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Case Report

High cervical spinal subdural hemorrhage as a harbinger of craniocervical arteriovenous fistula: an unusual clinical presentation

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Abstract

BACKGROUND CONTEXT: Craniocervical dural arteriovenous fistula (dAVF) is rare as compared with the typical thoracolumbar dAVFs of the spine and usually presents with hemorrhagic manifestation, predominantly intracranial subarachnoid hemorrhage.

PURPOSE: We describe the first case of craniocervical dAVF with initial presentation as neck pain and spinal subdural hemorrhage.

STUDY DESIGN: Case report.

METHODS: We present the case of a 59-year-old woman who presented with sudden onset of neck pain at an outside institution emergency department (ED) and was discharged after negative cervical spine radiographs. Magnetic resonance imaging of the cervical spine performed because of persistent pain demonstrated presence of high cervical spinal subdural hematoma and she was managed conservatively. She subsequently presented to our ED a week later with headache and was found to have an intraventricular hemorrhage on computed tomography scan of the head, which on subsequent workup with an angiography revealed the presence of a craniocervical dAVF.

RESULTS: Surgical obliteration of the fistula was performed with use of intraoperative angiography as an adjunct to confirm complete fistula obliteration. She had an excellent clinical outcome with no deficits at her last follow-up at 9 months.

CONCLUSIONS: Even though hemorrhagic presentation is fairly common in craniocervical dAVFs, there is no report of a craniocervical dAVF presenting with spinal subdural hemorrhage. The present case further highlights the propensity of these vascular lesions to bleed and emphasizes the clinical importance of including these lesions in the differential diagnosis of hemorrhage in the vicinity of foramen magnum region, whether subarachnoid or subdural in location. Physicians treating spinal pathologies should be aware of this entity and clinical presentation, as an angiography needs to be considered in these cases to direct appropriate referral and treatment.

Keywords: Craniocervical junction; Spine; Dural arteriovenous fistula; Perimedullary arteriovenous fistula; Subarachnoid hemorrhage; Subdural hemorrhage; Vascular disorders

Introduction

Dural arteriovenous fistula (dAVF) accounts for most cases of spinal arteriovenous malformation and most commonly occurs in the thoracolumbar region [1,2]. Hemorrhagic presentation is uncommon with dAVFs, which classically manifest as progressive congestive myelopathy secondary to stagnation of the venous outflow from the spinal cord, resulting in intramedullary venous hypertension and ischemic insult to the spinal cord [1–4]. In contrast, dAVFs located in the craniocervical region are rare, accounting for only up to 2% of such cases; albeit they fairly

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commonly present with hemorrhage [4–8]. Subarachnoid hemorrhage (SAH) remains the most common mode of presentation of craniocervical/high cervical dural and perimedullary AVFs and is reported to occur in up to 45% of cases, which forms the basis of obtaining vascular imaging of the neck to rule out high cervical vascular pathologies such as craniocervical dAVF in case of negative cerebral angiogram in a patient with SAH [4,6,9–12]. Spinal subdural hemorrhage (SDH) is less common as compared with cranial SDH, and usually occurs as a result of trauma, iatrogenic secondary to lumber puncture, or rarely spontaneously in cases of hematological disorders, tumors, or anticoagulant use [13]. No case of cervical spinal SDH secondary to a craniocervical dAVF has been reported earlier as per our review of the literature. Considering the common hemorrhagic presentation of dAVF involving the craniocervical region, the presence of high cervical spinal SDH should lead to a suspicion and appropriate workup to unravel the presence of craniocervical dAVF, as it can be a sentinel event before the usual clinical presentation as an SAH or an intraventricular hemorrhage (IVH), as happened in the present case. We report a case of craniocervical dAVF that presented with neck pain. The patient rebled and sustained an IVH. Further workup revealed presence of ventral spinal SDH, which was subsequently diagnosed to be secondary to a craniocervical dAVF.

