



Case Report

Spontaneous regression of extradural high-flow vascular malformation in spinal arteriovenous metamerism syndrome (SAMS): A unique case report

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Received : 02 January 2023

Accepted : 19 April 2023

Published : 05 May 2023

DOI

10.25259/SNI_4_2023

Quick Response Code:



ABSTRACT

Background: Spinal arteriovenous metamerism syndrome (SAMS) is a rare nonhereditary genetic vascular disorder, involving multiple layers of tissues in the same metamerism level. Spontaneous regression of SAMS has never been reported in the medical literature.

Case Description: A 42-year-old woman presented with intermittent low back pain for 6 months. Magnetic resonance imaging of the thoracolumbar spine incidentally found clusters of spinal vascular malformations involving spinal cord, vertebral bodies, epidural space, and paraspinal muscles. There was no sign of venous congestion. Magnetic resonance angiography and spinal angiography revealed intradural spinal cord arteriovenous malformation (SCAVM) at the level of T10-11 and extradural high-flow osseous arteriovenous fistula. Due to asymptomatic SAMS and a high risk of anterior spinal arterial compromise during treatment, conservative treatment was considered in our patient. Spinal angiography obtained 8 years after initial angiography demonstrated significant regression of extradural component of SAMS and stable intradural SCAVM.

Conclusion: We describe a unique case of SAMS with spontaneous regression of extradural component during a long-term observation period.

Keywords: Spinal arteriovenous metamerism syndrome, spinal vascular malformation, spinal cord arteriovenous malformation, spinal epidural arteriovenous fistula, spontaneous resolution

INTRODUCTION

Spinal arteriovenous metamerism syndrome (SAMS), a rare subtype of spinal cord arteriovenous malformations (SCAVMs), is a complex nonhereditary genetic vascular disorder, affecting spinal cord, bony structure, epidural space, and/or paraspinal soft tissue or muscles in the same metamerism level.^[2,17,19] Most patients harboring SAMS have intra and extradural vascular malformations.^[5] Spontaneous regression of SAMS has never been reported in the medical literature.^[13] The authors described a unique case of SAMS with spontaneous regression of extradural high-flow vascular malformation during a long-term observation period.

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CASE DESCRIPTION

A 42-year-old woman presented with intermittent low back pain for 6 months without history of trauma. She went to the local hospital and obtained plain radiography of lumbar spine, revealing scoliosis and mixed lytic and sclerotic lesions at L1 and L2 vertebral bodies with collapse of left side of L1 vertebral body. Magnetic resonance imaging (MRI) of the thoracolumbar spine was performed and incidentally found clusters of vascular malformations. SCAVM was presumably diagnosed, and the patient was transferred to our institute for further investigation and proper treatment. Physical examination showed a large well-circumscribed mass, which is approximately 14 cm in diameter, over the left side of the lower back [Figure 1]. The patient was no muscle weakness, urinary or fecal incontinence, or any neurological deficits.

MRI of the thoracolumbar spine demonstrated abnormal tortuous and dilated flow voids surrounding the lower spinal cord and conus medullaris, particularly at posterior surface of T10-11 level, and at left-side of L1 and L2 vertebral bodies, epidural space, neural foramina, and paraspinal muscles including psoas major, multifidus, erector spinae, and quadratus lumborum. The affected muscles from T12 to L4 vertebral levels were hypertrophy and markedly enhanced after administration of contrast material with mild hyperintense T2 signal change on T2-weighted sequence. There was no signal intensity change of the spinal cord [Figures 2-4]. Contrast-enhanced magnetic resonance angiography (MRA) of the thoracolumbar spine disclosed clusters of vascular malformations including intradural spinal cord arteriovenous malformation at the level of T10-



Figure 1: The photograph of the patient in posterior view shows a large well-circumscribed mass, which is approximately 14 cm in diameter, over the left side of the lower back.

