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Case Report

Completely Extradural Spinal Arteriovenous Fistula (Abnormal Direct Connection between Arteries and Extradural Venous Plexus) with Totally Extradural Venous Drainage Causing Compressive Cervical Myelopathy

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Abstract

Spinal extradural arteriovenous fistulas are rare vascular lesion which are defined as abnormal direct connection between an artery or arteries and the extradural venous plexus within the spinal canal and/or intravertebral foramen. These lesions with exclusive extradural venous drainage are even rarer. Because of the limited cases the natural history, demography and treatment understanding of these are limited. Endovascular treatment remains the mainstay of treatment for these lesions. This case and its management are thus presented for its rarity.

Keywords: *Arteriovenous fistula, glue embolization, spinal vascular malformation*

Introduction

Spinal Extradural arteriovenous (AV) fistula is rare vascular lesions and is defined as abnormal direct connection between the radicular artery or arteries and extradural venous plexus within the spinal canal and/or intervertebral foramen. They are divided into Type A and Type B depending on the presence or absence of intradural venous drainage, respectively. Because of the lesion's rarity the understanding of its natural history and treatment results is limited. The case and its management are thus presented for its rarity.

Case Report

A 55-year-old man presented with gradually progresive difficulty in walking over a period of 1 year with gradual affection of upper limbs for last 6 months. His clinical examination revealed spastic quadriparesis. Magnetic resonance imaging (MRI) of the cervical spine revealed large dilated extradural flow voids [Figure 1a-c] compressing the spinal canal and displacing the cord. Imaging features were suggestive of cervical vascular malformation and thus an angiographic evaluation was planned.

Treatment details

His digital subtraction angiography was then performed through the right femoral access. The left vertebral artery injection revealed a high flow AV fistula (AVF) fed from the left vertebral artery and draining into the dilated epidural venous plexus [Figure 2a and b]. Venous phase of intracranial circulation in the left vertebral injection revealed intracranial venous drainage through the right transverse sigmoid route, with no drainage through left transverse sinus [Figure 3]. The fistula also had supply from the right thyrocervical trunk [Figure 4]. Additional supply of the fistula was from the dilated left thyrocervical and costocervical trunk [Figure 5] both draining into the dilated epidural venous plexus with completely extradural venous drainage.

Endovascular embolization of the fistula was then planned using N-butyl cyanoacrylate. The left vertebral artery feeder was first microcatheterized using Headway Duo 167 (Microvention Terumo) microcatheter and fistula was embolized using a 50% mixture of lipiodol and N-butyl cyanoacrylate. Postembolization there was obliteration of supply from the vertebral artery and right thyrocervical artery [Figure 6a and b]. Further embolization was performed in the same sitting using another set of

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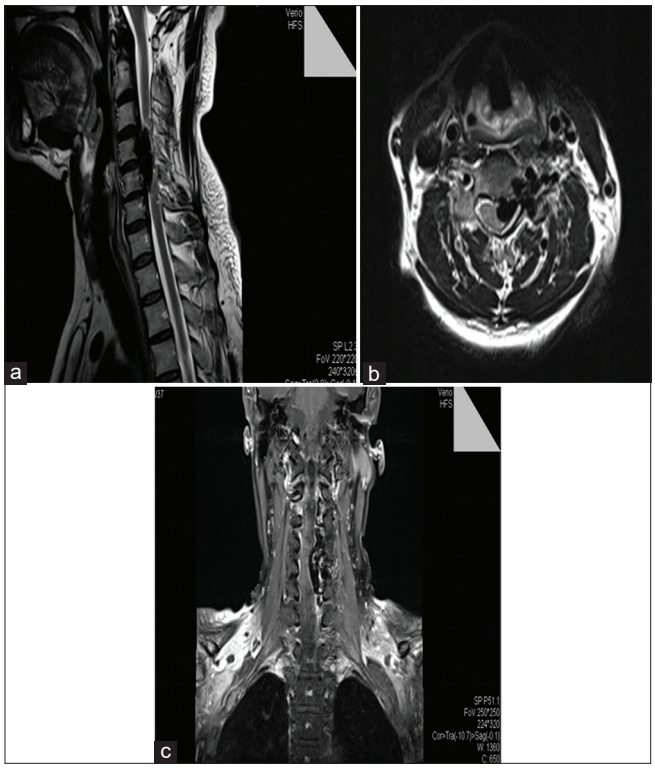

