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CASE REPORT

Cervical intraosseous arteriovenous malformation: report of a rare entity and its management dilemmas

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Abstract

Intraosseous occurrence of a spinal AVM is anecdotal, with only four such cases reported previously. This is the first report of a spinal intraosseous AVM in the cervical vertebrae. A 44-year-old male patient presented with a 2-month history of progressive quadriparesis and bladder dysfunction. Magnetic resonance imaging showed multiple flow voids within the C4 and C5 vertebral bodies, and an extradural component causing cord compression. CT showed extensive bony destruction at both levels. The diagnosis of an intraosseous AVM was confirmed with spinal angiography. The AVM was noted to be fed by branches from the ascending cervical arteries and the vertebral artery. The nidus was draining into the vertebral venous plexus and thence into the jugular vein through the marginal sinus. The patient underwent partial embolization of the AVM. Surgical resection was attempted but found to be unfeasible due to torrential bleeding. A 360-degree stabilization along with decompressive laminectomies was performed, resulting in clinical improvement and disease stabilization at one year follow-up. The case and its management dilemmas are discussed in light of a brief literature review.

Keywords Intraosseous · Spinal · Vertebral · Arteriovenous malformation

Introduction

Arteriovenous malformations (AVM) of the spine are unusual vascular pathologies, and pose a greater challenge to diagnose and manage compared to their intracranial counterparts. Since Di Chiro's first description of selective angiography for spinal AV shunts in 1971, the classification of spinal AVMs has undergone multiple modifications [7]. Amongst the various described AV shunts in these classifications, spinal AVMs are much less common than AV fistulae (AVF), and of them, 'paraspinal AV shunts' (with the shunt located outside the spinal canal) [8] are the rarest. Intraosseous occurrence of paraspinal AVMs is anecdotal, with only four such cases reported previously [1, 2, 4, 5]. This is the first report of an angiographically-characterized spinal intraosseous AVM within the vertebral bodies. The other unique features of the case were its cervical location, the extent and complexity of the pathology, and the dilemmas related to its management.

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Case report

A 44-year-old male patient presented with a 2-month history of progressive weakness of all four limbs, urgency of micturition and urge incontinence. He was wheel chair-bound at admission. His general physical examination was normal. Cardiovascular system evaluation was within normal limits. His neurological examination revealed spasticity in both the lower limbs, and diminished power (grade 2 to 3) in all four limbs. His deep tendon reflexes were exaggerated in the lower limbs, and he demonstrated bilateral extensor plantar responses. Magnetic resonance imaging (MRI) of the cervical spine (Fig. 1A and B) showed multiple flow voids within the C4 and C5 vertebral bodies extending anteriorly into the prevertebral space and posteriorly into the canal, causing significant compression and T2-weighted signal changes in the spinal cord. CT showed extensive lysis of the C4 and C5 vertebrae (Fig. 1C) as well as the pedicles and lateral masses on both sides (Fig. 1D). Spinal angiography confirmed the diagnosis of an intraosseous vertebral AVM at C5, C6 levels with feeders from bilateral ascending cervical artery branches (Fig. 2A and B) and vertebral artery (Fig. 2C). The nidus was draining into the vertebral venous

