Clinical Case Reports

ISSN: 2675-3626

Spinal vascular malformation in a 12 years old child. Diagnostic challenges and management strategies adopting a multidisciplinary approach.

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Background: Spinal vascular malformations (SVM) are a complex entity and rarely noted in children. They are commonly seen in the elderly with a predilection for the thoraco-lumbar region. The incidence varies from 3-11% and varies across all age groups. The presenting features in the initial stages are paraplegia, abnormal gait, paraesthesia, radicular pain and diffuse sensory symptoms. If not detected early they can lead to further spinal cord compression and cause urinary retention, bladder, bowel incontinence or erectile dysfunction. Despite the advances in diagnosis and treatment, the complexity of the pathology makes treatment challenging in a child. Spinal arteriovenous metameric syndrome (SAMS) is an extremely rare, non inherited neuro cutaneous condition characterised by a SVM and a vascular skin lesion affecting the corresponding dermatome as we saw in our patient.

Case presentation: We present a 12 years old female child who presented with lower limb weakness, diminished reflexes and urinary retention. Examination of her back revealed a pigmented nevus in her lumbar paraspinal region which was communicating with the SVM in the same metameric segment. Nerve conduction velocity was suggestive of motor axonal neuropathy. Magnetic resonance imaging of spine was suggestive of paraspinal vascular malformation. She has recovered after surgical intervention and embolization.

Conclusion: SAMS is extremely rare in children with very few reported cases. A multi-disciplinary approach beginning with a detailed history taking and clinical examination is invaluable in such complicated cases to deal with the various aspects of a complex pathology

Keywords: spinal vascular malformation, spinal arteriovenous metameric syndrome (SAMS), embolization

INTRODUCTION

Spinal vascular malformations (SVM) are a complex entity and rarely noted in children [1-2]. The incidence is 3-11% and varies across all age groups [3]. The lower thoracic cord is the most common site of SVM [3]. However the cervical region can also be involved. If not detected early, they can lead to spinal cord compression and cause urinary retention, bladder, bowel incontinence or erectile dysfunction. Despite the advances in diagnosis and treatment, the complexity of the pathology makes treatment challenging in a child.

We present a 12 years old female child who was diagnosed to have paraspinal vascular malformation and has recovered after surgical intervention and embolization. Our patient was eventually diagnosed as Spinal arteriovenous metameric syndrome (SAMS) which is an extremely rare, non inherited neuro cutaneous condition characterised by a SVM and a vascular skin lesion affecting the corresponding dermatome. This was first described by Cobb in 1915 as 'hemangiomata' of the spine associated with a skin nevi and was known as Cobb syndrome. Literature review reveals the number of cases to be less than 100. Actual incidence of SAMS may be higher as only symptomatic cases have been documented [4-7].

CASE REPORT

A 12 years old female child presented to the out-patient department with a history of on and off back pain for one month. There was no history of fall and trauma prior to this episode. She complained of sudden onset bilateral lower limb weakness for 3 days along with difficulty in walking due to weakness and pain in her lower limbs. Child was having urinary retention for 2 days. On examination her lower limbs were found to have hypotonia with diminished deep tendon reflexes. She had grade 2/5 power in her hips and 1/5 in her knees and ankles. Her tone, power and reflexes in the upper limbs were normal. There was no evidence of breathing

Received: 31 October 2022
Accepted: 06 May 2023
Published: 16 May 2023

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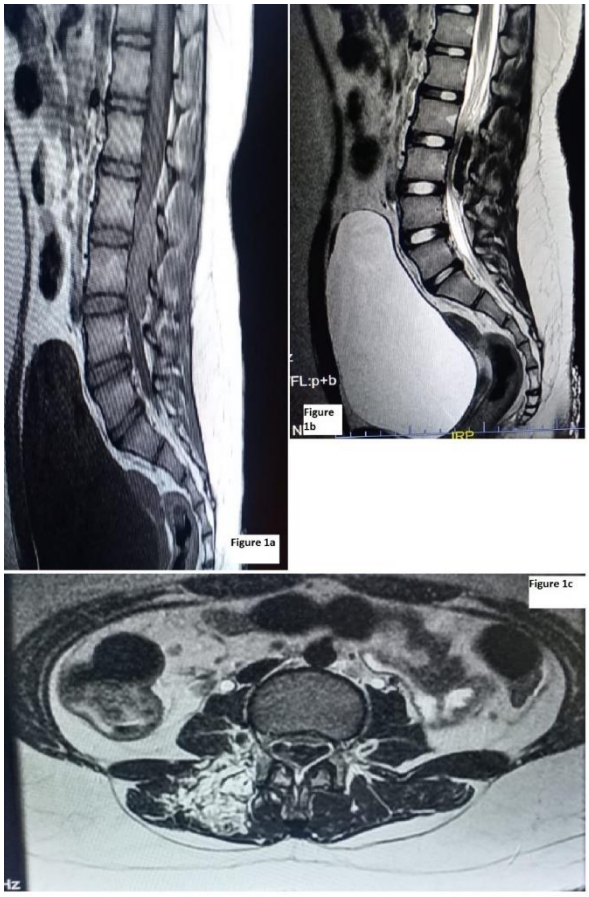


