consideration, at least, for genetic testing for CADASIL. This may be particularly important if there is a family history of stroke; given that stroke is so common, this family history could easily be dismissed as irrelevant in the assessment of suspected bvFTD. Finally, Patient 2 also suggests that CADASIL might possibly be an under-recognised diagnosis in apparent bvFTD. Screening Notch3 in a substantial and unselected cohort of FTD patients might be appropriate to investigate this possibility.

Conflict of interest/disclosure

The authors declare that they have no financial or other conflicts of interest in relation to this research and its publication.

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Spontaneous vertebral arteriovenous fistula causing cervical myelopathy and acute ischemic strokes treated by endovascular balloon–assisted coiling and Onyx embolization

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ABSTRACT

Vertebral arteriovenous fistulas (VAVF) are infrequent lesions characterized by abnormal communication of the extracranial vertebral artery or one of its branches to the surrounding venous plexuses, without the presence of any intervening vessels. We describe a rare occurrence of a patient with VAVF presenting with acute ischemic stroke, encephalomalacia from multiple prior embolic events, and cervical myelopathy, which was successfully treated by coil-assisted Onyx embolization (ev3 Endovascular, Plymouth, MN, USA) with balloon for flow arrest. Our patient demonstrates that point occlusion with embolization for VAVF can be a feasible, safe, and effective treatment option for complete obliteration of the fistula, with subsequent reduction in the volume of the intra-spinal canal venous plexus. Although it is postulated that thromboembolism is less common because of redirection of flow to the venous side of the fistula, our patient also illustrates the potential for to–fro flow in such a fistula to result in embolic injury to the distal circulation.

1. Introduction

Vertebral arteriovenous fistulas (VAVF) are rare lesions characterized by abnormal communication of the extracranial vertebral artery (VA) or one of its branches to the surrounding venous plexuses, without the presence of any intervening vessels or capillaries.[1] They can be either spontaneous or traumatic in origin. Neurological sequellae from vertebral arteriovenous (AV) fistulas can arise from a number of mechanisms including mechanical compression or venous hypertension of the spinal cord and secondary arterial steal. These in turn depend on the pattern of venous drainage, flow velocity, and chronicity of the AV fistula. Obliteration of the AV fistula with preservation of the parent artery is the primary goal of treatment.

We describe a rare occurrence of a patient with VAVF presenting with acute and chronic ischemic strokes and cervical myelopathy which was successfully treated by coil-assisted Onyx embolization (ev3 Endovascular, Plymouth, MN, USA) with balloon for flow arrest.

2. Case report

A right-handed hypertensive patient in their 60s presented with acute onset vertigo. Examination revealed left-appendicular ataxia, left C5 innervated muscle weakness, decreased left bicep reflex, and hyperreflexia in both legs. A review of systems was positive for pain and hyperesthesia in the left arm for 10 years.

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2.1. Neuroimaging

MRI of the brain showed scattered punctuate acute infarcts in the left cerebellum with background encephalomalacia (Fig. 1). MRI of the cervical spine revealed a lobulated, extramedullary enhancing structure adjacent to the left neural foramen at C2–3 with cord compression (Fig. 2). Digital subtraction angiography showed an ectatic left V3 segment with pseudoaneurysm measuring 3.8 × 3.4 × 2.3 cm, and slow drainage into the left suboccipital plexus and into an epidural vein within the spinal canal causing cord compression.

Disconnection of the fistula was performed with temporary occlusion using an ASCENT occlusion balloon (DePuy, Raynham, MA, USA), followed by the deployment of four 0.018'' bare platinum coils into the shunt followed by injection of Onyx LES-34 (eV3 Endovascular), which filled the coil frame selectively (Fig. 3).

Acetylsalicylic acid and atorvastatin were initiated for secondary stroke prevention. The cerebellar symptoms resolved and there was mild improvement in the left arm weakness. Follow-up electromyography (EMG) 2 months later showed normal sensory studies and severe chronic motor axon loss in the C5–7 myotomes bilaterally. There was active motor axonal loss in the left C5–7 myotomes suggesting ongoing compression of the spinal anterior horn cell or root. MRI after 4 months revealed continued prominent left sided epidural intra-canal flow voids, most notable at the C4–5 level. Repeat embolization was attempted, but minimal flow into the pouch was observed, and no significant change of angiographic flow could be achieved.

