

Concomitant intramedullary arteriovenous malformation and a vertebral hemangioma of cervical spine discovered by a pathologic fracture during bicycle accident

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Abstract



Introduction Spinal intramedullary arteriovenous malformations are uncommon and a challenging type of neurosurgical entities. They are rarely located to cervical segment. On the other hand, although hemangiomas are relatively common bone tumors, cervical involvement is again rare and clinically significant ones are infrequent.

Case presentation A 14 year-old-male patient referred to an academic tertiary care unit and presented with neck pain and left hand weakness. Neurological examination revealed motor strength deficit at intrinsic muscles and

hyperesthesia at the left hand. Furthermore the pathological reflexes were positive on the left hand side. Imaging studies showed compression fracture, lytic changes resembling a hemangioma at C7 vertebra, and also an intramedullary vascular pathology at C5-6 level which was shown to be an intradural-intramedullary arteriovenous malformation (AVM) on digital subtraction angiography. Based on neurological and radiological findings, the decision was to treat the patient. After embolization of the AVM, the neurological condition of the patient deteriorated and immediate MRI scan of the cervical spine revealed edema of the spinal cord at the C5-6 level. Thus an emergent surgery was performed and C5-6-7 laminectomies with C5-T2 posterior fixation and arthrodesis were implemented. A second stage operation was carried out as C7 corpectomy with a distractable titanium cage 2 weeks after initial surgery. A follow-up evaluation at five years revealed 4/5 motor strength on his left intrinsic hand muscles and mild hyperactive deep tendon reflexes. Imaging studies at the postoperative period showed stable placement of the construct and no evidence of contrast enhancement at the C5-6 level inside the spinal cord.

Conclusion A rare case of multiple pathologies affecting the cervical spine, coincidentally diagnosed after a pathological fracture during a bicycle accident as vertebral hemangioma and intradural-intramedullary AVM that was successfully treated with early detection, have been presented. One should assess such patients under multidisciplinary fashion and treat on a case-by-case basis for achieving the best results in patient care.

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Keywords Arteriovenous malformation · Cervical spine · Endovascular treatment · Spinal reconstruction · Vertebral hemangioma

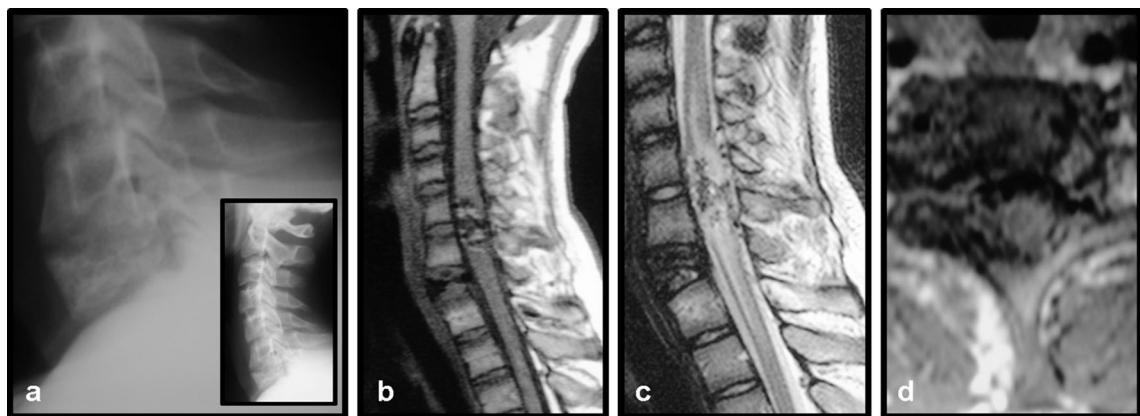


Fig. 1 Preoperative imaging studies of the patient. Plain lateral radiograph demonstrates destruction and loss of height at C7 (a) Sagittal T1 (b) T2 (c) and axial T1 (d) weighted cervical magnetic resonance imaging (MRI) studies show the pathological

compression fracture at C7 affecting the corpus and left pedicle, and the signal changes due to vascular abnormality; swelling and edema of the spinal cord at C5-6 levels, as well