11 and extradural paravertebral arteriovenous malformation from T12 to L2 vertebral levels [Figure 5]. Spinal angiography confirmed SCAVM, nidus type, at the level of T10-11 supplied by left posterior spinal arteries that receives radiculopial branches from the left T11intercostal and L1 segmental arteries, and sulco-commissural feeder arising from the anterior spinal artery of the left T9 with caudal drainage into perimedullary veins and subsequent drainage to the dilated left sacral radicular vein. Another high-flow spinal osseous epidural arteriovenous fistula (SOEAVF) was demonstrated and fed by multiple arterial feeders from bilateral T12 intercostal, L1 segmental, and left L2 segmental arteries converging into the venous lake at the level of L1 and L2 with venous drainage through enlarged epidural and paravertebral venous plexus and subsequent drainage to the inferior vena cava [Figures 6-8]. Computed tomography scan of the lumbar spine revealed multiple well-defined tubular and round-shaped osteolytic lesions and erosion with sclerotic rims involving particularly left-sided vertebral bodies and posterior elements of L1 and L2, corresponding with the dilated venous lake [Figure 9].

All above-mentioned imaging features contributed to a definite diagnosis of SAMS at T10-11–L2 levels. Surgical or endovascular treatment of these lesions was considered a high risk of anterior spinal arterial compromise. In addition, she was asymptomatic from vascular malformations. Therefore, the patient was kept for continuous observation and has attended our outpatient department every year without missing any follow-up visit. At 3rd year of the follow-up, MRI and MRA were performed and showed unchanged of the lesions. At 4th year of the follow-up, spinal angiography was scheduled to perform but the procedure was postponed due to the COVID-19 pandemic. Spinal angiography obtained 8 years after initial angiography surprisingly showed significant regression of extradural component of SAMS and stable intradural SCAVM [Figure 10].

DISCUSSION

Since 1915, Cobb syndrome was characterized by the concurrent findings of a spinal vascular malformation and a vascular skin lesion affecting the corresponding dermatome.^[3] Subsequently, Bhattacharya *et al.*^[1] proposed the new name of SAMS, arising from a somatic mutation developing in the region of the neural crest or adjacent mesoderm before migration and differentiation, producing multiple vascular malformations in the same metameric distribution.

SAMS is classified as extradural-intradural arteriovenous malformations with two or more vascular malformations involving spinal cord, bone, paraspinal musculature, subcutaneous tissues, and/or skin in one or several metameric segments.^[4,11,17,19] The intradural lesion associated

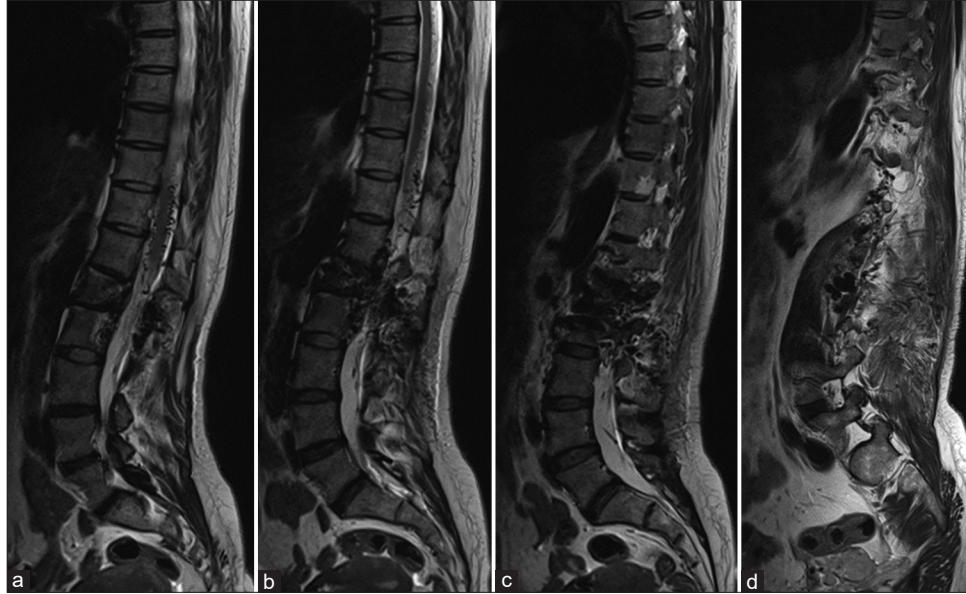


Figure 2: (a-d) Sequential sagittal T2-weighted images from midline to left side of the thoracolumbar spine reveal abnormal tortuous and dilated flow voids surrounding the lower spinal cord and conus medullaris, particularly at posterior surface of T10-11 level, and at left-side of L1 and L2 vertebral bodies, neural foramina, and paraspinal muscles. There is no signal intensity change of spinal cord.