Follow-up MRI after 11 months demonstrated involution of the epidural vein, with return of the spinal cord to near midline. There were no new areas of infarction when compared to the imaging at the onset of symptoms.

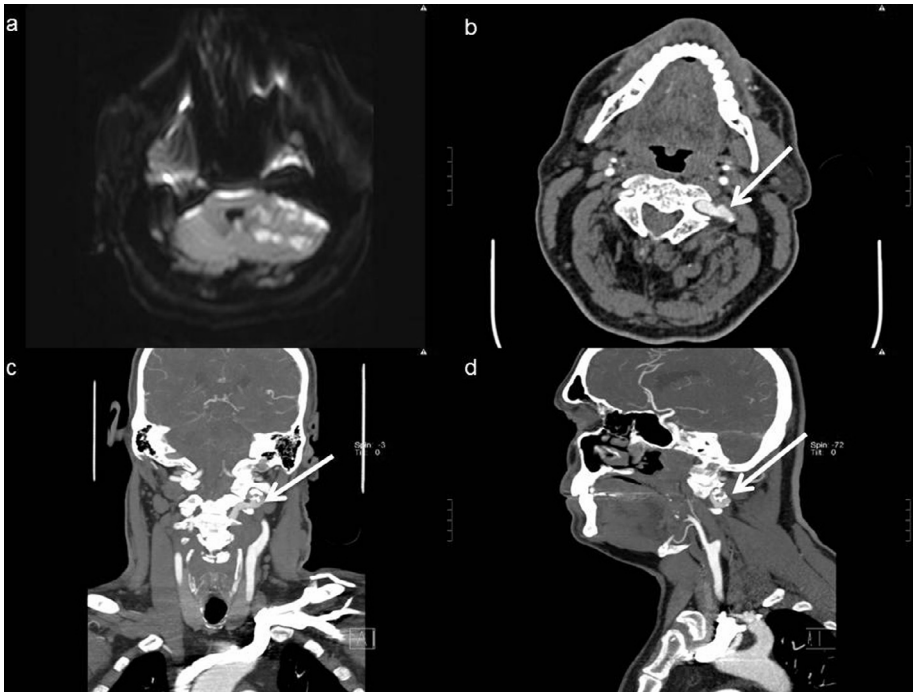

Fig. 1. (a) Axial diffusion weighted sequence MRI showing scattered acute ischemic infarcts in the superior cerebellar artery and posterior inferior cerebellar artery territory of the left cerebellar hemisphere. (b) Axial, (c) coronal and (d) sagittal CT angiography images showing a complex pseudoaneurysm (arrow) arising from the left vertebral artery between C1 and C2 on the left side.

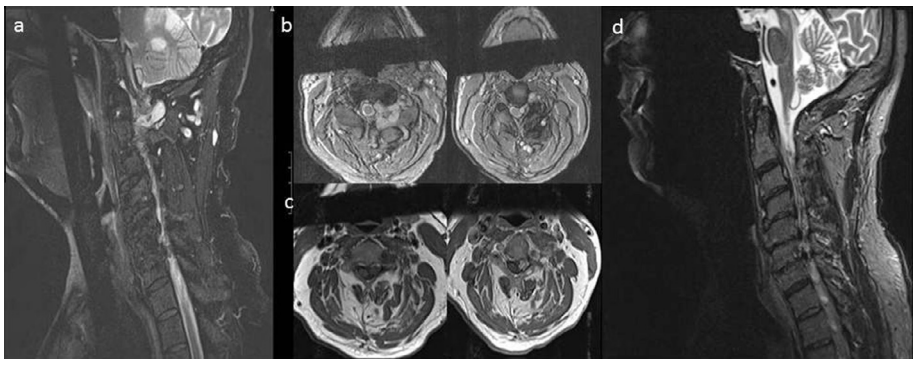

Fig. 2. (a) Sagittal fat-suppressed short T1 inversion recovery MRI of the cervical spine showing a lobulated soft tissue mass arising adjacent to C2–3, compressing the thecal sac and spinal cord on the left as seen on (b) axial gradient echo and (c) axial T1-weighted sequences. (d) Sagittal fat-suppressed short T1 inversion recovery MRI of the cervical spine post coil and Onyx embolization (ev3 Endovascular, Plymouth, MA, USA) showing obliteration of the vascular mass.