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Case Report

A rare case of a cervical dural arteriovenous fistula presenting in a younger patient with vertex subarachnoid hemorrhage: Case report and literature review ☆

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ABSTRACT

Spinal dural arteriovenous fistulas (SDAVF) are rare entities, which mainly occur in the thoracic and lumbar regions of older adults with a significant male predominance. The clinical manifestations are nonspecific such as myelopathy and this can make it challenging to diagnose. Rarely these have been described in the cervical region with even rare manifestations including subarachnoid hemorrhage. In the few reports of hemorrhage, it is usually infratentorial. We present a case of 40-year-old female (uncommon gender and younger age) who presented with headaches and was found to have supra tentorial subarachnoid hemorrhage and a suspicious lesion in the spinal canal. This proved to be a type 1 AVM of the spinal canal (per the American British and French classification). This was successfully treated endovascularly. With the discussion of the relevant literature we hope that this case can add to our medical knowledge as another presentation of this uncommon condition and ultimately help in diagnosing this illusive and rare entity.

Introduction

Spinal vascular shunts are rare entities (5%-9% of all central nervous system vascular malformations) [1] and comprise of fistulas and arteriovenous malformations [2]. The mean age is 50-60 years, with a male to female ratio of 5:1 and the common location being the thoracolumbar region [1]. The classical manifestation of these entities is progressive myelopathy [1]. There have been rare cases of these entities presenting as subarachnoid hemorrhage (SAH) where the blood is usually present either over the spinal cord, in the basal cisterns or posterior fossa [3]. We present a rare case of a cervical level dural arteriovenous fistula (dAVF) in a female who was below the mean age, presented with headaches as the main symptoms and was found to have non cisternal pattern of SAH.

☆ Conflicts of Interest: None.

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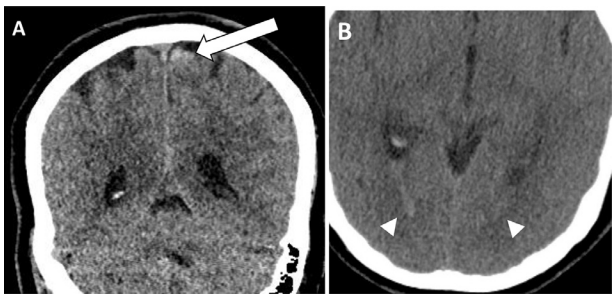

Fig. 1 – (A) Coronal noncontrast head computed tomography reveals trace subarachnoid hemorrhage over the left parietal lobe (white arrow). (B) Axial reconstructed image shows intraventricular hemorrhage in the occipital horns of lateral ventricles (arrow heads).

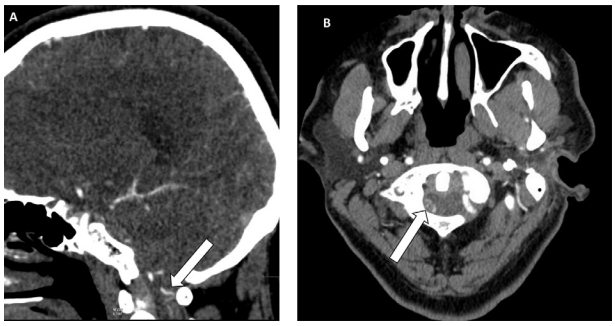

Fig. 2 – CT angiogram (A and B) shows early filling of serpiginous structures in the dorsal thecal sac (arrow) concerning for underlying spinal shunt. Compare to Fig. 6, post procedural exam.

Case Description

The patient, a 40-year-old female without significant medical history, presented with 2-day history of acute onset severe headache, accompanied by neck tightness, episodes of nausea and vomiting. On examination, the patient was in moderate distress and drowsy, but arousable and oriented. Vitals were within normal limits. Comprehensive neurologic exam was unremarkable. Computed tomography (CT) scan of the head showed trace acute SAH near the vertex as well as intraventricular hemorrhage in the occipital horns of lateral ventricles (Fig. 1). CT angiogram of the head and neck did not demonstrate an intracranial malformation. However, a subtle right sided arteriovenous shunt at the level of C2-C3 was seen, raising a suspicion for a spinal dAVF (Fig. 2). Magnetic resonance imaging of cervical spine revealed serpiginous flow voids in the right dorsal aspect of the thecal sac at the level of C2-C3, also supporting a spinal dAVF (Fig. 3). Digital subtraction angiography identified a fistulous communication between right C2 radicular branch of right vertebral artery and peri medullary vein, consistent with a Dural AVF, a type 1 AVM as per the American, British and French classification discussed below (Fig. 4).