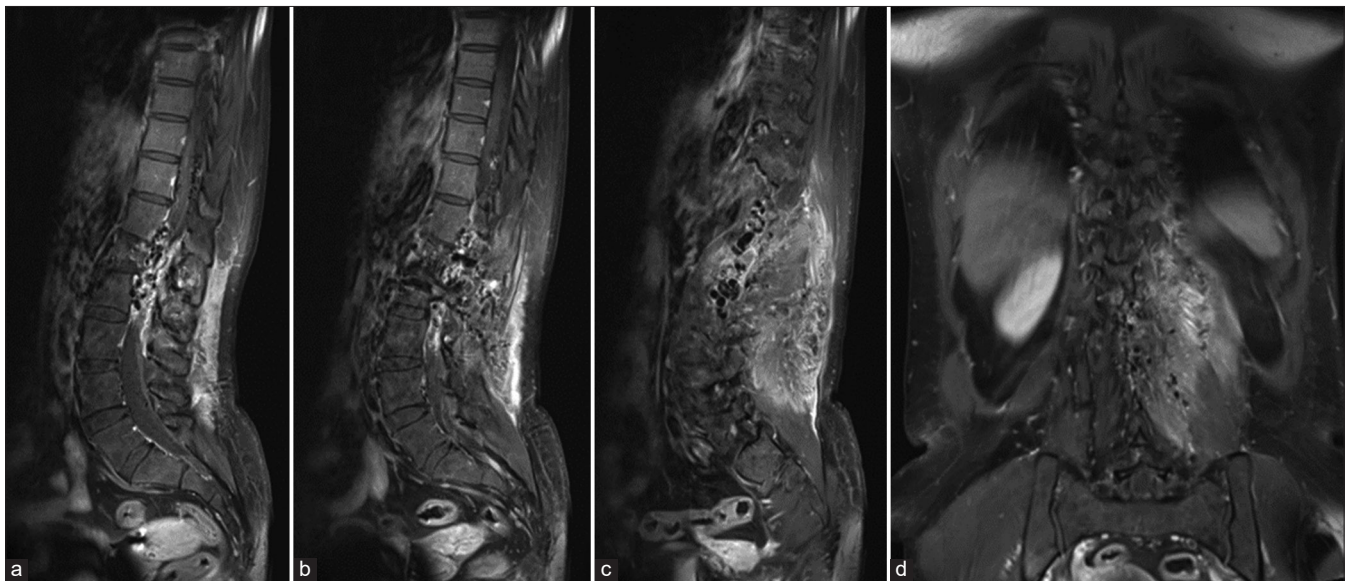


Figure 3: (a-c) Sequential sagittal and (d) coronal fat-suppressed contrast-enhanced T1-weighted images of the thoracolumbar spine illustrate associated intense enhancement and mild enlargement of epidural venous plexus, psoas, and paravertebral muscles from T12 to L4 vertebral levels.

with SAMS is mostly nidus-type SCAVMs.^[13] SAMS is rare nonhereditary genetic vascular lesions and account for 6.4% of all spinal arteriovenous lesions.^[16] There is significant female dominance in SAMS group, compared with non-SAMS. Intradural hemorrhage is the most common presentation and more common than in non-SAMS SCAVM. The recurrence rate of hemorrhage is 50%.^[13]

Due to the rarity and poorly understood pathophysiology of SAMS, optimal management remains unclear. Treatment options include endovascular treatment, surgical resection, a combined approach, or conservative management. Complete removal of SAMS is often difficult without significant risk of neurological morbidity. It is difficult that a complex SAMS could be cured by only endovascular treatment. For