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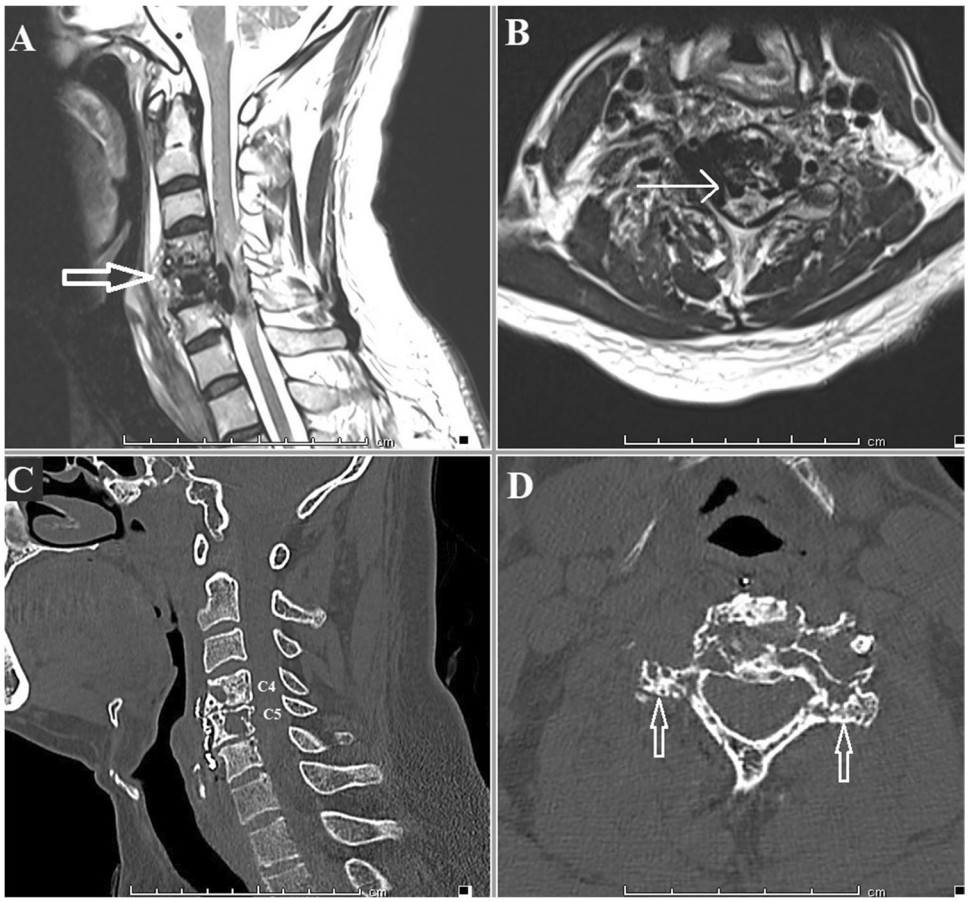

Fig. 1 Preoperative imaging **A)** T2-weighted sagittal showing multiple flow voids within the C4 and C5 vertebral bodies (arrow) extending anteriorly into the prevertebral space and posteriorly into the canal. Signal changes are noted within the spinal cord. **B** T2-weighted axial MRI sequence at C5 showing multiple flow voids in the vertebral body and an epidural component of the lesion (arrow) causing significant compression of the spinal cord. **C** CT cervical spine, sagittal section showing extensive lysis of the C4 and C5 vertebrae. **D** Axial CT section at C5 level showing destruction of the pedicles and lateral masses on both sides (arrows)

plexus (Fig. 2D) which was in turn draining into the jugular vein through the marginal sinus.

The patient was planned for embolization of the AVM followed by surgical resection and fixation of the involved spinal segments. While the larger feeders from the ascending cervical artery were successfully embolized bilaterally (Fig. 3A and B) using liquid embolic agent, smaller feeders of these vessels could not be cannulated. The feeders from the vertebral artery could not be embolized due to the risk of reflux of embolic material into the parent artery. The nidus was thus only partially embolized. Following this, he was taken for C4, C5 corpectomy and anterior cervical fixation. Intraoperatively, a leash of abnormal vessels were noted in the prevertebral area and within the longus coli bilaterally. A corpectomy was attempted, but had to be abandoned due to torrential bleeding from the nidus and abnormal vessels within the vertebral bodies that resulted in multiple episodes of hypotension. An in-situ C3-C6 anterior cervical plating was performed. This was followed by C4 to C6 decompressive laminectomies and C2 to T1 posterior fixation the following day (Fig. 3C). There was improvement in his neurological status after surgery, and he could walk with support at the time of discharge a week later. At a follow-up visit a year later, his neurological status had improved to normal. Repeat MRI (Fig. 3D) showed reduction in the flow voids at C4, C5 levels, and resolution of the epidural compression and signal changes in the spinal cord.