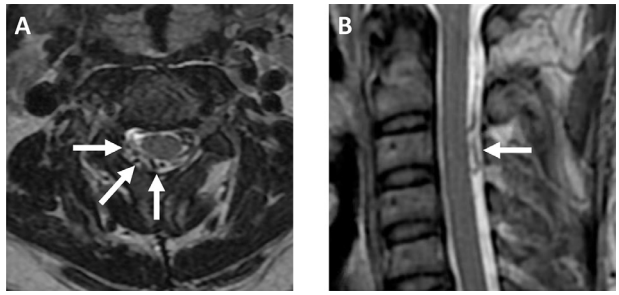

Fig. 3 – (A, B) Axial and sagittal T2 weighted magnetic resonance imaging serpiginous flow voids at the right dorsal aspect of dorsal thecal sac at the level of C2-C3.

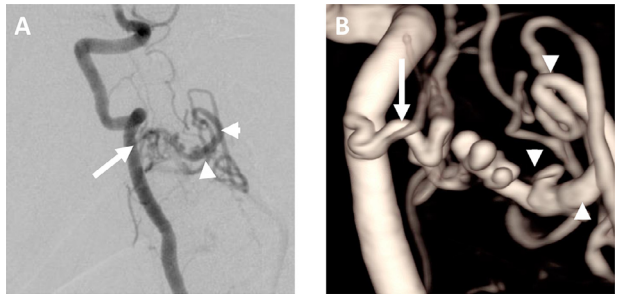

Fig. 4 – (A, B) Diagnostic digital subtraction angiography and 3D reconstruction of the right vertebral artery demonstrating an arteriovenous fistula between radicular branch of the vertebral artery (arrow) and peri-medullary vein (arrow heads).

The patient underwent endovascular treatment with n-Butyl cyanoacrylate glue embolization 2 days after the diagnostic angiography. The immediate post-embolization angiography demonstrated near-complete occlusion of the fistula with residual sluggish flow to the fistula (Fig. 5). A follow-up CT angiogram of the neck showed reduction in size of the feeding radicular artery without filling of peri-medullary vein, indicating successful treatment of the AVF (Fig. 6). The post-treatment course was uneventful, and the patient was discharged after 10-days of admission.

Discussion

The vascular supply of the spinal cord is in the form of a single anterior spinal artery (ASA); which supplies the anterior two thirds and paired posterior spinal arteries (PSA) which supply the posterior cord [2]. The ASA originates from the 2 branches of the vertebral arteries and travels along the anterior median fissure [2]. The PSA originate from the intradural portion of the vertebral artery or the posterior inferior cerebellar artery, with contributions from 10 to 23 radiculopial branches [2].

Spinal shunts (dAVF and arteriovenous malformations) are type of vascular malformation which comprise of anomalous connections between the high-pressure arterial blood flow entering into the valve less venous system without the presence

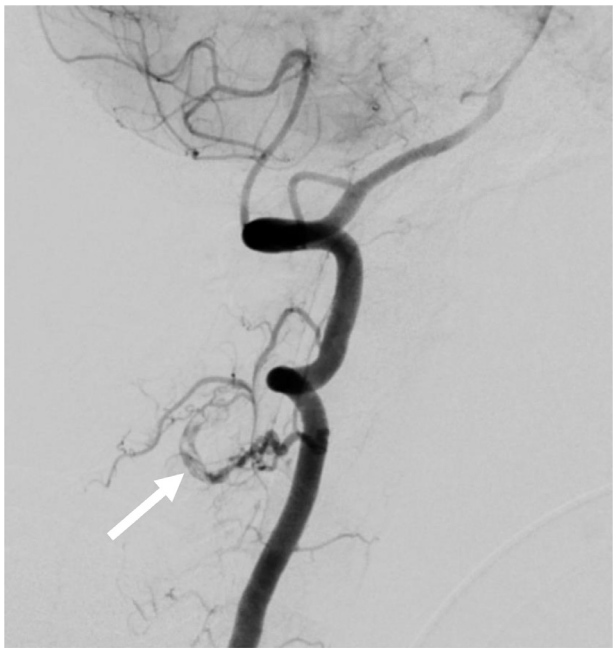

Fig. 5 – Endovascular treatment was performed with n-Butyl cyanoacrylate embolization (arrow), achieving near-complete occlusion.

