


CASE REPORT

Open Access



# A rare case report of endovascular management of pediatric spinal arteriovenous malformation

Srikanth Amaravadi<sup>1</sup>, Kovilapu Uday Bhanu<sup>1</sup>, Saurabh Maheshwari<sup>2\*</sup> , Peeyush Dhagat<sup>1</sup>, Shweta Pandey<sup>1</sup> and Sumesh Menon<sup>3</sup>

## Abstract

**Background:** Spinal arteriovenous malformations (AVM) consist of a heterogeneous group of pathological vascular entities that affect the spinal cord parenchyma either directly or indirectly.

**Case presentation:** We present an unusual case of spinal arteriovenous malformation (type 2) in an 18-month-old girl who presented with weakness of both lower limbs and urinary incontinence. She was diagnosed with a spinal AVM with large intramedullary nidus and paraspinal extension which was managed with endovascular embolization.

**Conclusion:** Spinal cord AVM in children can be debilitating. If presented early, patient can be taken up for embolization, which is a relatively safe procedure with better neurological outcome. Our case illustrates the successful role of interventional radiology in the treatment of this rare condition.

**Keywords:** Spinal AVM, Nidus, Embolization, Interventional radiology, Paediatrics

## Background

Spinal arteriovenous malformations (AVM) consist of a heterogeneous group of pathological vascular entities that affect the spinal cord parenchyma either directly or indirectly [1]. Male predominance is noted in the pediatric population, but in the adult population, no sex predominance was documented [2, 3]. The earliest clinical report of a spinal vascular malformation was in 1890 by Berenbruch, who did not acknowledge the lesion as a vascular pathology at surgery but only later at autopsy. Later on, Krause identified a spinal vascular malformation during surgery [3]. Typically, there is a delayed presentation, resulting in a lapse of the significant period between the onset of symptoms and diagnosis, leading to

deterioration in neurological function, which could have been prevented with a timely intervention [4].

Spinal arteriovenous malformations have been classified into four types: dural arteriovenous fistula (type 1), intramedullary glomus (type 2), intramedullary juvenile type arteriovenous malformation (type 3), and intradural-extramedullary arteriovenous fistula (type 4) [5]. Partial embolization and ligation of feeding vessels remain the mainstay of managing these enigmatic lesions [6, 7].

Here, we present an unusual case of spinal arteriovenous malformation (type 2) in a pediatric patient with large intramedullary nidus with paraspinal extension managed with embolization.

## Case presentation

An 18-month-old female child presented to the neurology outpatient department with complaints of weakness of both lower limbs and urinary incontinence for 4 days. The child had been toilet trained at the age of 12 months. The onset of weakness was acute in nature and non-progressive. There was no history of trauma to the spine, no

\*Correspondence: saurabhshwari@yahoo.co.in

<sup>2</sup> Department of Radiodiagnosis and Imaging, Military Hospital Shimla, Shimla, India

Full list of author information is available at the end of the article

history of fever, and no history of muscular dystrophies. The child was a full-term normal vaginal delivery at the hospital with no intrapartum or postpartum complications. Developmental milestones were age-appropriate and immunized per the Indian Academy of Pediatrics schedule. There was no history of marriage consanguinity. General and other systems examinations were found to be normal. MRI of the spine was requested to look for the cause of her weakness.

MRI spine with standard pre- and post-contrast sequences sagittal T1, T2, STIR, HASTE T2, MEDIC, Axial T1, T1 FS, and T2, Coronal STIR, and MR myelograms was included in the protocol. IV contrast was administered, and multiplanar post-contrast imaging was done. The MR images revealed: Abnormal intramedullary bunch of flow voids with a sac-like structure measuring 9.0 mm × 10.5 mm × 15 mm (AP × TR × CC) was seen extending from D9 to D10 Intravertebral disk level to mid D11 vertebral body level. Associated cord edema was seen extending superiorly till the D1 vertebral level and till conus medullaris inferiorly (Fig. 1).

Based on MRI findings, the child was diagnosed with a spinal arteriovenous malformation type 2 with intra- and extramedullary components of the vascular sac and evidence of intramedullary hematoma.