Discussion

Intraosseous AVMs account for less than 1% of all bony vascular lesions [3]. While there are multiple reports of this pathology occurring in the oro-maxillo-facial region and limbs, its occurrence in the spine is very unusual. Preoperative radiographic differentials of a spinal intraosseous AVM includes vascular osseous tumours such as aneurysmal bone cyst (ABC), hemangioma, osteoblastoma, giant cell tumour, chondrosarcoma, metastasis or myeloma. Since expansile or lytic changes on CT may be seen in many of these pathologies, the presence of specific tumour-defining characteristics should be closely looked for. For example, the presence of a 'fluid–fluid levels' in a multi-cystic lesion would favour a diagnosis of ABC, while the demonstration of flow voids in a lesion with a 'salt and pepper' appearance would be indicative of a hemangioma. Due to the rarity of a spinal intraosseous AVM, its preoperative diagnosis is elusive, and the diagnosis of an AVM is made only at surgery or on

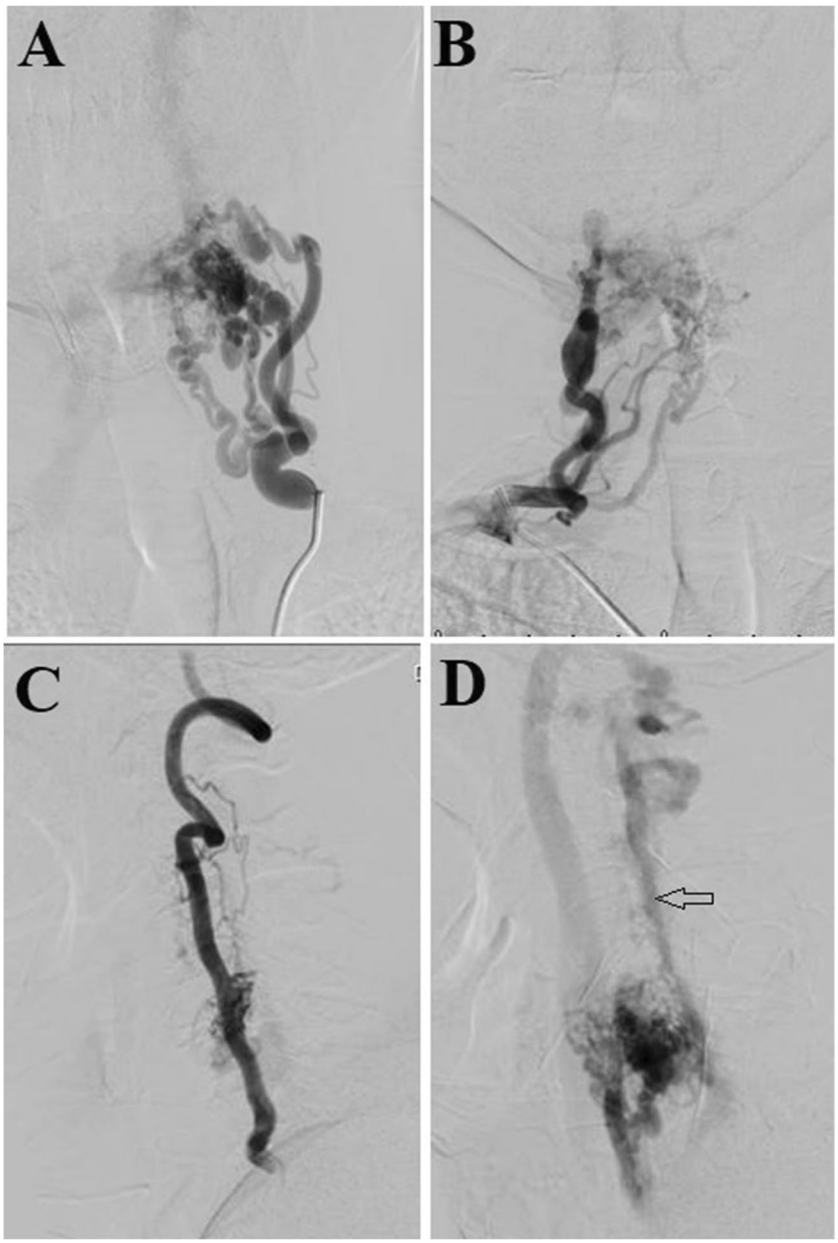

Fig. 2 Spinal DSA showing an intraosseous vertebral AVM at C5, C6 levels. **A** Feeders from the left ascending cervical artery, **B** right ascending cervical artery and **C**) vertebral artery. **D** The nidus was seen to be draining into the vertebral venous plexus (arrow) which was in turn draining into the jugular vein through the marginal sinus

