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Case report

Giant intradural cervical spine arteriovenous malformations – A case and review of literature

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ABSTRACT

Introduction: Spinal arteriovenous malformations (SAVMs) are very rare and can be very challenging to treat since none of the therapeutic options does provide a definitive cure to these lesions. We believe that incorporation of intraoperative angiography during surgery in a hybrid theatre can help achieve a better cure.

Case presentation: We present a 45 years old woman with three (3) years history of weakness and ten (10) days' history of acute pain in right upper extremity. Magnetic resonance angiography (MRA) of the cervical segment of spinal cord revealed tortuous vascular masses from foramen magnum to the inferior margin of fourth cervical (C4) vertebral. Spinal digital subtracting angiography (DSA) confirmed vascular malformation at the cervical segment of the spinal cord with their origin from bilateral posterior spinal arteries. She was successfully operated on with the aid of intraoperative angiography without any neurological deficient.

Conclusion: Spinal angiography is the gold standard for all-inclusive assessment of SAVMs. Surgery and endovascular techniques equally have key therapeutic valves in treatment of SAVMs but a combination of the two gives a more accurate and reliable cure to this disorder.

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Abbreviations: AV, arteriovenous; AVF, arteriovenous fistulas; AVMs, arteriovenous malformations; C, cervical vertebral; CT-scan, computer tomographic scan; L, lumber vertebral; DSA, digital subtracting angiography; MRA, magnetic resonance angiography; MRI, magnetic resonance imaging; SAH, subarachnoid haemorrhage; SAVM, spinal arteriovenous malformations; SDAVF, spinal dural arteriovenous fistula; T, thoracic vertebral.

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1. Introduction

Spinal arteriovenous malformations (SAVM) are rare [1–4] and in conjunction with other spinal cord vascular disorders constitutes about 3–4%, and can be related to significant neurological disabilities and even death if not detected and treated early [1,5]. Cervical SAVMs are even more rarer and so far, only a hand full of literature have reported arteriovenous malformations (AVMs) in this location. Cervical SAVMs are very challenging to detect since their clinical course imitate a diverse range of neurological conditions [6]. They can be grouped into high-flow and slow-flow base on their pathophysiology. While high-flow lesions may lead to ischaemia or haemorrhage, slow flow on the other hand may lead to venous congestion, compression of spinal cord and ischaemia [7]. Based on their location in the cord, angioarchitecture, intradural or extradural and/or presence or absence of arteriovenous (AV) shunts, SAVMs can be categorized into four (4) types [7,8]. SAVMs are curable disorders [9] and recent improvements in neuro-imaging and endovascular techniques outlining the angioarchitecture of spinal vascular anatomy have led to enhanced treatment of this disorders [10]. We report a case of giant intradural cervical SAVM which we successfully treated by incorporates both open surgery and intraoperative angiography without any neurological deficient.

2. Case report

We present a 45 years old woman with three (3) years history of weakness and ten (10) days' history of acute pain in right upper extremity. She was apparently diagnosed 3 years ago with spinal vascular malformations in a local hospital with no treatment. She was visiting the local hospital for pain relievers intermittently until ten (10) days prior to presentation at our facility when her condition aggravated. The weakness and pain at right upper limb radiated to the neck with persistent needle-like pain (Paresthesia). She however has no numbness as well as incontinence. Her bladder and bowel habits have not changed. She was put on Gabapentin capsules for the pain relieve. Past medical and surgical history was unremarkable.

General physical examination did not yield much. All the systems were grossly normal. Neurological examination revealed intact cranial nerves. On the upper limbs, the right arm had corresponding weakness of 3/5 proximal strength and 4/5 strength in his distal muscles while the left arm had corresponding weakness of 4/5 proximal strength and 4/5 strength in his distal muscles. Corresponding dermatomes were normal. She had brisk and bizarre sensations to pin prick, cold and hot stimuli on the upper arms but normal in lower limbs. Proprioception and vibratory sense were markedly reduced in the upper arm but normal in the lower limbs. Abdominal reflexes were absent and rectal tone intact. Her low limbs however had bilateral corresponding 5/5 proximal strength and 5/5 strength in his distal muscles. She could walk about with any form of support. All reflexes were present and normal. Routine laboratory and other ancillary investigation were normal.