Figure 1a, 1b – Sagittal T1W and T2W MR images showing acute to sub acute hematoma in the dorsal epidural space in lumbar spinal canal causing severe canal stenosis at L2-3 level to L3-4 level.
Figure 1c – Axial T2W MRI shows dilated vessels in right lumbar paraspinal muscles with extension through right neural foramina into spinal canal. Acute to sub acute haematoma in dorsal epidural space causing lumbar canal stenosis.

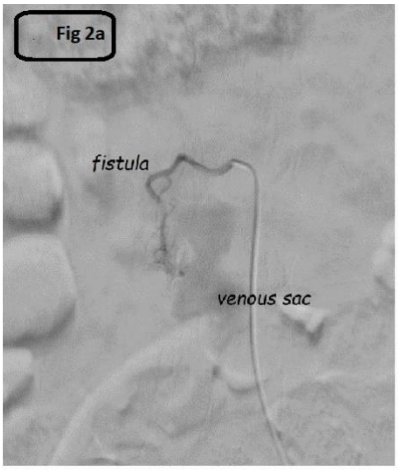
difficulty or nasal intonation of voice. Child was hemodynamically stable on examination. There was no recent history of fever, diarrhoea or vomiting prior to this episode. There was no history of similar illness in the family.

Nerve conduction velocity (NCV) was suggestive of motor axonal neuropathy. Routine biochemistry was within normal limits. Examination of her back revealed a pigmented nevus in her lumbar paraspinal region. Magnetic resonance imaging (MRI) of her spine showed posterior spinal canal acute epidural hematoma from L2-L4 vertebral body causing neural compression. A lumbar paraspinal vascular malformation was seen extending to extradural space and right L2-L3 and L3-L4 neural foraminae [Figure 1a, 1b, 1c]. She underwent emergency L1-L4 laminectomy and decompression of extradural hematoma. During surgery, a large abnormal dilated vascular channel was seen crossing obliquely through the spinal canal, causing neural compression. This was excised and sent for biopsy. In the acute phase of presentation, the priority was to decompress the spinal cord at the earliest. Laminoplasty was the initial intention. During surgery, as the anatomical findings became evident, laminectomy was done to maximise the space and reduce the risk of injuring the abnormal dilated fragile epidural vascular structures by the repositioned lamina. The paraspinal vascular malformations were not targeted surgically at this stage to avoid further blood loss and the technical challenges of chasing elastic vessels of a large vascular malformation that retract and bleed from tissue depths. Following decompression, the patient started improving neurologically. Spinal stability was less of a concern in the emergency scenario and can be addressed in the future by instrumental stabilisation if the clinical progress at follow-up warrants so. In practice, the

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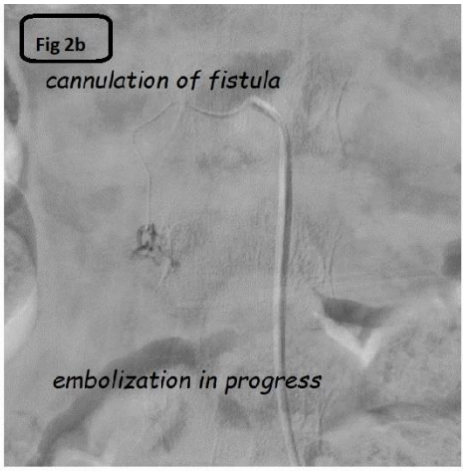


Figure 2a and 2b - Angiography study – pre procedure.