histopathology [1, 2, 4, 5]. In our case, the diagnosis of an intraosseous AVM was clear from a combination of radiographic findings i.e. MRI detection of multiple flow voids in and around the vertebrae, CT demonstration of extensive bony lysis and angiographic confirmation of a nidus.

Paraspinal AVMs cause neurological symptoms due to venous congestion, cord compression by engorged draining veins mimicking a tumour, or due to perimedullary venous reflux causing an arterial steal phenomenon. Spinal instability may be seen in extensive lesions like ours with widespread bony destruction. In the case reported by Molina et al. [5], an intraosseous AVM in the L4 lamina resulted in a scoliotic deformity. Like in other spinal AVMs, symptoms of intraosseous AVMs are often insidious in onset and slowly progressive. Acute exacerbations may occur due to sudden changes in venous hemodynamics, or due to rupture of AVM-associated intranidal aneurysms. Given their rarity, the exact risk of bleeding from these AVMs is unclear at present.

Paraspinal AVFs or AVMs generally occur in the dorsal or lumbar regions [8], and a cervical location has not been reported. In three of the four published cases of spinal intraosseous AVMs, the lesion was located in the posterior spinal elements [1, 4, 5]. In the fourth case [2], the lesion was centred in the extradural compartment, and though the vertebra demonstrated signal changes, it had no demonstrable flow voids like in our case. The diagnosis of an intraosseous AVM was made retrospectively on histological examination