Magnetic resonance angiography (MRA) of the cervical segment of spinal cord revealed tortuous vascular masses from foramen magnum to the inferior margin of C4 vertebral. The lesions were significantly heterogeneously enhanced. The spinal cord significantly compressed the masses, so parts of the spinal cord are not visible. The vertebra disc from C2–C7 is somehow collapsed with posterior protrusion of the edges of vertebral bodies resulting in the relative compression of the dura sac. On T2-MRI, cervical intervertebral discs have iso to hyper-intensity. There are no para-spinal soft tissue swellings. A working diagnosis of intra-spinal vascular malformation from foramen magnum to the inferior margin of C4 vertebral was made (Fig. 1A and B). Spinal digital subtracting angiography (DSA) was done at the local hospital and repeated at our facility showed vascular malformation at the cervical segment of the spinal cord with their origin from bilateral posterior spinal arteries (Fig. 2A–D). After educating and counselling the patient as well as her relatives, operation was scheduled.

The operation was done in our hybrid surgical theatre that incorporates both open surgery and intraoperative angiography. The aim of our surgery was to excise this giant vascular malformation in cervical vertebral canal with selected angioplasty, decompression of vertebral canal and spinal nerve root as well as fixation of fused posterior C2–C6. The patient was put on the prone position with the head rested on the doughnut head support after general anaesthesia. The skin marking was drawn from mid occipital hairline to about C6 vertebral. The endovascular surgeon also draped the left inguinal region and cannulated the left femora artery. Draping of the neck was done after securing the artery and the cannula inserted.

The skin incision was made and the subcutaneous, intramuscular, along the C2–C7 was dissected carefully. The para-spinal muscles were separated from the spinous processes at both sides followed by exposition of the lower part of the occiput at the foramen magnum, the atlas arch C2–C7 removed and laminectomy done. The dura was opened and we saw huge tortuous intradural vascular malformation that were more localized to the left side and extended from C1–C5. These abnormal vascular masses were originating from both posterior spinal arteries. They cause enlargement of the spinal canal with extensive adhesions and compressing of nerve roots as well as deformation (Fig. 3A).

Intraoperative angiography was used to identify the feeding arteries intra-operatively and they were resected with no complication. The microscopy was used throughout the operation. After total resection of the lesion, intraoperative angiography was done again which confirm total resection of the lesions. Intraoperative images also show total resection (Fig. 3B and C). After securing total hemostasis within spinal cord, the dura was repaired with artificial dura mater and posterior C2–C4 fixation done with titanium screws. A drainage tube was inserted and suturing of muscle, fascia, as well as subcutaneous tissue and skin. (The resected SAVM is as shown in Fig. 3D.) The patient recovered very quickly with no neurological deficits and the pain at the left arm was improved. Postoperative MRI was done and this revealed no residual SAVM (Fig. 4A and B). She was put on a rigid cervical colour for six (6) months. She was discharged home two (2) weeks after the operation and schedule outpatient visits arranged every three (3) months.

