60 years.

On examination he was systemically well but with marked tenderness and swelling over the left temple and orbit (*figure 1A*). Ophthalmological examination including acuity, fields and eye movements was unremarkable. There was no chemosis or proptosis.

Figure 1 Photographs of the patient on initial presentation (A) and at 6-month follow-up (B).

Although complaining of feeling lethargic, he was alert and orientated with a Glasgow Coma Scale score of 15. There were no cranial or peripheral nerve deficits on neurological examination and he did not have photophobia or neck stiffness.

INVESTIGATIONS

Standard hematological and biochemistry tests were normal. An additional erythrocyte sedimentation rate test was performed but this was not significantly raised. A diagnosis of periorbital cellulitis was made and he was discharged home with a course of oral flucloxacillin.

He returned to the ED the following day, however, with worsening of his symptoms and complaining of an additional sensation of fullness in the left ear, although he denied any tinnitus. There was associated dizziness (rotational vertigo) and nausea. Ear examination revealed a bulging tympanic membrane and there was concern that his constellation of symptoms could be due to a severe sinusitis leading to otitis media and periorbital cellulitis.

A non-enhanced CT scan of the sinuses, which included the brain, was performed. This demonstrated normal sinuses but, more alarmingly, acute subarachnoid blood in the left Sylvian fissure, extending inferiorly around the anterolateral margin of the temporal lobe (*figure 2*). There was additional swelling of the soft tissues overlying the left temporal region. CT angiography was

CrossMark

To cite: Macdonald A, Plaha P, Byrne J. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-011103

Figure 2 Axial unenhanced CT showing acute subarachnoid hemorrhage in the left Sylvian fissure (A) and extracranial swelling in the left temporal region (B).

performed following transfer to the regional neurosciences center. This did not show an aneurysm or arteriovenous malformation. However, it did show abnormal vessels spanning the bone behind the left orbit and anterior to the middle cranial fossa, extending deep to the temporalis muscle and deep vascular grooves in the inner table of the left squamous temporal bone. A DAVF was suspected and the patient proceeded to catheter angiography.

Digital subtraction angiography was performed with injections to both the internal and external carotid arteries and the left vertebral artery. These demonstrated a DAVF supplied by the left middle meningeal artery which drained to the sphenoparietal sinus with enlarged dural viens. Early filling of the cavernous sinus was evident, with drainage to both inferior petrosal sinuses and to the left ophthalmic and facial veins (figure 3A,B). Shunting to cortical veins was not evident, but this was inferred as the cause for the spontaneous subarachnoid hemorrhage. A small saccular aneurysm was found arising from the left superficial temporal artery remote from the intracranial fistula (figure 3C).

DIFFERENTIAL DIAGNOSIS

Facial pain and swelling around the temporal and periorbital region has a broad differential diagnosis including cellulitis, erysipelas and temporal arteritis. The difficulty and delay in diagnosis was therefore understandable in this instance. However, in retrospect, the sudden onset of these symptoms waking the patient from sleep may have given some indication of an acute vascular event.

TREATMENT

Transarterial embolization was proposed after multidisciplinary team review and endovascular treatment performed under general anesthesia. Two abnormal arteriovenous connections

Figure 4 Lateral radiograph showing high-density embolization material (Onyx) in both dural veins and arteries as well as the sphenoparietal sinus (arrow).

were identified on selective angiography of the middle meningeal artery and these were occluded by injection of Onyx 18 (Covidien) (figure 4). The patient was discharged home the following day.

OUTCOME AND FOLLOW-UP

At 6-month follow up the patient reported complete resolution of his symptoms and all swelling had resolved (figure 1B). He underwent repeat angiography, which confirmed stable occlusion of the fistula.

DISCUSSION

Most DAVFs are thought to be acquired lesions, related to a previous significant head injury or sinus thrombosis where the insult opens a pre-existing physiological site of arteriovenous shunting in the dura mater. However, many patients present without such history, and a recent report highlighted an association that suggests a genetic component to their formation. In a recent analysis of a cohort of 56 patients with DAVFs, Gross *et al*³ found that 14% had additional aneurysms at sites remote from the DAVF.

Our patient had no history of previous significant head trauma and, given his family history of maternal death due to aneurysmal subarachnoid hemorrhage, the DAVF may have a

Figure 3 Lateral digital subtraction angiogram after injections of the left external carotid artery. (A) Two shunt sites (short arrows) between the middle meningeal artery and dural veins. These drain to the sphenoparietal sinus (long arrow, B) during the late arterial phase, when the distal superficial temporal artery also fills and a saccular aneurysm is seen (arrowhead, B). In the venous phase (C), enlarged dural veins (short arrows) and the cavernous sinus are seen filling (dotted arrow).

congenital rather than acquired etiology. The presence of bone erosion implies that it was longstanding.

While it is unclear what triggered spontaneous bleeding from this presumably longstanding fistula, the anatomical correlation between his symptoms, imaging findings and its resolution following intervention show this to be a new and unusual presentation of a DAVF.

The patient's unusual presentation adds to a body of cases of idiosyncratic presentations of DAVFs in the literature. These include dementia and hearing loss.^{4 5} While a rare cause of

facial swelling and pain, this case is a reminder of the ability of DAVFs to produce a plethora of symptomatology and the diagnosis should be considered more readily in differential diagnosis. In particular, clinicians should be alert to a history of cranial trauma, surgery or family history of aneurysms.

Contributors All authors contributed significantly to the preparation and editing of this manuscript.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Learning points

- ▶ Dural arteriovenous fistulae (DAVFs) can present in a variety of idiosyncratic fashions.
- ▶ Spontaneous onset facial swelling and bruising is a newly described presentation of a DAVF.
- ▶ Clinicians should be mindful to consider DAVF in their differential diagnosis and to consider appropriate vascular imaging.

REFERENCES

- 1 Piippo A, Niemela M, van Popta J, *et al.* Characteristics and long-term outcome of 251 patients with dural arteriovenous fistulas in a defined population. *J Neurosurg* 2012;118:923–34.
- 2 Pakarinen S. Arteriovenous fistula between the middle meningeal artery and the sphenoparietal sinus. A case report. *J Neurosurg* 1965;23:438–9.
- 3 Gross BA, Ropper AE, Du R. Cerebral dural arteriovenous fistulas and aneurysms. *Neurosurg Focus* 2012;32:E2.
- 4 Hurst RW, Bagley LJ, Galetta S, *et al.* Dementia resulting from dural arteriovenous fistulas: the pathologic findings of venous hypertensive encephalopathy. *AJNR Am J Neuroradiol* 1998;19:1267–73.
- 5 Kim MS, Oh CW, Han DH, *et al.* Intraosseous dural arteriovenous fistula of the skull base associated with hearing loss. Case report. *J Neurosurg* 2002;96:952–5.

Copyright 2014 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow