A rare case of unilateral hemifacial spasm and facial palsy associated with an abnormal anatomical variant of the posterior basilar circulation

J Chan¹, K Jolly², A Darr³, DJ Bowyer¹

¹Department of Otolaryngology, Princess Royal Hospital, Shrewsbury and Telford Hospital NHS Trust, Telford, UK

²University Hospitals Birmingham NHS Foundation Trust, West Midlands, UK

³Royal Wolverhampton NHS Trust, West Midlands, UK

ABSTRACT

Tortuous vertebral arteries are a rare anatomical variant. Mild tortuosity is usually asymptomatic whereas severe tortuosity may present with ischaemic symptoms or compressive symptoms (focal neurological deficit). While a resulting hemifacial spasm has been previously described, sparse literature exists for its association with facial palsy. We present a rare case of facial spasm along with facial palsy in a 67-year-old woman who was found to have an anatomical variant in the posterior basilar circulation with an ectatic basilar artery and significantly displaced posterior vertebral artery impinging on the facial nerve.

KEYWORDS

Facial nerve disorders – Hemifacial spasm – Vertebrobasilar dolichoectasia

Accepted: 24 March 2019

CORRESPONDENCE TO

Jacqueline Chan, E: jacqueline.chan@nhs.net

Case history

A 67-year-old woman presented to the otolaryngology outpatient department with a two-year history of intermittent left hemifacial spasms and more recent facial palsy. At the onset of the hemifacial spasms the patient had been assessed by the neurology team; magnetic resonance imaging (MRI) had identified significant vertebrobasilar dolichoectasia with a tortuous loop of the left vertebral artery causing compression of the brain stem and displacement of the left seventh cranial nerve (fig 1).

Three-months prior to presentation to otolaryngology she had also developed left perioral paraesthesia, which progressed over 12 hours to a left lower motor neurone grade IV (House–Brackmann scale) facial palsy. These more recent symptoms of facial weakness had resolved completely over three to four weeks, prior to presentation at the otolaryngology clinic.

Clinical examination of the ears, cranial nerves and cerebellar function were normal. Pure tone audiometry demonstrated a symmetrical bilateral mild to moderate sensorineural hearing loss, which was unchanged compared with previous audiometric testing two years previously.

Laboratory tests were undertaken for causes of facial palsy. This included a vasculitis screen (antineutrophil cytoplasmic antibodies and antinuclear antibodies), Lyme's disease serology, serum angiotensin-converting enzyme, serum calcium, inflammatory markers and random glucose. All laboratory test results were normal.

Further neuroimaging was undertaken (fig 1). This once again highlighted an anatomical variant in the posterior basilar circulation, with an ectatic basilar artery and significantly lengthened and tortuous vertebral artery that was indenting the brain stem and impinging on the facial nerve; this had not changed from the earlier MRI in 2016 (fig 2).

The patient had previously had neurology-directed trials of gabapentin and carbamazepine for her hemifacial spasms. These drugs had been unsuccessful, and the patient had declined an offer of botulinum toxin injections into the facial musculature. Following the onset of the facial weakness, the patient was referred for a neurovascular surgical opinion to consider decompressive surgery. This was considered to be inappropriate for the clinical features and imaging findings. The patient's hemifacial spasms are non-progressive and there have been no further episodes of facial weakness.

Figure 1 Axial T2 weighted magnetic resonance image of 2016 showing tortuous vertebral and basilar arteries impinging on seventh cranial nerve.

Figure 2 Axial T2 weighted magnetic resonance image of 2018 showing ectatic basilar artery and significantly displaced posterior vertebral artery impinging the seventh cranial nerve.