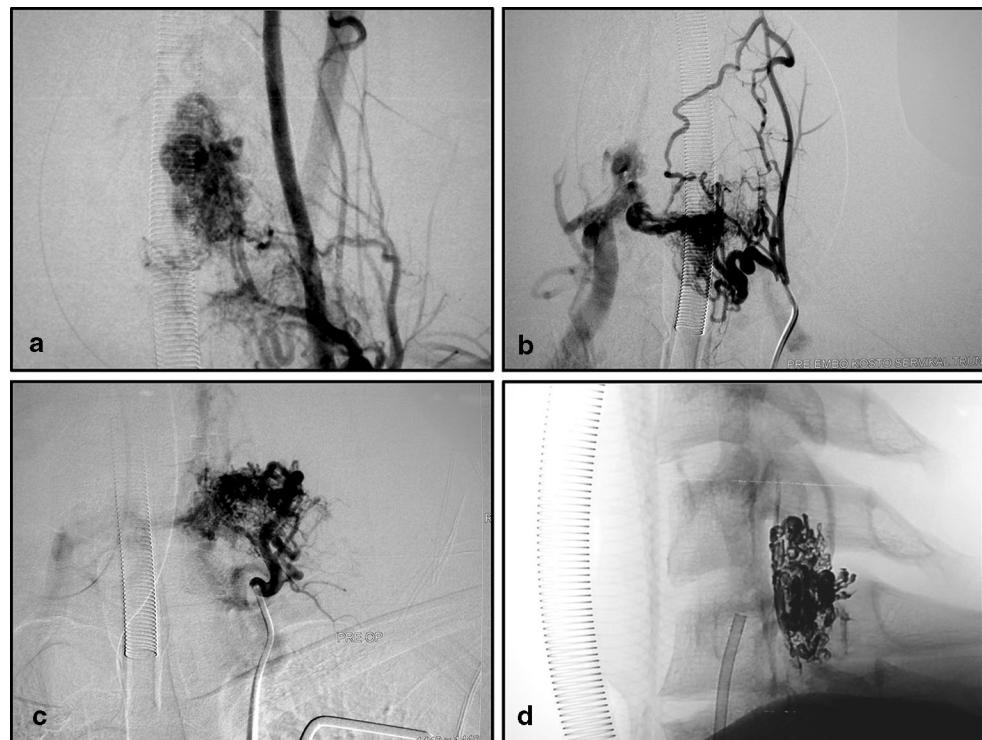


Fig. 2 The pre- and perioperative pictures taken during the embolization process. Left anterior oblique projection angiogram of the left vertebral artery revealing the feeding branches that are coming from just distal part of the left vertebral artery origin as well as costocervical trunk (a). Left anterior oblique projection digital subtraction angiography (DSA), selective catheterization of costocervical trunk demonstrating an enlarged tortuous feeding artery that is

supplying the caudal portion of the AVM (better visualized compared to Fig. 2a). There is also another serpiginous arising more cranially from the left deep cervical artery (b). A different angle view of left anterior oblique projection DSA showing stasis of contrast material just after application of onyx to the nidus (c). Non-contrast lateral angiogram showing the onyx cast after the procedure (d)

Case presentation

A 14 year-old-male was referred to an academic tertiary care unit with an initial diagnosis of C7 compression

fracture. His main complaints were neck pain and left hand weakness starting after a bicycle accident. However, the patient stated that his left hand was sluggish and he had some difficulty in grasping with left side over the last

several months. The neurological examination revealed 1/5 motor strength deficit at intrinsic hand muscles on the left side and hyperesthesia at C8 and T1 dermatomes. Deep tendon reflexes were hyperactive, the toe was upgoing and there was ++ clonus on the left side at the time of admission.

Diagnostic imaging section

Computerized tomography (CT) and magnetic resonance imaging (MRI) scans of the cervical spine revealed a pathologic compression fracture and lytic changes at C7 vertebra resembling a hemangioma; furthermore there were signal changes, swelling and edema of the cervical spinal cord coexisting with an intramedullary vascular pathology at C5-6 level, later shown to be a Type II arteriovenous malformation (AVM) with a large shunting volume, also classified as an intradural-intramedullary AVM according to Spetzler's modified classification of spinal cord vascular lesions by digital subtraction angiography (DSA) (Figs. 1, 2).