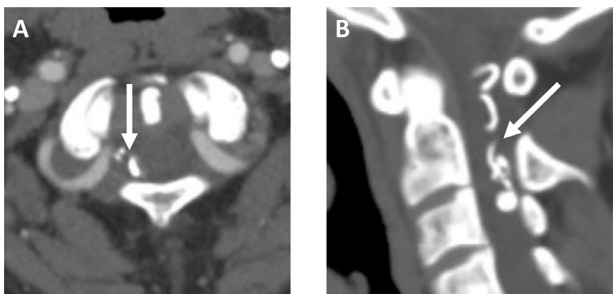

Fig. 6 – (A, B) Axial and sagittal postcontrast neck computed tomography angiography demonstrates successful occlusion of the arteriovenous fistula with radio dense embolization material (arrow).

of an intervening capillary bed [4]. The first clinical observation of a spinal vascular malformation was published in Germany in 1890 by Berenbruch [1]. Whereas the first description by clinical angiography was in 1967 by Di Chiro from National Institute of Health, USA [5]. To date there have been seven classification systems for these entities [5], with the 2 leading systems in use being the American/British/French system and the Spetzler classification [2]. As per the American/British/French system the Type I lesions, or the "single coiled vessel" type, are dAVFs and consist of a radicular artery draining into an engorged spinal vein on the dorsal aspect of the dural sheath of a nerve root [2]. These are the most commonly encountered malformations [4] with the Spetzler classification dividing them into extradural and intradural types (further classified into dorsal and ventral lesions) [4].

The dAVF is thought to originate from venous outflow obstruction (eg, spontaneous thrombosis, trauma) and subsequent arterialization of the coronal venous plexus [2]. In majority of the case the exact etiology cannot be found with some rare cases of postoperative fistulas also described [6].

The spinal dAVFs commonly occur in the thoracolumbar region and are more common in the 6th decade and in males [1]. These generally have a progressive course with clinical deterioration usually taking months to years (19% disability in 6 months and 50% in 3 years as per 1 study) [7]. The underlying mechanism is venous hypertension with broad clinical presentations including lower-extremity weakness and sensory disturbance that may manifest in gait abnormalities, sensory loss, ill-defined lower-back pain, loss of bowel and/or bladder control, as well as sexual dysfunction [4]. The rarer incidence of cervical dAVF vs the thoracolumbar variants has been ascribed to anatomical differences in venous drainage patterns, with small caliber radiculospinal veins [8]. When they do occur in cervical region the dAVF, they have the same demographics in general with presenting symptoms including myelopathy, radiculopathy, and cranial nerve dysfunction [9]. There are a few cases with SAH as the manifestation with a comprehensive review by Aviv et al. demonstrating five such cases at the C3-C6 levels [9].

Detection on imaging depends on the secondary changes produced by the dAVF and the malformation itself [10]. MRI is the modality of choice as both of these can be detected together. This should ideally be performed with contrast [10]. The various changes that the dAVF can produce are hemorrhage, arterial steal, mass effect, or venous hypertension [11]. The most common manifestation of these secondary changes are best visualized on T2 weighted images, manifesting as cord edema and with cord atrophy with long standing disease [10]. Another important imaging hallmark is dilated and coiled perimedullary vessels, which can also be observed on the T2-weighted images as flow voids, which are often more pronounced on the dorsal surface compared with the ventral surface [10]. If the shunt volume is low these may be less conspicuous and are best seen on post contrast imaging and hence the usefulness of adding contrast to these studies [10]. Apart from diagnosis noninvasive evaluation of the shunt location is extremely helpful to guide the invasive conventional angiography [10]. Spinal contrast-enhanced MRA has greatly contributed to localizing these lesions, with techniques such as time resolved MRA adding to the sensitivity of these findings as well as reducing the time for finding the appropriate arterial feeders to these malformations [12]. Digital subtraction angiography remains the gold standard for this technique [12]. On selective angiography, stasis of contrast material in the radiculomedullary arteries, especially the ASA, can be seen [10]. After injection into the segmental artery harboring the AV fistula, early venous filling and retrograde contrast uptake of the radiculomedullary veins is seen, usually with an extensive network of dilated perimedullary veins [10]. With the classic appearance on MRI the differential is limited. If the primary malformation is small and only edema is seen, entities such as glioma, ischemia, or demyelination are included in the differential [10].

The two options in the treatment of SDAVFs are surgical occlusion of the intradural vein that received the blood from the

shunt or endovascular therapy using a liquid embolic agent after super selective catheterization of the feeding radiculomeningeal artery [10]. The key principle is obliteration of the fistulous connection with restoration of normal anterograde arterial flow and venous drainage [4]. For some lesions, surgery remains the treatment of choice, particularly when the arterial supply is in close association with the ASA, PSA, or artery of Adamkiewicz; in those cases, the risk of spinal cord ischemia and worsening of neurological function with curative embolization may be prohibitive [1]. The liquid embolic agents include n-butyl cyanoacrylate and Onyx with these agents showing low recurrence rates (0%-25%) [4,5]. Our case was unique because of the younger age, no preceding symptoms and SAH that occurred over the convexity rather than the cisterns or posterior fossa, which has been described [3]. In our patient endovascular therapy yielded a very good response.

Conclusion

Spinal dAVF are rare malformations which can have nonspecific clinical findings. We present a case of an atypical presentation of this entity in an unusual age group and anatomical location. It is our hope that this case will add to the awareness and consideration of this entity even when the hemorrhage is atypical in location.

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