Discussion

Tortuous vertebral arteries form part of a condition known as vertebrobasilar dolichoectasia (VBD) and are a rare entity. VBD is characterised by tortuous, elongated and dilated vertebral or basilar arteries. The estimated incidence of VBD varies between 0.06% and 5.8%.¹ The clinical presentation can vary from asymptomatic patients to those presenting with compressive manifestations and ischaemic symptoms. A common compressive manifestation is impingement of cranial nerves. Multiple case reports have reported involvement of cranial nerves III, IV, V, VI, VII and VIII; the majority of these patients present with symptoms of trigeminal neuralgia or hemifacial spasm.¹ There have also been reports of concurrent cranial nerve neuropathies associated with VBD; Lakhan described concurrent trigeminal and facial nerve involvement secondary to VBD leading to hemifacial spasms and facial pain.² While the literature has extensively described a resultant hemifacial spasm from VBD, sparse literature exists in its association with facial palsy. Additionally, no literature has described VBD associated with hemifacial spasm and subsequent facial palsy.

The underlying aetiology and pathophysiology of VBD is not clearly understood. It has been suggested that the disease process is multifactorial and involves the tunica

media. Risk factors, including age, male sex, hypertension, smoking and coronary artery disease,³ are associated with disruption of the internal elastic lamina, which in turn leads to activation of metalloproteinases, causing breakdown of extracellular proteins located within the tunica media.

Historically, investigation of VBD involved cerebral angiography, an invasive method. However with advances in medical imaging, MRI and magnetic resonance angiography have become the investigations of choice. These investigations allow for accurate assessment of the wall of the artery and the presence of intraluminal thrombus, together with the anatomical relations of the affected artery with surrounding structures, including cranial nerves, the brain stem and third ventricle.

Significant complications can arise as a result of VBD, including ischaemic stroke, brainstem compression and obstructive hydrocephalus. The frequency of complications was found to be associated with increased arterial diameter. A systematic review of 375 patients found that the five-year risk of transient ischemic attack was 10.1%, 17.6% for ischaemic stroke, 10% for brainstem compression and 5% for intracerebral haemorrhage.³

Medical management of hemifacial spasm may be considered in early stages of the condition when symptoms are mild and spasms occur infrequently. Drugs that have been reported to be effective include carbamazepine and

benzodiazepines such as clonazepam.⁴ The treatment of choice for hemifacial spasms however, is the use of botulinum toxin injections. The method effectively relieves spasms three to five days following the injection and is effective for approximately six months.⁴

Surgical management of VBD remains challenging due to surgical access and the close association with neurovascular structures. Microsurgery for management of trigeminal neuralgia and hemifacial spasm has previously been described. Ferreira *et al* described management of compression of cranial nerve VII via microsurgical decompression by repositioning of the vertebral artery and undertaking a pexy.⁵

Conclusions

This rare case illustrates an uncommon cause of facial palsy. It is also a rare case of vertebrobasilar dolichoectasia causing hemifacial spasm and subsequently facial palsy. Although rare, anatomical variants in cerebral vasculature should be considered in the evaluation of patients presenting with facial palsy, hemifacial spasm, or a combination of

both. Studies have suggested that VBD is associated with significant complications and thus serious morbidity and mortality. This further highlights the importance of identifying the condition, if present. If VBD is found to be present, patients should be referred for further neurosurgical evaluation.

References

1. Mohammed K, Iqbal J, Kamel H *et al*. Obstructive hydrocephalus and facial nerve palsy secondary to vertebrobasilar dolichoectasia: case report. *Surg Neurol Int* 2018; **9**: 60.
2. Lakhan S. Teaching neuroimages: concurrent hemifacial spasm and trigeminal neuralgia due to vertebrobasilar dolichoectasia. *Neurology* 2013; **81**(8): e52.
3. Wolters FJ, Rinkel GJE, Vergouwen MDI. Clinical course and treatment of vertebrobasilar dolichoectasia: a systematic review of the literature. *Neurol Res* 2013; **35**(2): 131–137.
4. Rosenstengel C, Matthes M, Baldauf J *et al*. Hemifacial spasm. *Dtsch Arztebl Int* 2012; **109**(41): 667–673.
5. Ferreira M, Walcott BP, Nahed BV, Sekhar LN. Vertebral artery pexy for microvascular decompression of the facial nerve in the treatment of hemifacial spasm. *J Neurosurg* 2011; **114**(6): 1,800–1,804.