Digital subtraction angiography (DSA) was done, which showed hypertrophied intercostal artery with a large intramedullary and extramedullary tangle of vessels. Given complex anatomy, the neurosurgeon and

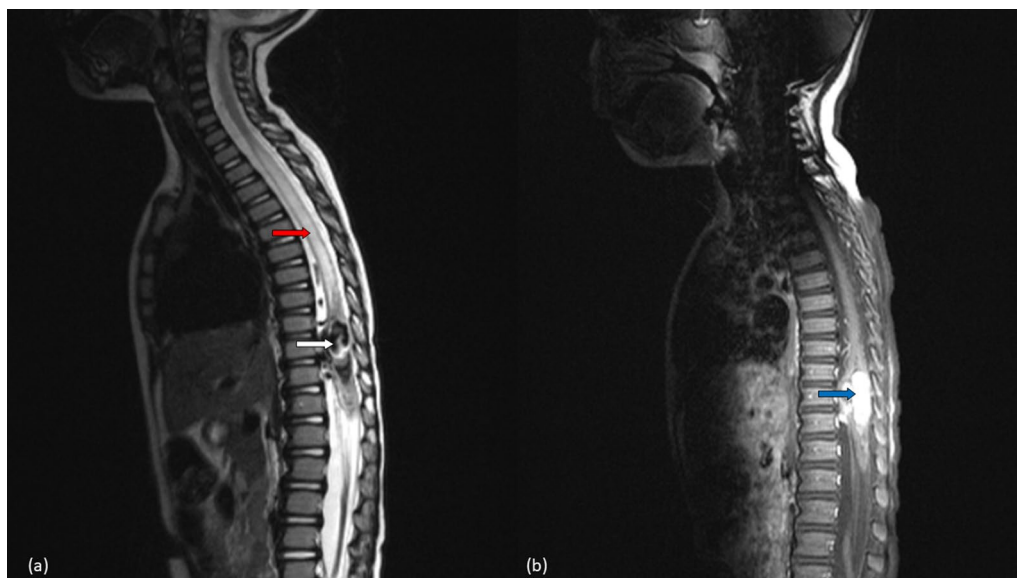
neuro-intervention team took the decision of targeted endovascular embolization.

The child was taken up for endovascular embolization under general anesthesia in a separate sitting after appropriate counseling of the parents and obtaining informed consent from them. A diagnostic spinal vessel angiogram revealed an abnormal hypertrophied feeder from the left D8 intercostal artery supplying the AV Fistula with an aneurysmal sac. Superselective cannulation of the feeding artery was done using a flow-directed microcatheter (Marathon, EV3), and 2 ml of 30% NBCA glue (n-butyl cyanoacrylate) was injected at the fistulous site. Complete obliteration of the AV fistula and intramedullary aneurysmal sac was achieved (Fig. 2).

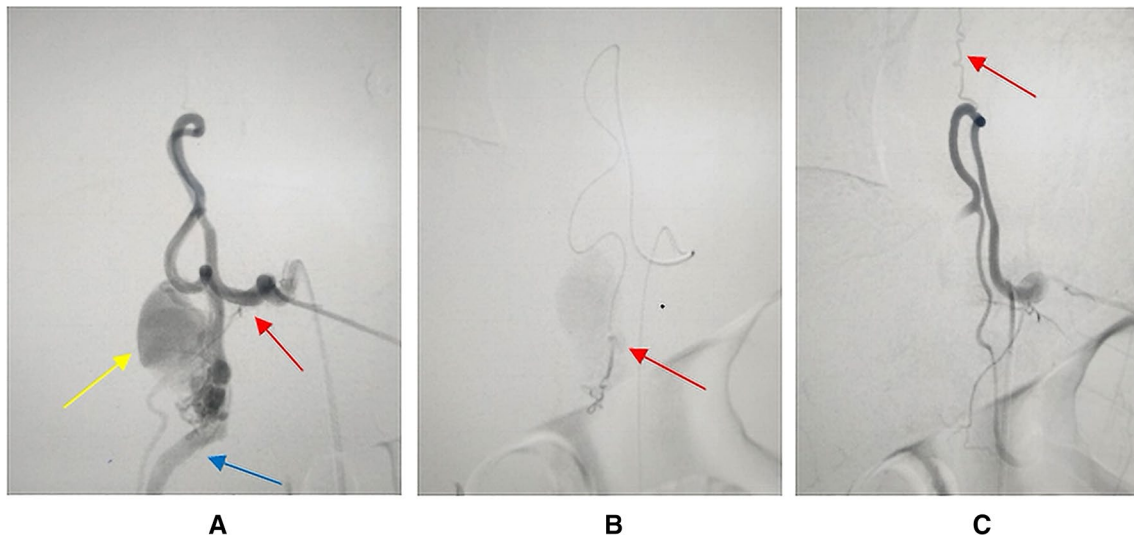
Post-embolization MRI was done, and it revealed a significant reduction in nidus and obliteration of sac. There was also resolution of the cord edema (Fig. 3). On follow-up, the child gradually improved over the next 1 year and could walk with assistance and regain complete bladder control. At 36 months, the patient could walk with support, regained complete bladder control, and residual wriggling gait was present.

