



# Isolated fourth ventricle haemorrhage: “think beyond intracranial source” unusual presentation of lumbosacral spine arteriovenous malformation presentation

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## ABSTRACT

Spinal arteriovenous malformations (SAVMs) are rare vascular lesions and account for about 4% of primary intraspinal masses. Since SAVMs can involve any location along the spinal column and produce a host of different problems, the symptoms are extremely variable. There are few reports of simultaneous cerebral SAH and intraventricular hemorrhage (IVH) following rupture of a spinal AVM (SAVMs). Herein, we present a rare case of Lumbo Sacral spine arteriovenous malformation, which clinically manifests as sudden onset of severe headache and vomiting due to isolated fourth ventricle Hemorrhage (IVH) without cerebral subarachnoid hemorrhage.

## INTRODUCTION

Being a rare category, spinal vascular malformations (SAVMs) account for about 4 % of primary intraspinal masses (3). Rosenblum and coworkers described two major types of spinal AVM intradural versus dural on the basis of location of nidus: intradural versus dural. Intradural AVMs were classified as intramedullary and dural AVFs (Arteriovenous fistula). They usually present with pain, numbness, and weakness, loss of bowel/bladder control, incoordination, and impotence is few of the issues. In majority of cases SAVMs first come to medical attention by bleeding - this usually presents as acute, severe back pain, followed by sudden onset weakness, numbness, incontinence; severity ranges from no neurologic dysfunction to complete paralysis, depending on location and extent of bleeding. The natural history of spinal vascular malformations (SAVMs) is unpredictable and varies from acute subarachnoid hemorrhage of spine to venous congestion of cord.

As far as cranial manifestation of SAVMs concerns cerebral subarachnoid hemorrhage (SAH) has been reported in English literature, and seems to be caused by rupture of same intra ventricular bleed due

## Keywords

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to SAVMs has been reported four times in last decade. We believe this to be the first reported case of a Lumbo-sacral SAVMs presenting as isolated fourth ventricle hemorrhage (IVH.) We also need a kind attention regarding left lower limb swelling; it might be due to raised venous pressure which was resolved spontaneously after definitive surgical management of SAVMs.

### CASE REPORT

A twenty-year-old adult male experienced sudden onset severe headache associated with episodes of vomiting in a morning during defecation, which was preceded by severe lower back pain. There was no history or signs of trauma, no previous history of back pain, radiculopathy, or myelopathy were reported. On examination it was revealed that patient was conscious, oriented and without any focal neurological sign. He had left lower limb swelling which was misdiagnosed as varicose vein of limb and was operated 6 months back for same, but swelling did not subside.



Non contrast computed tomography (CT) of head was done, which showed fourth ventricle hemorrhage with concomitant sparing of bilateral lateral and third ventricle, all cisterns were seen normally and had no evidence of cisternal bleeding and sub arachnoid hemorrhage (FIGURE 1). Magnetic Resonance imaging (MRI) brain, CT Angiography brain and digital subtraction angiography (DSA) brain did not reveal any intra cranial source of bleeding, but MRI Lumbo sacral region (FIGURE 2) and spine Digital subtraction angiography revealed spinal perimedullary AVM (SAVMs) at L1 to S1 level (conus medullaris) with feeding artery aneurysm and venous drainage from spinal vein to perimedullary vein and extending cranially (figure-3). The lesion was treated surgically as cauterization and disconnection of the vein. Till the writing of this report 12 month follow up has been completed and he is neurologically intact and most importantly lower limb swelling subside within first month of surgery (FIGURE 4).

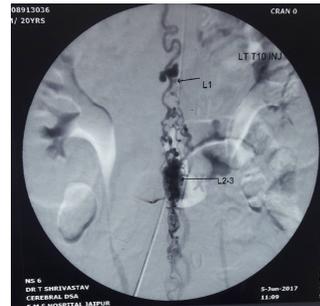


FIGURE 1. NCCT Head showed small hyper density in 4th ventricle suggestive of recent intraventricular bleed.

FIGURE 2. MRI L S Spine T2 weighted with GRE shows multiple flow void in intradural lumbo-sacral region extending from L1 to S1 levels. Subtle enhancement is seen in blood vessel with slow flow. No enhancement is seen with blood vessel with high flow suggestive of spinal dural AVF.

FIGURE 3. Spinal DSA showing Lumbo-sacral dural AVF with pre-nidal aneurysms with feeder from left T10 radiculomeningeal artery.

FIGURE 4. Swelling left leg, which reduced drastically after treating primary pathology along with hyper pigmentation on dorsum of foot.