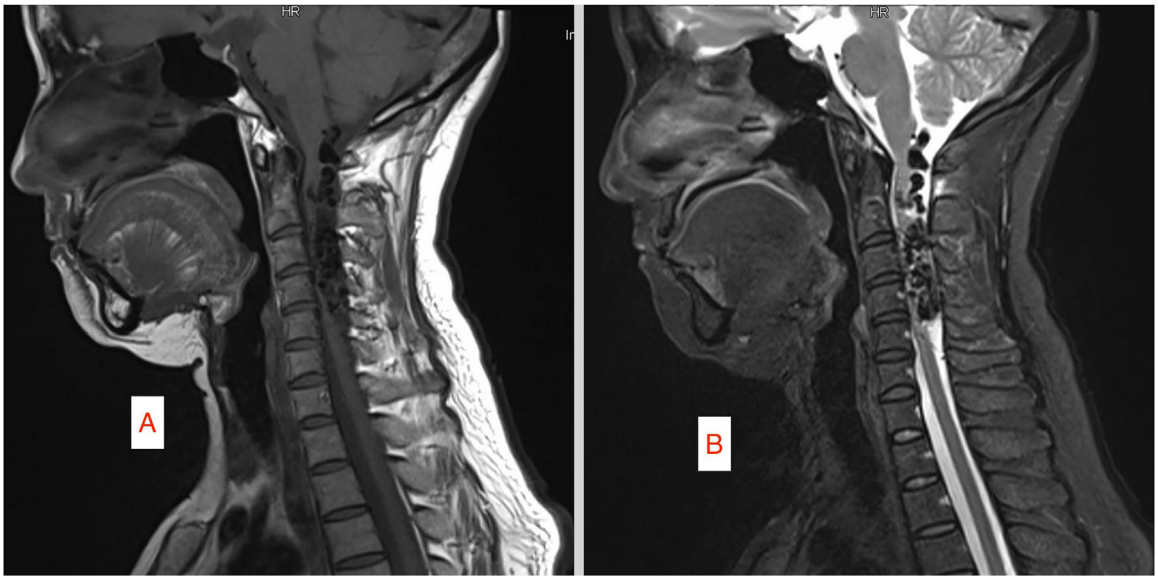

Fig. 1 – A and B are T1 and T2 MRI respectively showing the spinal arteriovenous malformation (SAVM).

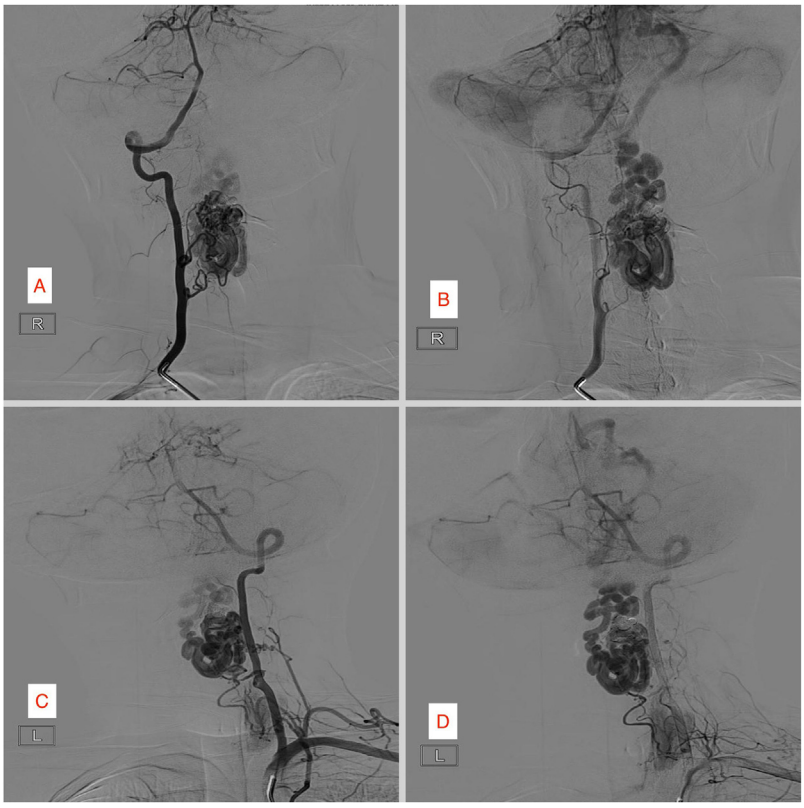

Fig. 2 – A, B, C and D are digital subtracting angiogram (DSA) images giant spinal arteriovenous malformation (SAVM).