## Discussion

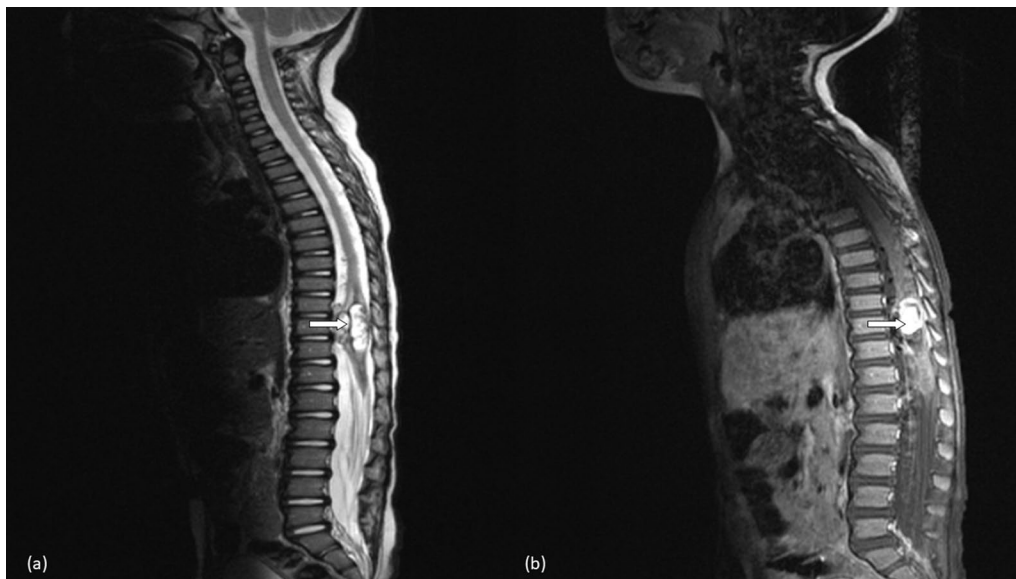
Spinal arteriovenous lesions are rare entities, with several complications if untreated. These account for 3–16% of all space-occupying lesions in the spinal cord [8]. Although spinal cord AVMs are common during the third decade of life, 20% of the cases occur before



**Fig. 1** **a** T2 weighted and **b** T1 weighted post-contrast sagittal Magnetic Resonance Imaging (MRI) images of the whole spine show peri- and intramedullary spinal AVM at D9–D11 spinal level (white arrow) with surrounding spinal cord edema extending from D1–D11 spinal level (red arrow). The post-contrast images show diffuse enhancement of the nidus (blue arrow)



**Fig. 2** Digital Subtraction Angiography (DSA) images: **A** Left D8 intercostal digital subtraction angiogram shows abnormal hypertrophied feeder (red arrow) with intranidal aneurysm (yellow arrow) and draining vein (blue arrow). **B** Super selective microcatheter run at a fistulous site (red arrow). **C** Post-glue embolization shows the obliteration of AVM and normal opacification of the anterior spinal artery (red arrow)



**Fig. 3** Post-embolization **a** T2 weighted sagittal and **b** T1 weighted post-contrast Magnetic Resonance Imaging (MRI) images of the whole spine show regression of nidus at D9–D11 level (white arrows in **a** and **b**) with the resolution of the cord edema seen in Fig. 1a

age 16 [9]. Spinal AVM patients generally present with symptoms of backache and progressive myelopathy, gait abnormalities, sensory involvement, and bladder/bowel incontinence [10]. Management of spinal AVMs includes vascular embolization, surgery, or a combination of both. Conservative management is offered to a specific type of lesions [11].