### DISCUSSION

Spinal arteriovenous malformation (SAVM) is a rare, abnormal tangle of blood vessels on, in or near the spinal cord and account approximately 10% of CNS vascular malformations in all age groups. These lesions are directly supplied by radicular arteries and drained by spinal cord veins, although dural supply can occur as with dural arteriovenous fistulas. (3, 7)

On the basis of location these lesions are divided into either intradural or extradural, and intradural further subdivided into intramedullary or extramedullary. Most are thoracolumbar, posterior, and outside the cord (extramedullary). The rest are cervical or upper thoracic and often inside the cord

(intramedullary). AVMs may be small and localized or may affect up to half the cord. (3,6)

In majority of patients the ultimate fate of spinal AVM is progressive neurological deficit in terms of sensory, motor or bladder/ bowel involvement. In

some situation rarely, high cervical AVMs rupture into the subarachnoid space, causing subarachnoid hemorrhage with sudden and severe headache, nuchal rigidity, and impaired consciousness (4). The natural history of SAVMs is characterized by venous congestion causing progressive neurological deficits in majority of patients. (3,8)

In the literature, a thoracolumbar SAVMs presenting with both SAH and IVH appears to be a rare occurrence. Although there are few reports of concomitant cerebral SAH and intraventricular hemorrhage (IVH) following rupture of a spinal SAVMs (2,3,5). There have been three case reports of SAVMs presenting in the adult population and two in pediatric age group with intraventricular hemorrhage (IVH) exist in the literature. In both groups, the clinical, radiographic, and surgical findings suggested that the SAVM was the source of the hemorrhage.

In 1999 P Bazro et al reported first case of intraventricular hemorrhage attributed by the

rupture of conus medullaris AVM in a young patient. (1). ES Marlin et al reported a case of intraventricular hemorrhage in both lateral, third and fourth ventricle in a 2-year-old female child caused by rupture thoraco lumber SAVMs, who died in next few days probably due to re rupture. (3). Recognition of such cases in future may allow earlier diagnosis and treatment before catastrophic re hemorrhage. Masanori et al reported another case who presented with intraventricular hemorrhaging (IVH) into the fourth and third ventricles that was caused by a cervical intramedullary arteriovenous malformation. (4)

To the best of our knowledge, this is the first case report of an adult patient in whom initial imaging demonstrated isolated fourth ventricle hemorrhage (IVH) without Sub arachnoid hemorrhage (SAH) secondary to a ruptured low lumbosacral SAVMs. Till the date only five cases of Intraventricular hemorrhage is reported in literature.

TABLE 1. Patients presenting with IVH caused by rupture of Spinal AVMs

S.NO.	Author(s)	Year	Age/sex	Ventricular hemorrhage	Location of spinal AVM
1	H Baharvahdat et al (2)	2016	48 Year/Male	B/L Lateral, Third & fourth ventricle and SAH	Conus Medullaris AVM
2	E. S. Marlin et al. (3)	2014	2 Year/ Female child	B/L Lateral, Third & fourth ventricle	Thoraco lumber Spinal AVM
3.	Kenning. T.et al (5)	2009	1 Year 2 month /female childe	B/L Lateral, Third & fourth ventricle and SAH	Thoraco lumber spinal perimedullary AVM
4	Masanori Ito et al (4)	2007	33 Year /male	Third & fourth ventricle	Cervical intramedullary spinal AVM
5	P Barzó et al (1)	1999	28 Year /male	B/L Lateral, Third & fourth ventricle	Conus medullaris AVM
6	Present case	2017	20 Year/male	Isolated fourth ventricle	Lumbo sacral Spinal AVM

Although to kept Lumbosacral SAVMs as a differential diagnosis for isolated spontaneous fourth ventricle IVH is beyond the imagination

particularly in adult patient, where the distance between the bleeding source and presenting site is significant. The present case suggests that MRI of

the entire spine with dedicated blood detected sequence GRE & SWI should be considered if cranial angiography does not reveal a source. (2,3). There must be some possible hypothesis of raised venous pressure behind the swelling of left leg due to SAVMs being wrongly operated for varicocities. So, such possibilities in these cases should also be considered.

### CONCLUSIONS

The case reported raises necessity of complete spinal neuraxis evaluation especially in young group of patients presenting with angiographically negative intraventricular hemorrhage. Evaluation for thoracolumbar spinal vascular malformations must be included in the initial work up. A whole spinal workup should be considered, when bleeding from intracranial origin is carefully excluded.

This is reminder to treating neurosurgeon along with concern medical fraternity as neurointerventionalist for careful consideration this rare differential and it has to be kept in mind that presentation can varied from head to toe.

### ABBREVIATIONS

IVH - Intra Ventricular hemorrhage  
 SAH - Sub arachnoid hemorrhage  
 SAVM - Spinal arterio vascular malformation  
 DAVF - Dural arterio venous fistula  
 MRI - Magnetic Resonance imaging (MRI)  
 DSA - Digital subtraction angiography  
 SWI - Susceptibility weighted imaging

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