3. Discussion

Spinal arteriovenous malformations (SAVM) are rare [1–4] and in conjunction with other spinal cord vascular disorders constitutes about 3–4%, and can be related to significant neurological disabilities and even death if not detected and treated early [1,5]. They can be grouped into high-flow and slow-flow base on their pathophysiology. While high-flow

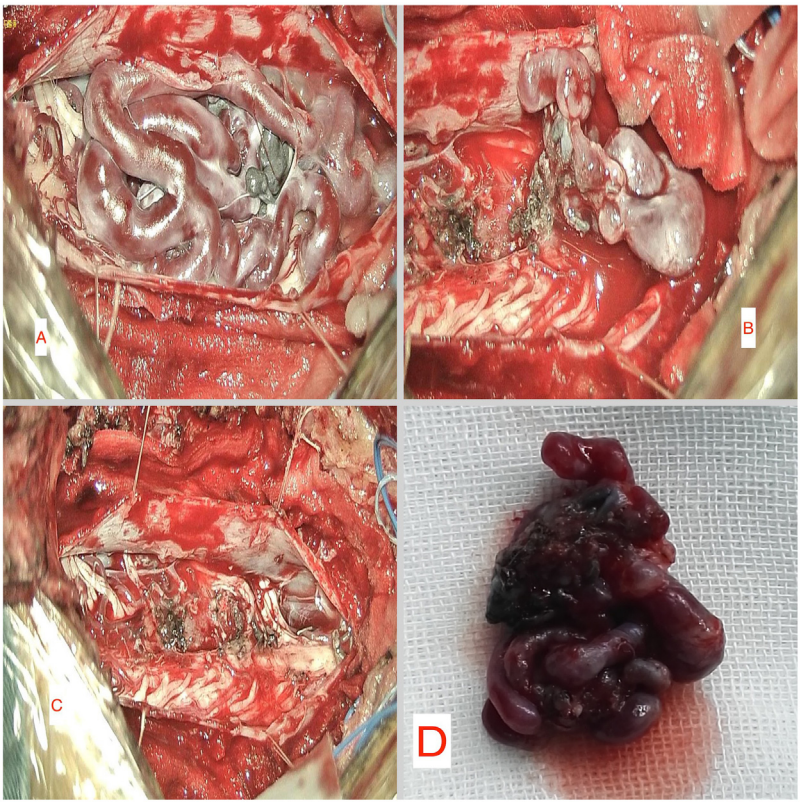

Fig. 3 – A, B, C and D are intraoperative images. A shows the lesion before resection, B shows the lesion partially resected, C shows total resection of the lesion and D shows the resected AVM.