The spinal cord may be edematous on MRI, which appears as a hyperintense signal on T2WI MRI. There are associated low signal areas at the dorsal surface of the cord. These low signal intense areas represent the engorged perimedullary venous plexus. The spinal cord is bulky and hypointense on T1-weighted images. Post-contrast images show intense enhancement representing engorged veins with disruption of the blood barrier of

the spinal cord. Magnetic resonance angiography (MRA) may provide additional information as it could show the possible level of fistula. However, DSA plays a crucial role before management as it can provide the micro-architecture of the lesion. DSA can identify the number, morphology, and exact anatomical location of the arterial feeders, nidus, and venous drainage of the malformation [12].

Spinal cord AVMs are associated with a poor prognosis if left untreated. Djindjian et al. [13] found that 36% of individuals younger than 40 progress to severe impairment after 3 years of evolution.

Embolization plays a crucial role in managing intramedullary spinal AVMs either as a primary treatment or as an adjunct to surgery. Even partial embolization plays a significant role in the outcome [14]. Pediatric age group spinal arteriovenous shunts account for thirteen percent of all cases. Although 17% of the pediatric population showed symptoms, they were undetected till adulthood. Intramedullary-Extramedullary AVM and complex Angiomatosis are complicated to treat because of the complex architecture of these lesions involving both intradural and extradural compartments and, at times, that of normal cord parenchyma with the actual AVM. Even though rare, some case reports in the literature document the complete regression of AVMs by using combined angiographic embolization and extensive surgical resection.

## Conclusions

In our case, endovascular embolization was chosen as the primary treatment option for the expected risk of neurological deterioration due to surgical resection. Post-embolization, complete obliteration of the spinal cord AVM was obtained. On follow-up, the child had satisfactory improvement in the neurological status. Spinal cord AVM in children can be debilitating. If presented early, patient can be taken up for endovascular embolization, which is a relatively safe procedure with better neurological outcome.

## Abbreviations

AVM: Arteriovenous malformations; TSE: Turbo spin echo; STIR: Short tau inversion recovery; HASTE: Half Fourier single-shot turbo spin-echo; MEDIC: Multiple echo data image combination; MRI: Magnetic resonance imaging; DSA: Digital subtraction angiography; NBCA: *N*-butyl cyanoacrylate.

## Acknowledgements

We would like to thank Dr. Samaresh Sahu for his support in conducting this study.

## Author contributions

SA conceptualized the design of this case report. SA and KUB contributed to the acquisition and analysis of patient data and images. SM and SP contributed to the drafting and revision of the case report. PD and SM2 contributed to the interpretation of images. All authors have agreed both to be personally

accountable for their own contributions, and they have ensured that questions related to the accuracy or integrity of any part of this work, even ones in which they were not personally involved, were appropriately investigated, resolved, and the resolution was documented in the literature. All authors have read and approved the manuscript.

## Funding

No one was paid during this study. The study did not have a source of funding. This study was not supported by a grant.

## Availability of data and materials

All data are available based on a reasonable request.

## Declarations

### Ethics approval and consent to participate

The present study was approved by the ethical board of the hospital in which the study was performed. The patient reported in this article had signed a written informed consent form. This case report was a reporting of a case in a medical educational center, in which all patients are informed that they may be subjects of scientific experiments and are informed of the ethical codes of conducts. This study was in compliance to the latest version of the Declaration of Helsinki.

### Consent for publication

The patient had written and signed an informed consent note that the findings may be published without any personal detail.

### Competing interests

The authors declare that they have no competing interests.

### Author details

<sup>1</sup>Department of Radiodiagnosis and Imaging, Armed Forces Medical College, Pune, India. <sup>2</sup>Department of Radiodiagnosis and Imaging, Military Hospital Shimla, Shimla, India. <sup>3</sup>Department of Accident and Emergency, Military Hospital Trivandrum, Thiruvananthapuram, India.