Historical review of the condition, epidemiology, diagnosis, pathology, differential diagnosis

AVMs of the spinal cord are considered to be developmental abnormality composed of clusters of blood vessels. The occurrence of vascular malformations of the spinal cord and dura are uncommon; compose 3–4 % of spinal cord masses, and are extremely rare in the cervical region [1, 2]. Out of 17 patients treated with Onyx from the series of Corkill et al. [1], only four cases found to be located at the cervical spinal cord. Recently, Velat et al. [3] reported 20 patients with glomus spinal AVMs, underwent micro-surgical pial resection within 17 years, half located at the cervical spine, being the largest series. The longest established and the most widely accepted classification categorizes spinal cord vascular malformations into the four categories based on the angioarchitecture and flow patterns as Type I to Type IV as following: Type I, dural arteriovenous fistulas (AVFs); Type II, intramedullary glomus AVMs; Type III, juvenile or combined AVMs; and Type IV, intradural perimedullary AVFs. Spetzler et al. have updated and proposed a modified system of classification in 2002, which divides the vascular lesions into three broad categories: Neoplasms, Aneurysms and Arteriovenous lesions. Arteriovenous lesions include AV fistulas and AV malformations with further subdivisions (Table 1) [2, 4]. The clinical findings of such lesions vary from pain, paresthesia, myelopathy and paresis to paralysis because of spinal cord compression, venous congestion and vascular

Table 1 Modified classification of spinal cord vascular malformations [4]

Classification of spinal cord vascular malformations

- I. Neoplastic vascular lesions
 - A. Hemangioblastoma
 - B. Cavernous malformation
- II. Spinal aneurysms
- III. Arteriovenous lesions
 - A. Arteriovenous fistulas
 - Extradural
 - Intradural
 - Ventral (small shunt/medium shunt/large shunt)
 - Dorsal (single feeder/multiple feeders)
 - B. Arteriovenous malformations
 - Extradural–intradural
 - Intradural
 - Intramedullary
 - Compact
 - Diffuse
 - Conus medullaris

steal [2, 3]; deterioration of spinal cord functions due to hemorrhage should also be considered [5]. To exclude metameric neurocutaneous syndromes which may contain AVMs in spine such as Cobb syndrome or Klippel-Trenaunay-Weber syndrome, is important while making the differential diagnosis. A small but important subset of patients with Klippel-Trenaunay-Weber syndrome may harbor AVMs of the spine [6]. Cobb syndrome is a neurocutaneous syndrome in which there are metameric cutaneous and spinal AVMs coexist [7]. In this patient, absence of any dermatomal cutaneous nevus and family history exclude these possibilities.

Hemangiomas are benign vascular neoplasms consisting of vascular spaces lined with endothelium. They are relatively common bone tumors with approximately 10 % vertebral involvement comprise the most common vertebral neoplasms, usually discovered incidentally. Frequently occur at the lower thoracic and upper lumbar spine, cervical involvement is rare with a reported incidence of 7 % [8–10]. The lesions are most often solitary, but they may be multiple in up to one third of cases. Spinal hemangiomas are usually localized to the vertebral body, less frequently extending into or exclusively affecting the posterior arch. The large majority of lesions are asymptomatic; clinically significant symptoms develop in only 0.9–1.2 % of patients. When symptoms occur, they can be vague and nonspecific. Vertebral collapse and epidural and/or extraosseous extension can result in back pain. Neural compression can produce paralysis and/or plegia or bladder and bowel dysfunction, whereas radicular symptoms occur