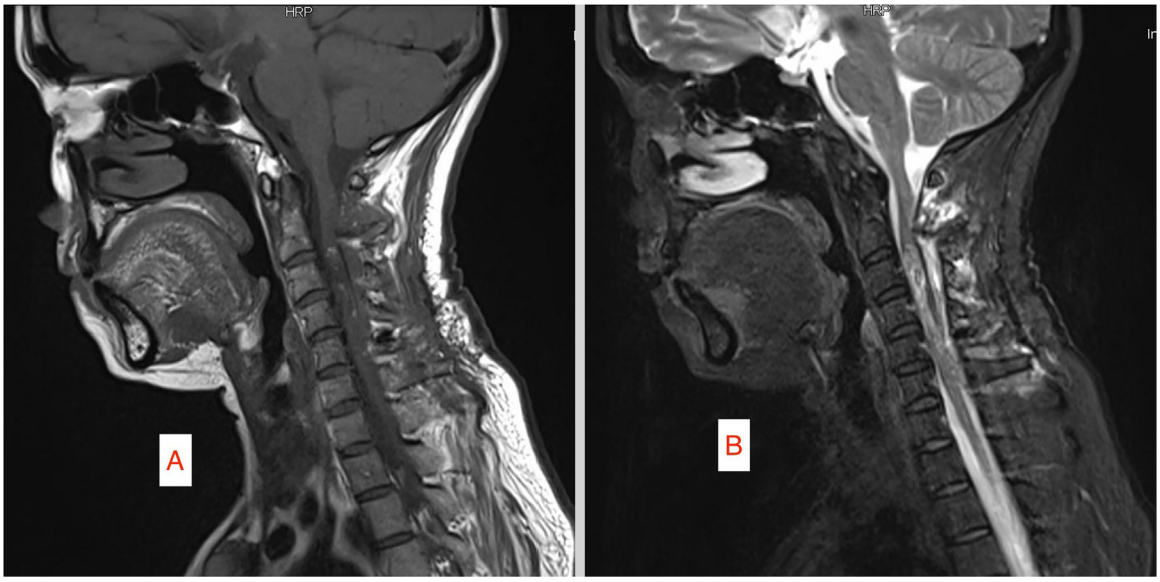

Fig. 4 – A and B are postoperative T1 and T2 MRI respectively indicating no residual AVM.

lesions may lead to ischaemia or haemorrhage, slow flow on the other hand may lead to venous congestion, compression of spinal cord and ischaemia [7]. Our case is much more interesting because of the size of the lesions and fair of neurological defect after treatment. With the aid of our hybrid technique of incorporating both open surgery and intraoperative angiography, we successfully treated this case without any neurological deficient.

The management of SAVMs depends largely on the understanding of the vascular anatomy of the spine. The

spinal vasculature consists of anterior and posterior arterial networks. The anterior spinal artery runs beside the anterior median fissure and make-up the anterior arterial plexus. The sulcal branch of arteries originates from the anterior spinal artery and supplies the anterior horns, corticospinal tracts as well as the spinothalamic tracts [1,11]. The posterior arterial structure comprises of plexiform collaterals connecting the two posterior spinal arteries and they supply the posterior third of the spinal cord as well as the dorsal columns. In the cervical region, the medullary arteries originate from the vertebral arteries and divisions of the thyrocervical trunk with about 6–10 of them feeding the anterior and posterior spinal arteries in adults [1,11]. On the other hand, these medullary arteries originate from the intercostal arteries, which are divisions of the aorta and iliac arteries at the thoracic and lumbar region. Among these medullary arteries, Adamkiewicz is the largest and classically arises from the left side to supply the spinal cord between T8 and L2 [1,11]. The sulcal and radial veins constitutes the spinal cord venous system and they drain into the coronal venous plexus on the cord surface. Medullar veins are exclusive in the sense that they are valveless and drains the pial venous plexus into the epidural (Batson's) venous plexus [1,11].

Rosenblum et al. indicated that intradural SAVMs should come to mind when a patient symptoms affects the arm, associated with acute unset of subarachnoid haemorrhage (SAH) with spinal bruit and the he or she is less than 30 years [3]. Although our patient systems affected the arm they were however no associated SAH and her was more than 30 years. The most classical history of intradural cervical SAVM paresis, sensory abnormalities, sphincter disturbances, and pain although dura cervical SAVM may also mimic this presentation [3].

On magnetic resonance imaging (MRI), SAVMs are seen as aberrantly compact, frequently tortuous vessels with angioma-like nidus on the surface of the spinal cord. MRI aids in disguising spinal dural arteriovenous fistula (SDAVF) and intramedullary SAVMs. Contrast-enhanced MRI is now the most accurate noninvasive imaging method of choice in making diagnosis of SAVMs, assessment of the cord enclosing the lesions as well as distinguish areas of acute or chronic hematomyelia [12]. Furthermore, spinal angiography provides more precise features of the arterial feeders, angioarchitecture of the SAVM nidus as well as venous drainage. However, MRA is not able to specifically identify smaller size feeding spinal arteries [12]. Therefore, spinal angiography is the gold-standard in making diagnosis of SAVMs since it able to differentiate other vascular abnormalities as well especially acute spinal cord ischaemia [1,12,13]. In situations where the SAVM is associated with SDAVF, myelography and post-myelographic CT-scan should certainly be done after MRI. Moreover, in cases of low-flow fistulas, myelography is extra sensitive in revealing pathologic perimedullary vessels than MRI [12,14]. SAVM characteristically seen on myelography as a ''bag of worms'' and broadening of the spinal cord. Furthermore, post-myelography CT-scan is much more useful in identifying the malformations inside or outside the spinal cord and assessing the local bony changes linked with the persistent compression by the enormous tortuous dilated vessels [1].