Figure 1: (a) Sagittal view of cervical spine magnetic resonance imaging showing large dilated epidural flow void. (b) Axial view of the cervical spine showing dilated epidural flow voids. (c) Coronal view of cervical spine showing dilated epidural flow voids

Figure 3: Venous phase of the intracranial injection of left vertebral injection showing drainage through the right transverse–sigmoid sinus

Headway Duo 167 (Microvention Terumo) microcatheter, though the thyrocervical and costocervical trunk, using mixture of lipiodol and N-butyl cyanoacrylate [Figure 7]. Postembolization angiogram revealed complete obliteration of the fistula and left vertebral intracranial injection revealed resumption of antegrade intracranial venous flow through the left transverse and sigmoid sinus, which was not present in preembolization stage [Figure 8].

Figure 2: (a) Digital subtraction angiography (DSA) (lateral view) of left vertebral artery injection showing a high flow arteriovenous fistula fed from the left vertebral artery (b) DSA AP view of the left vertebral artery showing a high flow arteriovenous fistula draining into the epidural venous plexus

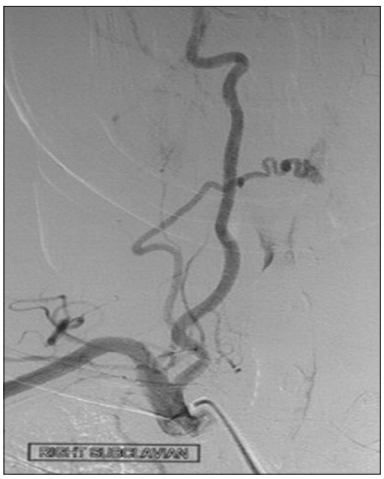

Figure 4: DSA of right subclavian artery showing right thyrocervical supplying the fistula

Outcome and follow-up

Post procedure, the patient had marked clinical improvement in his symptoms and complete resolution of the weakness and 1 month follow-up mRS score was zero. His follow-up MRI revealed complete collapse of dilated flow voids [Figure 9a and b].

Discussion

Spinal extradural AVF is a rare distinct clinical entity which needs to be identified and differentiated from more common seen spinal dural AVF. Spinal epidural AVF is excluded from the most common classification scheme of spinal AV shunts.[1]

Spetzler *et al*.,[2] in his modified classification system of spinal vascular malformation categorized AVF into extradural and intradural AVFs. They are typically supplied by radicular artery and drains into epidural venous plexus. Based on the venous drainage Takai and Taniguchi,[3] classified it into Type A – epidural AVF with and Type B – epidural AVF without intradural venous drainage. Our present case is type B spinal epidural AVF.

Figure 5: DSA showing the dilated thyrocervical and costocervical trunk supplying the fistula

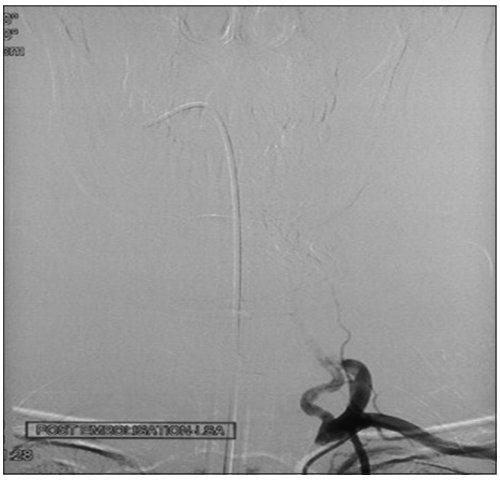

Figure 7: DSA following embolization of from the left thyrocervical and costocervical trunk