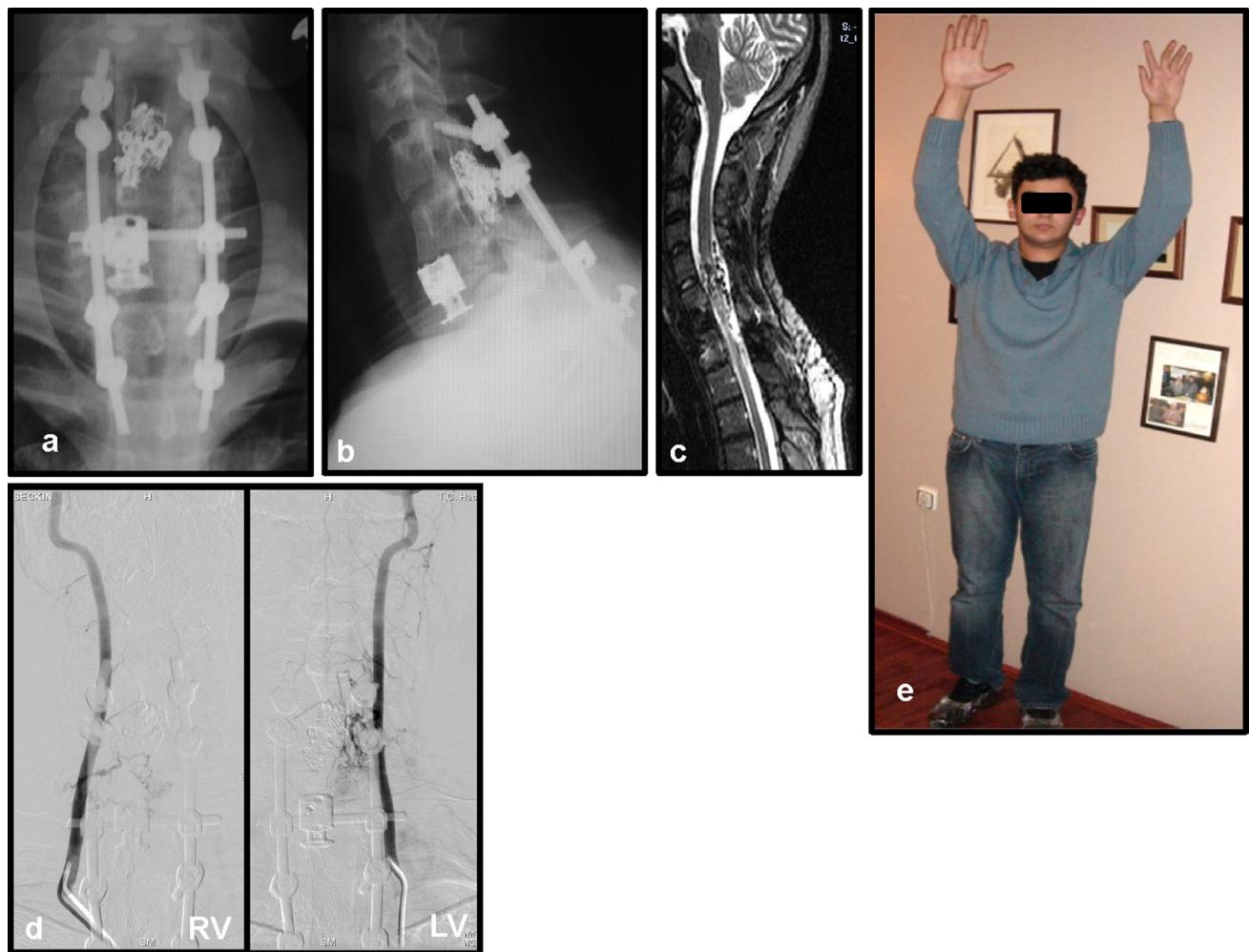


Fig. 3 Postoperative images and clinical picture of the patient. Anteroposterior (a) and lateral (b) radiograph depicting adequate positioning of the C7 titanium expandable cage after corpectomy and stable placement of C5–6-T1–2 posterior instrumentation. Radiopaque embolic substance can also be seen at C5–6 levels. Sagittal T2 weighted cervical MRI reveals glue cast and total resolution of

cervical spinal cord swelling and edema (e). Anteroposterior DSA views show completely occluded intramedullary AVM with perilesional angiogenesis and no sign of venous filling (RV right vertebral artery, LV left vertebral artery angiogram) (d). Clinical picture of the patient 5 years after treatment (e)

from nerve-root impingement [8, 10–14]. The differential diagnosis of vertebral hemangioma includes metastases, multiple myeloma, aneurysmal bone cyst, osteosarcoma, lymphoma and Paget's disease. The bony involvement and soft-tissue extension can be demonstrated by CT scan and MRI, respectively [11, 12].