Case report

A 59-year-old woman with significant past medical history of breast carcinoma presented with sudden onset of neck pain. She was evaluated at an outside institution emergency department and was sent home after negative cervical spine radiographs. Because of persistent neck pain over the next few days, she underwent magnetic resonance imaging (MRI) of the cervical spine, which revealed presence of a ventral high cervical spinal subdural collection with signal intensity suggestive of a small subdural hematoma (Fig. 1). In view of previous history of breast carcinoma in the past, meningeal metastasis was suspected and she underwent contrast MRI of the spinal column and brain, along with magnetic resonance angiography of the brain to rule out any vascular anomaly. All were negative apart from the presence of ventral spinal SDH. She subsequently presented to our emergency department a couple of days later with worsening headaches and a computed tomography scan showed the presence of an isolated IVH involving the left occipital horn with no evidence of SAH (Fig. 2). A digital subtraction angiography was performed, which showed the presence of an early draining vein following left vertebral artery injection, suggestive of a craniocervical dAVF supplied by meningeal branches of radicular artery originating from the left vertebral artery with no evidence of an aneurysm or intracranial arteriovenous malformation (Fig. 3). A transcondylar far lateral approach was performed with clipping of the arteriovenous fistula. Intraoperative angiogram was performed during surgery to confirm the obliteration of AVF. The patient had an uneventful postoperative course and was discharged home with no neurologic deficits. She was doing well at 9 months follow-up with an intact neurologic examination and a modified Rankin scale score of 0.

Discussion

Spinal arteriovenous malformations are rare vascular lesions with spinal dAVFs being the most common type [2]. Classically, a spinal dAVF is located in the thoracolumbar region, has an arteriovenous shunt within the dural root sleeve fed by the radiculo-meningeal arteries, drains through a single radicular vein, and generally presents in

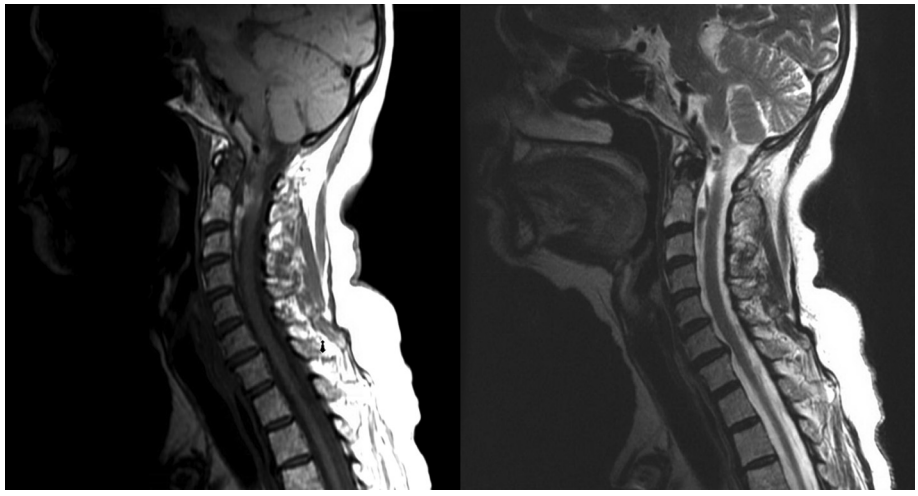

Fig. 1. Sagittal T1-weighted (Left) and T2-weighted (Right) magnetic resonance image of the cervical spine showing presence of a ventral high cervical spinal subdural collection with signal intensity suggestive of a subacute subdural hematoma.

