Spinal Arteriovenous Malformation: A Case Report and Review of Literature

Abstract

Spinal arteriovenous malformations are rare vascular anomalies within the paediatric age group. These anomalies are associated with devastating consequences and require prompt management to prevent the long-term neurological sequelae.

We report a case of a 10-year-old boy with tetraparesis secondary to spinal arterio- venous malformation type III (Juvenile AVM) with rapidly deteriorating neurological signs who had to be managed conservatively due to lack of advanced neurosurgical facilities and interventional radiological services in our facility and sub region.

Keywords: *Arteriovenous, conservative, deteriorating, juvenile, malformations, spinal, tetraparesis*

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Introduction

Spinal arteriovenous malformations (AVM) in the paediatric population are rare vascular malformations with devastating consequences from the pathology as an entity as well from poor management.[1,2] It requires prompt diagnosis and institution of the appropriate management to forestall the long term neurologic sequelae.[3-5]

We report a case of haemorrhagic type III or juvenile spinal AVM in a 10-year-old boy with rapidly progressing neurologic sequelae who was managed conservatively due to unavailability of advanced neurosurgical care in our facility.

Case

A 10- year- old male admitted with a day's history of upper back pain, inability to walk and pass urine, following an alleged assault by playmates at school three days prior to presentation.

On examination, patient was conscious and alert, oriented to time, place and person. Spine examination was unremarkable. Patient had normal grade of power in the upper limbs however, it was zero for all muscle groups in the lower limb. The Sensory level was at T1; reflexes were normal in the upper limbs, but absent in the lower limbs. The Bulbocavernosus reflex was absent. Anal sphincter tone was decreased. There was a palpable suprapubic mass with features suggestive of a full bladder.

Based on the presenting symptoms and examination findings, a diagnoses of cervical spine injury with tetraparesis (ASIA type B) was made. An MRI with contrast was ordered [Figure 1] and it showed an ill-defined serpentine flow noted in the substance of the cervical cord at the level of C4 vertebral body with associated extra-medullary tortuous feeding vessels. A large extra medullary vessel at T2 vertebrae with compression and displacement of the cord. There were areas of high signal intensity within the extramedullary and intramedullary component at T1 and T2 suggestive of a haemorrhage; as well as features suggestive of spinal arterio- venous malformation type III (Juvenile AVM).

On the second day of admission, his neurological status deteriorated with sensory level limited to C7 and motor function limited to C7 in the upper limbs. They were counselled for advanced neurosurgical intervention abroad but declined due to financial constraints.

He was cared for by a multidisciplinary team, with the parents counseled on the prognosis of his condition.

Discussion

Spinal vascular malformations have been noted to be rare and associated with significant heterogeneity within the

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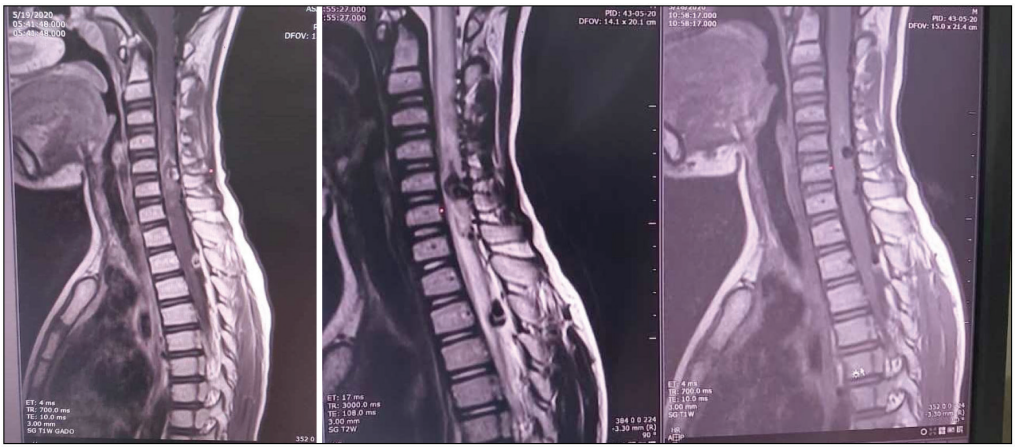

Figure 1: Showing sagittal views of MRI of cervical portion of the spinal cord

paediatric population characterized by shunting of blood from the arteries to veins in an abnormal capillary bed.[6] These vascular malformations tend to occur early in life.[7]

The classification developed by Anson and Spetzler is widely used to characterize the lesion into four types.[7] Type III AVM are regarded as rare and tend to be aggressive at presentation.[8] The index case had radiological features suggestive of the juvenile type and falls within the classical age group.

The presentation of spinal AVM comprises motor and organ dysfunction ranging from back pain, progressive muscle weakness, bladder and bowel dysfunction, sensory deficits to difficulty in walking all typified in the patient.[3-5] Similarly Patchana *et al.* and Thiele *et al.* reported motor and organ dysfunction.[1,9] It has been postulated that the neurological sequelae may be as a result of haemorrhage, mass effect, venous congestion or redistribution of blood.[3,10] Haemorrhage was identified in the index case and possibly the cause of the neurologic deficits.

The preliminary investigation of choice in the evaluation of AVM is the magnetic resonance imaging which in our case was done.[3,11-15] In preparation for intervention, digital subtraction angiography is regarded as the gold standard in characterizing the lesion and planning treatment.[1]

The cervico-thoracic location in this case varies from common sites of AVM reported in literature.[16] González described the lower thoracic and thoracolumbar regions as the common site of occurrence.[16] Rodesch *et al.* noted either the cervical or thoracic locations without extensions between the two regions of the spinal cord.[17] The presentation of tetraparesis from AVM associated with abnormal cord features spanning from C2 to T4 ultimately worsens the prognosis. Rapid deterioration in the neurological status from ASIA B to ASIA A was very notable in this case comparable data in existing literature and even worse after a rebleed.[18-22]

Treatment of AVM involves the endovascular techniques with embolization, surgery, combination of both or conservative management depending on the morphology of the lesion.[3,5,10,23-25] This case was managed conservatively with a multidisciplinary approach to tackle the various deficits due to unavailability of funds as well as highly resourced neurosurgical facilities or interventional radiological services hampering the possibility of attempting intervention in our sub region.[26,27]

Conclusion

Juvenile Spinal AVM is a rare vascular malformation associated with significant morbidity and requires timely intervention. The emergence of such a rare pathology in our setting underscores the need to establish advance neurosurgical facilities and interventional radiological services to treat and mitigate the devastating neurological sequelae.

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Conflict of interest

There are no conflicts of interest.

Ethical consideration

Consent was sought from guardians and patient.

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