All above, since we did not have any imaging studies before the accident, the differentiation of the cause of the intramedullary signal changes, edema and swelling of the spinal cord on MRI is difficult based on only imaging findings. However, progressive myelopathic signs mostly related to vascular steal and possible compression because of the spinal cord AVM; radicular findings in accordance with the coexisting vertebral hemangioma involving the corpus and left pedicle of the C7, might also be the cause of

the neck pain due to spinal instability, were the possible proposed mechanisms of such symptoms.

Rationale for treatment

Alone or in combination with each other; microsurgery, endovascular therapy, radiosurgery or radiation therapy are recommended treatment modalities for AVMs [1, 2, 15]. On the other hand, various treatment strategies and management algorithms for symptomatic vertebral hemangiomas are reported in the literature. Surgery (decompressive laminectomy ± stabilization; corpectomy or vertebrectomy + reconstruction and stabilization), transarterial embolization, vertebroplasty, ethanol injection and

radiotherapy should be performed according to the patients' symptoms, neurological status and radiological findings [8, 10, 11].

As was the case presented, given the progressive myeloradiculopathic findings secondary to intramedullary and osseous pathology; and likely instability due to the pathological fracture at cervical spine, the decision was to treat the patient rather than observation. Although microsurgical excision with or without embolization is recommended treatment modality for intradural intramedullary AVMs; this treatment may have relatively high treatment related morbidity [1] due to the deep seating and complex angiography and further co-existing vascular bony pathology. The authors tended to treat the dual pathologies endovascular and surgically. However the timing for the question what to do first is a matter of debate. The complex angioarchitecture of the lesion and possible related clinical findings resulted from vascular steal-large shunting volume- that was detected in DSA (Fig. 2), were thought to be the major causes of neurological deterioration and led the authors to prioritize the treatment of AVM. Although total occlusion may result in treatment related morbidity, the aim was complete obliteration of the AVM, if possible, due to the angiography to minimize-eliminate the risk of future bleeding [16]. As an embolic agent, ethylene vinyl alcohol copolymer, Onyx (Micro Therapeutics, Inc., Irvine, CA), was opted for the treatment of AVM due to its outstanding features over *n*-butyl cyanoacrylate (NBCA) such as controllable slow transmission, lower risk of both premature venous occlusion and a retained catheter [5]; as well as possible curative outcomes [15, 17], and also the endovascular team's high experience [18]. However the surgeons were ready for excision of the AVM in the case of complication or unsuccessful embolization, yet this might have risked major morbidity and caused immediate neurological deterioration requiring emergency surgery. For the cervical spinal osseous pathology, a surgical decompression and stabilization procedure was planned, posteriorly and anteriorly, respectively.

Procedure and procedure imaging section

After an extensive embolization of the AVM using the liquid embolic agent, Onyx (Micro Therapeutics, Inc., Irvine, CA), by interventional neuroradiology team (Fig. 2); the neurological condition of the patient deteriorated and neuro-examination revealed 3/5 hemiparesis on the left side. Immediate MRI scan of the cervical spine showed edema of the spinal cord at the C5-6 level. At this time, the goal of the surgical treatment by posterior approach was not only excise the bony tumor at C7, but also decompress the spinal cord at the edematous level,

emergently. For this purpose, C5-6-7 laminectomies and C5-6-T1-T2 posterior fixation and arthrodesis was performed. After two weeks from the initial surgery, second stage operation was carried out as C7 corpectomy preceded by transarterial embolization and followed by fixation/fusion with a distractable titanium cage. Then the patient was discharged with outpatient physical therapy referrals.

Outcome, follow-up

A follow-up evaluation at five years revealed 4/5 motor strength on his left intrinsic hand muscles and; mild hyperactive deep tendon reflexes, upgoing toe and + clonus also on the left side. Plain X-rays, MR images and DSA at the postoperative period showed stable placement of the construct without subsidence and fracture; resolution of cervical spinal cord swelling, edema, and no evidence of contrast enhancement at the C5-6 level inside the spinal cord; with totally occluded intradural intramedullary AVM along with peri-lesional angiogenesis and no sign of venous filling, respectively (Fig. 3).

Conflict of interest None.

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