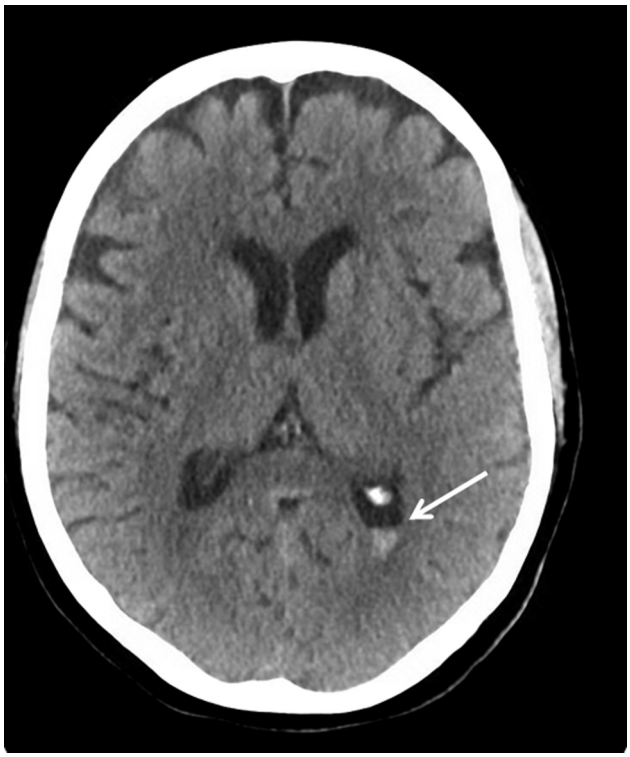

Fig. 2. Computed tomography of the head showing an isolated intraventricular hemorrhage involving the left occipital horn (white arrow) with no evidence of subarachnoid hemorrhage.

a middle-aged man with progressive myelopathy [1,2,5]. Cervical spine is a rare location for spinal dAVFs and when involved, the craniocervical junction is the most favored site. Differences in venous anatomy and drainage pattern between the cervical and thoracolumbar spine have been implicated as a plausible cause for the disproportionate distribution of dAVF with greater preponderance of these in the thoracolumbar spine as compared with the lumbar spine [5]. Although rare, it has been recognized that a dAVF involving the craniocervical regions differs from those occurring in the thoracolumbar regions both in terms of angio-architecture, clinical presentation, and complexity in management [2,4,5,7–9,11,12,14–17]. Although atypical, the angio-architecture of craniocervical dAVF has been described in some recent publications with excellent delineation of the anatomy [8,9,11]. The present case is being reported to highlight the unusual clinical presentation of this rare vascular lesion.

Various case reports and small case series have clearly demonstrated the significantly high rate of hemorrhagic presentation in dAVF involving the craniocervical region [2,4,6,7,9–11,14,16]. Although presence of venous hypertension is suspected as leading to the hemorrhagic presentation in these cases, the exact pathophysiology is not well defined. The increased spinal venous pressure as a result of arterializations diminishes the AV pressure gradient and leads to a decreased drainage of normal spinal veins (eg, the medullary veins, the valveless coronal venous plexus) and venous congestion with intramedullary edema. With spontaneous thrombosis in the dilated vessels, the pressure of the draining veins can rise rapidly, accompanied by venous hypertension and dilatation of anastomotic intracranial veins, which has been significantly associated with SAH. The higher flow rate in the lesions of the cervical area compared with the thoracolumbar area has been proposed as a mechanism for increased bleeding tendency [18]. Angiographic studies have shown that the spinal dAVFs at the craniocervical region are fed by dural branches of the radicular arteries that arise from the vertebral artery and generally drain into the medullary veins.