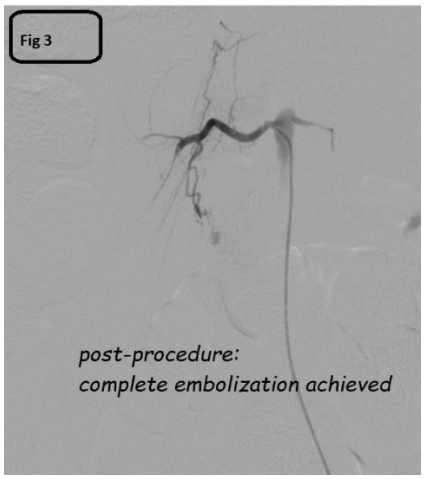


Figure 3 - Angiography study – post embolization.

neurosurgery team, here, tries to avoid prophylactic or anticipatory instrumentation as much as possible.

Following the day of surgery, power in her lower limbs improved slightly and her pain subsided. She remained catheterised for her voiding difficulty. Child was taken home against medical advice by relatives due to logistical reasons. She was readmitted after 2 months for further treatment.

Digital subtraction angiography was done which delineated the architecture of the spinal vascular malformation [Figure 2a -2b]. She had a moderate flow, perimedullary pial fistula. There was a large venous sac having multiple arterial feeders from right side of L3. The anterior spinal artery was seen from left L1 level. Subsequently embolisation of spinal vascular malformation was done with glue [Figure 3].

The child continued to have gradual improvement of motor function. She started walking with support one month after surgical decompression. She was troubled by a nagging allodynia and radicular pain along lower limbs, which started subsiding after embolisation. At 20 months follow-up, after spinal decompression, she is able to walk at normal pace and gait, with mild residual hypertonia of the left lower limb evident on close clinical examination. There is no pain and she is off all medications. Bladder control has normalised.

DISCUSSION:

SVMs include capillary telangiectasia, cavernous malformation, arterio-venous malformations and arterio-venous fistulas [8]. They have varied age of presentation, different clinical features and different radiological appearances. SVMs can be intra medullary, perimedullary, extra dural or radicular depending on their location or high-flow, low-flow or with no arteriovenous shunt depending on their flow pattern [9]. High flow SVMs are more commonly seen in children and young adults. They are predominantly congenital lesions. Our patient had a similar presentation in her second decade of life. Clinical examination revealed a pigmented nevus in her lumbar paraspinal region which had been previously ignored. This was communicating with the paraspinal SVM. They may be detected incidentally or may present with haemorrhage or mass effect as we saw in our patient who presented with radicular pain, hypotonia in her lower limbs and urinary and bowel retention. Increased spinal venous pressure, leads to venous congestion which in turn reduces the drainage of the spinal veins contributing to progressive myelopathy as we saw in our case [2].

Our patient had a vascular skin lesion in the same metameric segment as the SVM and was diagnosed as SAMS. These are often diagnosed late, as patients remain asymptomatic and disregard the skin lesions. Cutaneous manifestation can include port wine stain, angioma, angiolipoma or angio keratoma. The vascular lesion does not involute spontaneously like a capillary hemangioma [4-7]. The embryological origin of the blood supply to the spinal cord and verterbra arises from the segmental dorsal arteries which explains the similar metameric distribution in SAMS.

MRI is a useful tool to detect low flow SVMs as we saw in our patient. Digital subtraction angiography was done later, for better delineation. Angiography is the gold standard to evaluate SVM [9].

Treatment involves surgical approach, embolisation or a combination of both [10]. Our patient underwent an emergency decompressive surgery to reduce the neural compression. Embolisation later, resulted in significant improvement.

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DOI: 10.46900/apn.v5i2.172

CONCLUSION:

SAMS are extremely rare in children with very few reported cases [11]. They may undergo a rapid progression to monoparesis, paraplegia or quadriplegia if not diagnosed early. A multi-disciplinary approach beginning with a detailed history taking and clinical examination is invaluable in such complicated cases to deal with the various aspects of a complex pathology.

ABBREVIATIONS:

SVM – spinal vascular malformation

SAMS- Spinal arteriovenous metameric syndrom

NVC- Nerve conduction velocity

MRI – Magnetic resonance imaging

DISCLOSURES

Ethical approval

This study was performed in line with the principles of the Declaration of Helsinki. Ethics approval was not required for this study

Consent to participate

The patients gave consent to use their information and images for research purposes. *Consent for publication*

The patient gave consent to use his information and images for publication.

Conflict of interest

The authors declare no conflicts of interest with respect to the content, authorship, and/or publication of this article.

Funding

This research received no specific grant from any funding agency in the public, commercial or not-for-profit sectors

CONTRIBUTIONS

Satyajit Das: Conceptualization, Investigation ,Data curation, Writing – original draft

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