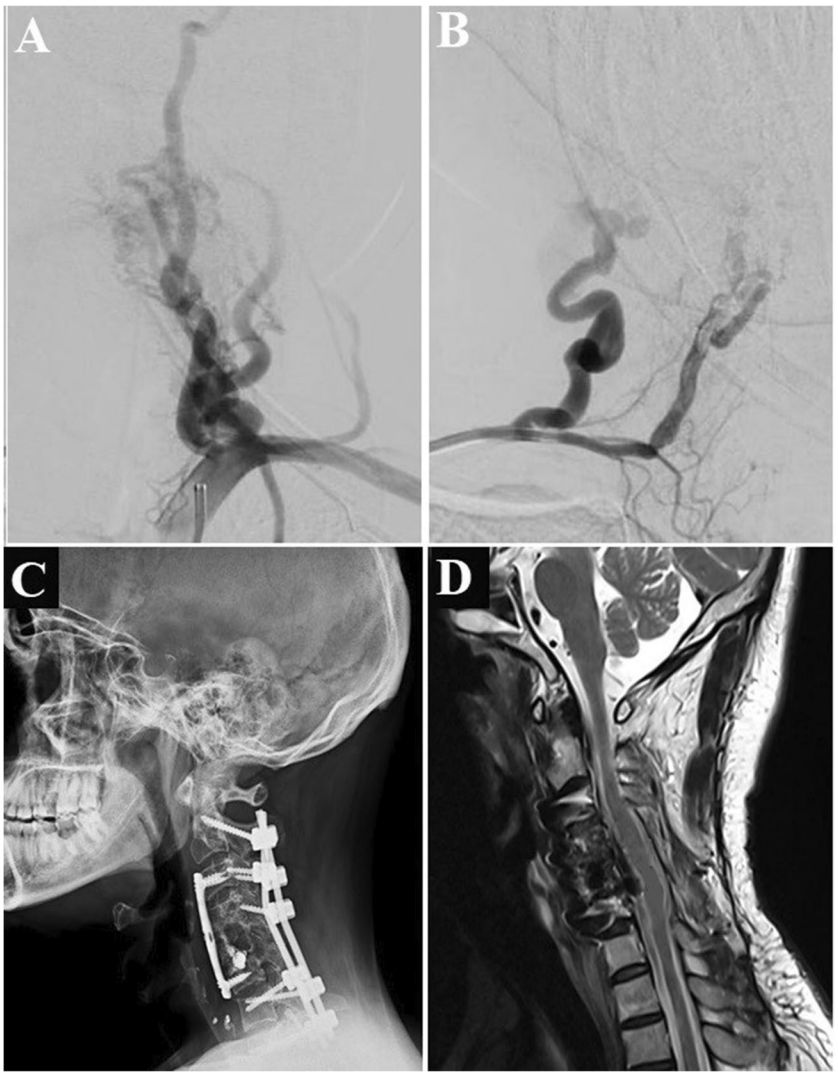

Fig. 3 Post-embolization DSA showing **A)** partial obliteration of the nidus with residual feeders noted from the left ascending cervical and vertebral arteries and **B)** complete obliteration of the right ascending cervical artery feeders. No nidus is visible in this injection. **C** Postoperative lateral radiograph showing C3-C6 anterior cervical plate fixation and posterior fixation with lateral mass and pedicle screws; **D** One-year follow-up sagittal T2-weighted MRI sequence showing reduction in the flow voids at C4, C5 levels, and resolution of the epidural compression and signal changes in the spinal cord

and a negative Wilms tumour staining of the resected specimen [2].

The optimal treatment of spinal intraosseous AVMs entails a multimodality approach with a combination of endovascular intervention, surgical resection with or without fixation, and radiosurgery. None of the previously reported four cases underwent preoperative embolization, and surgical resection of the lesions along with their epidural components was possible and proved curative. The dilemma in our case was that the AVM was not amenable to either complete embolization or resection. Only a partial embolization was possible due to the small calibre of some of the feeders, and inability to embolize the vertebral artery feeders due to inherent risk of parent artery occlusion. The tortuosity of the venous drainage precluded trans-venous embolization and usage of the 'pressure- cooker' technique [6] that reportedly achieve higher occlusion rates with lesser chances of ischemic complications. As a palliative measure, we performed a 360-degree stabilization of the spine along with decompressive laminectomies. This proved to be adequate in our case; not only did the patient improve clinically, but the follow-up MRI showed disease stabilization. However, he remains at potential risk of progression of the AVM with recruitment of new feeders over time. Further treatment options in such an inoperable, complex AVM could possibly include spinal radiosurgery or fractionated radiotherapy, similar to what has been described for other spinal AVMs and vascular spinal tumours [9].

Conclusion

This report describes a rare, vertebral intraosseous AVM in the cervical region. Due to its complex nature, the AVM was amenable only to partial embolization, and was found to be surgically unresectable. Partial treatment of the AVM resulted in neurological improvement and disease-stabilization at one year follow-up.

Author contributions Khurram Khan and Sumit Thakar contributed to the study conception and design. Material preparation and data collection were performed by Sumit Thakar, Tejus M.N. Rao and Vidyasagar Kanneganti. The first draft of the manuscript was written by Khurram Khan. The manuscript was edited by Sumit Thakar, Tejus M.N. Rao and Saritha Aryan. All authors read and approved the final manuscript. The study was supervised by Saritha Aryan.

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Code availability Not applicable.

Declarations

Ethics approval This being a retrospective study with de-identified data, ethical approval was not required as per the rules of the SSSIHMS, Bangalore Ethics Committee.

Consent to participate Informed consent was obtained from the participant included in the study.

Consent for publication The participant has consented to the submission of the case report to the journal.

Conflicts of interest/Competing interests All authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest or non-financial interest in the subject matter or materials discussed in this manuscript.

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