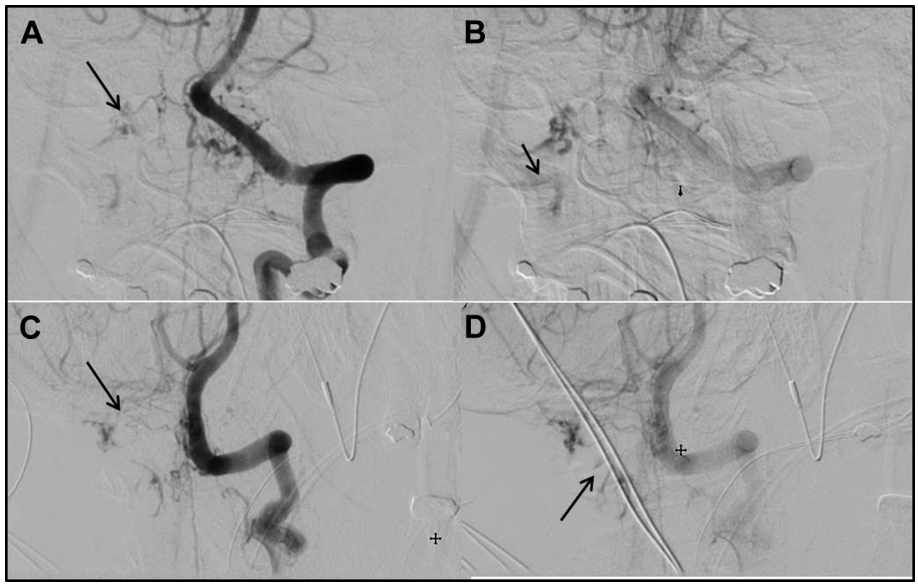

Fig. 3. Anteroposterior (A, B) and lateral (C, D) views after left vertebral injection showing a dural arteriovenous fistula supplied by meningeal branches of radicular artery with an early draining vein. The arrows depict the site of fistula in images A and C and the draining vein in images B and D.

The presence of significant anastomosis between the medullary and pontomesencephalic veins in the foramen magnum region with rostral venous flow of the shunt draining into the cranium may explain the increased tendency of these lesions to present as posterior fossa SAH [3,6,10]. In fact, it has been seen in various studies that venous drainage is generally directed caudally in patients with myelopathy in contrast to the flow of the shunt into the intracranial venous system in patients who present with SAH [10].

Even though no previous case of craniocervical dAVF presenting with spinal SDH has been reported, we assume that the pathophysiology of such a presentation remains the same with leakage of blood into the subdural space. Although conjectural, the possible explanation of this might be that patients with craniocervical dAVF probably had a spinal SDH that went unrecognized because of its asymptomatic nature and the dAVF was finally diagnosed when these patients developed SAH. Nevertheless, as spinal SDH is rare in the absence of any obvious predisposing factor such as trauma or coagulopathy, the presence of a high cervical SDH should lead to a strong suspicion of craniocervical dAVF, considering their common hemorrhagic presentation. Although there are no data, these lesions should be treated sooner rather than later, as they can rebleed again and lead to SAH, the most common presentation of high cervical dAVF. Our patient did sustain an intracranial hemorrhage; albeit the location was intraventricular with no SAH. In fact, the presence of SDH may portend the presence of craniocervical dAVF akin to sentinel bleeds in patients with SAH due to intracranial aneurysms [19].

The complex anatomy of these dAVFs highlights the technical challenges associated with treatment. Henceforth, even though endovascular treatment has been becoming increasingly common for thoracolumbar dAVFs, these lesions render themselves very unsuitable for endovascular treatment options for various reasons, such as the risk of infarction by occlusion of normal anastomotic vessels, the difficulty of introducing a microcatheter into the fine arteries feeding the dAVF, the presence of multiple small fistulas with potential for recanalization after embolization, and incomplete occlusion [7]. Often endovascular treatment needs to be combined with surgery to achieve complete obliteration of the fistula, as reported by Kim et al. [11] Surgery alone has been shown to result in excellent outcome as reported in other small series of patients as well [9,10,14]. In fact, surgical obliteration of the shunts has been the most common modality of treatment in most of the cases of craniocervical dAVF reported in the literature [4]. The importance of intraoperative angiography cannot be overemphasized, as these vascular lesions can often consist of multiple dural feeders both intradurally and extradurally and intraoperative angiography if present can act as a useful adjunct to facilitate total obliteration of the fistula and/or shunts.

Conclusions

Craniocervical dAVFs are a rare subgroup of spinal dAVFs and have different anatomy and clinical presentation as compared with typical thoracolumbar dAVFs. They are often suspected in cases with negative angiography for intracranial aneurysms in cases with SAH, leading to the common recommendation of including bilateral vertebral arteries to rule out these lesions as the cause of SAH especially that restricted to the posterior fossa. Even though hemorrhagic presentation is fairly common in craniocervical dAVFs, there is no report of a craniocervical dAVF presenting with spinal SDH. The present case further highlights the propensity of these vascular lesions to bleed and emphasizes the clinical importance of including these lesions in the differential diagnosis of hemorrhage in the vicinity of the foramen magnum region, whether subarachnoid or subdural in location. An angiography should be considered, as these lesions may be occult on MRI alone and may not be clearly identifiable as was seen in the present case. As spinal pathologies are commonly treated across multiple disciplines, the clinical importance of recognizing this presentation of craniocervical dAVF cannot be overemphasized.

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