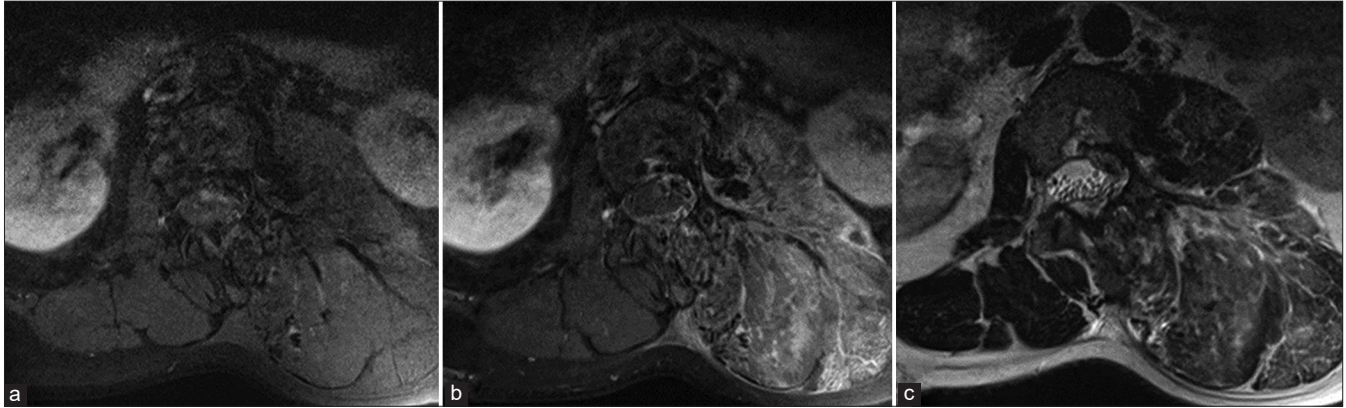


Figure 4: Axial T1-weighted (a) without and (b) with contrast-enhanced, and (c) T2-weighted images at the level of L2 vertebral body disclose multiple abnormal dilated and tortuous flow voids at left side of vertebral body, epidural space, and nearby paraspinal muscles with associated diffuse enlargement and enhancement of affected muscles.



Figure 5: Contrast-enhanced magnetic resonance angiography of the thoracolumbar spine. (a) Coronal and (b) sagittal maximum intensity projection images reveal clusters of vascular malformations including intradural spinal cord arteriovenous malformation (arrows) at the level of T10-11 and extradural paravertebral arteriovenous malformation from T12 to L2 vertebral levels with significant enlargement of bilateral T 12 intercostal, L1, and L2 segmental arteries, particularly on the left.

this reason, the aim of endovascular treatment is symptom improvement or stabilization of a progressive disease with poor long-term functional prognosis. Spinal angiography remains the gold standard to evaluate the angioarchitecture for therapeutic decisions. Careful assessment of symptoms of patients and causative vascular pathology is required to perform treatment at the appropriate time. Long-term

clinical follow-up and periodic spinal angiography are imperative to control this dynamic disease.^[2,5,13,17]

In the present study, SAMS included intradural SCAVM and extradural high-flow SOEAVF. Spinal epidural arteriovenous fistulas (SEAVFs) are likely acquired lesions and usually present with congestive myelopathy secondary to intradural venous drainage and may present with compressive symptoms resulting from exclusive extradural venous drainage.^[6] High-flow SOEAVF may be congenital in origin usually seen in young patient. This osseous fistula supplies mainly by intercostal or segmental arteries, from upper or lower segments of the ipsilateral and contralateral arteries, with associated small feeders converging into venous sac or lake at the level of the body defect.^[5,7]

Drake *et al.*^[4] reported a case of SAMS diagnosed with SCAVM and SOEAVF with shared common venous drainage. The patient manifested with acute worsening of chronic back pain and progressive neurological deficits from spinal venous congestion. She was successfully treated by targeted embolization of the extradural component led to dramatic improvement of the symptoms. Similarly, SAMS in the present study included SCAVM and SOEAVF but their drainage into different pathways without sharing. Due to asymptomatic SAMS and a high risk of anterior spinal arterial compromise during treatment, conservative treatment was considered in our patient.

To the best of our knowledge, spontaneous regression of SAMS has never been reported in the medical literature. Spontaneous regression of extradural high-flow vascular malformation in SAMS occurred in our patient during a long-term observation period. Based on the study of Kang *et al.*,^[9] they found that spontaneous resolution of spinal arteriovenous fistulas (AVFs) appeared to selectively involve low-flow fistulas, notably SEAVFs. When long-standing lesions disappeared shortly following an angiographic procedure, spinal angiography seemed to be a significant contributing factor, probably through