Received: 4 July 2022 Accepted: 5 December 2022

Published online: 09 December 2022

## References

- Baylor College of Medicine. Spinal vascular malformations. <https://www.bcm.edu/healthcare/care-centers/neurosurgery/conditions/spinal-vascular-malformations>. Accessed 20 June 2022
- Rodesch G, Hurth M, Alvarez H, Ducot B, Tadie M, Lasjaunias P (2004) Angio-architecture of spinal cord arteriovenous shunts at presentation. Clinical correlations in adults and children. The Bicêtre experience on 155 consecutive patients seen between 1981 and 1999. *Acta Neurochir (Wien)* 146:217–226. <https://doi.org/10.1007/s00701-003-0192-1>. (discussion 226–7)
- Krause, F. (1912) Surgery of the brain and spinal cord based on personal experience. In: Haubold A, Thorek M (ed) *Trans. Rebman*, New York
- Kramer CL (2018) Vascular disorders of the spinal cord. *Continuum (Minneapolis)* 24(2, Spinal Cord Disorders):407–426
- Anson JA, Spetzler RF (1992) Interventional neuroradiology for spinal pathology. *Clin Neurosurg* 39:388–417
- Djindjian R, Merland JJ, Djindjian M, Houdart R (1978) Place de l'embolisation dans le traitement des malformations artério-veineuses Médullaires à propos de 38 Cas. In: *Proceedings of the XI. Symposium neuroradiologicum*. Springer, Berlin, Heidelberg, pp 428–429
- Lin N, Smith ER, Scott RM, Orbach DB (2015) Safety of neuroangiography and embolization in children: complication analysis of 697 consecutive procedures in 394 patients. *J Neurosurg Pediatr* 16(4):432–438. <https://doi.org/10.3171/2015.2.peds14431>
- Cogen P, Stein BM (1983) Spinal cord arteriovenous malformations with significant intramedullary components. *J Neurosurg* 59:471–478. <https://doi.org/10.3171/jns.1983.59.3.0471>

9. Yaşargil MG, Symon L, Teddy PJ (1984) Arteriovenous malformations of the spinal cord. *Adv Tech Stand Neurosurg* 11:61–102. [https://doi.org/10.1007/978-3-7091-7015-1\\_4](https://doi.org/10.1007/978-3-7091-7015-1_4)
10. Park JE, Koo H-W, Liu H, Jung SC, Park D, Suh DC (2018) Clinical characteristics and treatment outcomes of spinal arteriovenous malformations. *Clin Neuroradiol* 28:39–46. <https://doi.org/10.1007/s00062-016-0541-0>
11. Patsalides A, Knopman J, Santillan A, Tsiouris AJ, Riina H, Gobin YP (2011) Endovascular treatment of spinal arteriovenous lesions: beyond the dural fistula. *AJNR Am J Neuroradiol* 32:798–808. <https://doi.org/10.3174/ajnr.A2190>
12. González AL, Tjahjadi M, Nuñez AM, Rivas FJM (2019) Spinal arteriovenous malformations. In: *Neurovascular surgery*. Springer, Singapore, pp 239–247
13. Djindjian R, Cophignon J, Rey A, Thron J, Merland JJ, Houdart R (1973) Superselective arteriographic embolization by the femoral route in neuroradiology: Study of 50 cases. III. Embolization in craniocerebral pathology. *Neuroradiology* 6:143–153. <https://doi.org/10.1007/bf00340442>
14. Ozpinar A, Weiner GM, Ducruet AF (2017) Epidemiology, clinical presentation, diagnostic evaluation, and prognosis of spinal arteriovenous malformations. *Handb Clin Neurol* 143:145–152. <https://doi.org/10.1016/B978-0-444-63640-9.00014-X>

### Publisher's Note

Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

Submit your manuscript to a SpringerOpen<sup>®</sup> journal and benefit from:

- ▶ Convenient online submission
- ▶ Rigorous peer review
- ▶ Open access: articles freely available online
- ▶ High visibility within the field
- ▶ Retaining the copyright to your article

---

Submit your next manuscript at ▶ [springeropen.com](https://www.springeropen.com)

---