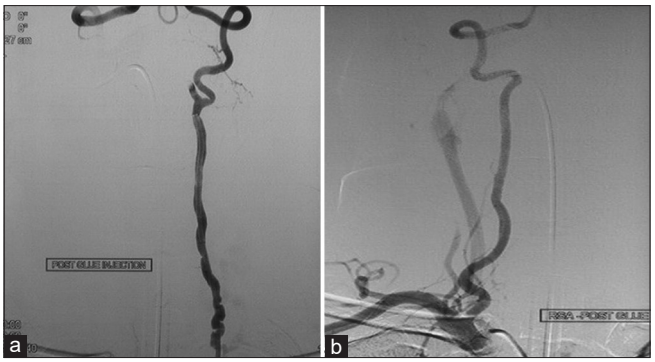

Figure 6: (a) DSA following embolization of the vertebral artery feeder. (b) DSA showing following embolization of the vertebral artery feeder collapse of supply from the right thyrocervical trunk

Figure 8: DSA showing venous phase of left vertebral intracranial injection following complete embolization. The left transverse and sigmoid sinus are now filling antegradely which was not seen in preembolization phase

Type B spinal epidural AVF are more common at the cervical level as in our case they are also associated with neurofibromatosis Type-1.

Takai and Taniguchi[3] in there literature review of spontaneous spinal epidural AVF analyzed 45 cases of spinal epidural AVF till 2011 out of which 23 were Type B.

These lesions are mostly symptomatic due to mass effect, occasionally due to hemorrhage. Recruitment of intradural veins and resultant venous hypertension can serve as a considerable source of morbidity.

Huang *et al*.[4] performed a pooled analysis of spinal epidural AVF in 2013 and identified 101 patients which also included post trauma and post spine surgery cases. In their analysis, 68% endovascular embolization while 32% underwent surgery. The obliteration rates were 87% for endovascular group and 92% for surgical group. These lesions require multiple session of embolization as there are multiple feeders, though in our case, we could achieve complete obliteration of the fistula in single session.

Following treatment, these patients showed marked improvement in clinical status, similar to what we observed in our case.

Asai *et al*.[5] reported a similar case where they used liquid embolic agent and coils to embolise the fistula which they surgically removed 1 week later. In 2015, Kasliwal *et al*.[6] had a similar case which was treated with endovascular method using coils and liquid embolic agent with stable results and clinical improvement.

To conclude, we reported a rare case of spinal epidural AVF with totally extradural venous drainage which was also causing intracranial venous hypertension. It was treated with endovascular embolization which remains the main stay of treatment in such cases following which excellent clinical recovery is expected. These cases need to be differentiated from other commonly encountered spinal AVF notably dural AVF. Complete obliteration should be target to avoid treatment failure or recurrence. The importance of understanding of vascular anatomy to avoid treatment failure and complications cannot be overemphasized.

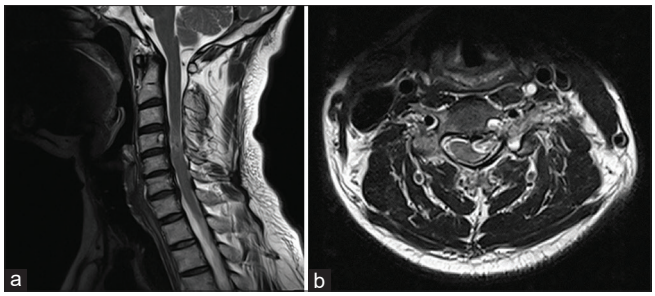

Figure 9: (a) Postembolization Magnetic resonance imaging showing complete collapse of the dilated flow void (b) Postembolization axial magnetic resonance imaging showing complete collapse of the dilated flow void with axial section showing glue cast in the flow voids

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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