Based on their location in the cord, angioarchitecture, intradural or extradural and/or presence or absence of AV-shunts, SAVMs can be categorized into four (4) types. This was proposed by Ansen and Spetzler in 1992 [8,15]. Type 1 is usually confirmed to dura of nerve sheath in the thoracolumbar region and constitutes about 60% of all SAVMs including spinal arteriovenous fistulas (AVF). Patients are usually males in their fifth and eighth decades of life and become symptomatic because the fistulae result in venous congestion, hypertension, hypoperfusion, oedema as well as hypoxia. Other presentation includes progressive spinal cord ischaemia and seldomly haemorrhage [8,16]. Type 2 or Glomus SAVMs on the hand are confined in the intramedullary at cervicothoracic junction, consisting of tightly packed nidus of dysmorphic arteries inside a short segment of the cord and constitutes about 20% of all SAVMs with mortality of 17.6%. Patients are usually younger and become symptomatic because of mass effect that lead to myelopathy or radiculopathy and occasional haemorrhage [8,17–19]. Type 3 or Juvenile SAVMs are very rare and originate may have single or multiple location which can be intradural and/or extradural. They are usually having intramedullary nidus the may beset the whole spinal canal at the level of the lesion [8,20]. Type 4 is usually periampullary AVF [8,15,21]. Our case although falls into type 2 does not fit well because of the age and clinical presentation.

The main objectives of treating SAVM are to reduction the chances of haemorrhage and prevent the progression of neurologic deficits [1,22]. It has been shown that decreasing of the arterial flow within a SAVM can reverse neurologic deficit. The precise blood supply to the malformation is frequently challenging to determine although the lesion may be supplied by a single or a few arterial feeders due to the high-flow shunts [1]. Furthermore, many of the arterial feeders may have an ancillary blood supply to the SAVM. Surgical or endovascular occlusion of proximal arterial feeders often do not completely treatment the malformation, but rather make management more challenging due to non-occlude the SAVM nidus [1]. Additionally, surgical management of an intramedullary SAVM usually lead to major impediments. On the other hand, embolization has proven be the main therapy of choice for intramedullary SAVMs, both as primary therapy and as an adjunct to surgical excision. Interestingly, fractional embolization has proven to be of good valve to these disorders [1,23]. The most ideal agent for endovascular treatment is liquid embolization agents. Liquid embolic agents like cyanoacrylate and Onyx attain a more permanent occlusion with extremely little recanalization rate, but has a simultaneous risk of involuntary embolization of the collaterals branches that are not seen on angiography [1]. Particle embolization is simpler to carryout, but has severe drawbacks due to recanalization that may necessitate several re-embolization [1]. The choice of a particulate therapeutic agent above may be considered a palliative therapy and not conclusive because it does not total ameliorates the signs and symptoms of the disorder but rather improves the natural history as well as offer a good clinical outcome [1].

4. Conclusions

Spinal angiography is the gold standard for all-inclusive assessment of SAVMs. Surgery and endovascular techniques

equally have key therapeutic valves in treatment of SAVMs but a combination of the two gives a more accurate and reliable cure to this disorder. We therefore suggest that neurosurgeon who choices the open surgical option in the treatment of SAVM incorporates intraoperative angiography to be sure that they accurately and total resect the lesions.

Authors' contributions

All the authors contributed equally to the manuscript design and writing.

Ethics approval and consent to participate

The ethical committee of the hospital full approved our case study. The patient was informed about our intension to involve her in a case study and she agreed to partake in the study. She signed the concern form before the operation was carried out according to all surgical protocols.

Consent for publication

The patient was dually informed about our intention to publish her case and she fully concerted to the use of these documents. The hospital also concerted to the use of this information for publication.

Availability of data and material

The folder or data is the property of the hospital and readily available with proper concern but for confidentiality of the patient the folder cannot to release to third party.

Conflict of interest

None declared.

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