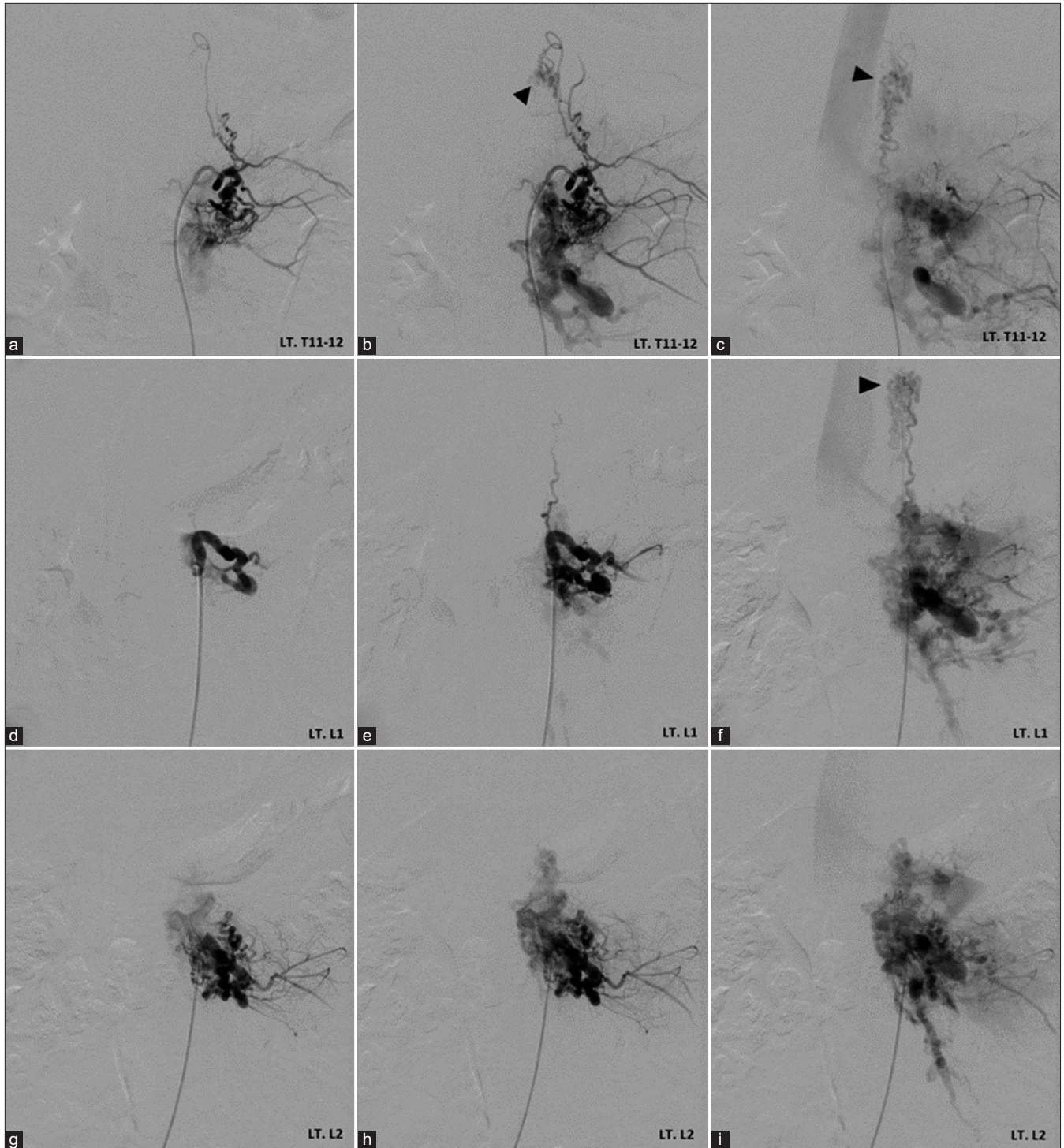


Figure 6: Anteroposterior views of the left (a-c) T11-12 intercostal, (d-f) L1, and (g-i) L2 segmental arteries angiography demonstrate spinal cord arteriovenous malformation (arrowheads) at the T10-11 level, supplied by left posterior spinal arteries that receives radiculopial branches from the left T11 intercostal and L1 segmental arteries, and spinal osseous epidural arteriovenous fistula fed by dilated radicular arteries from left T12 intercostal, L1, and L2 segmental arteries converging to the venous lake at the level of L1 and L2 with venous drainage through enlarged epidural and paravertebral venous plexus.

the prothrombotic properties of nonionic contrast material. However, the delay between the initial angiography and the study

subsequently documenting regression is too long to support this mechanism in high-flow SOEAVF in the present study.



Figure 7: Spinal angiography of (a and b) the right T11-12 intercostal and (c and d) L1 segmental arteries in arterial and venous phases reveals associated arterial feeders from contralateral T12 and L1 converging to the left dilated venous lake at the level of L1 and L2.

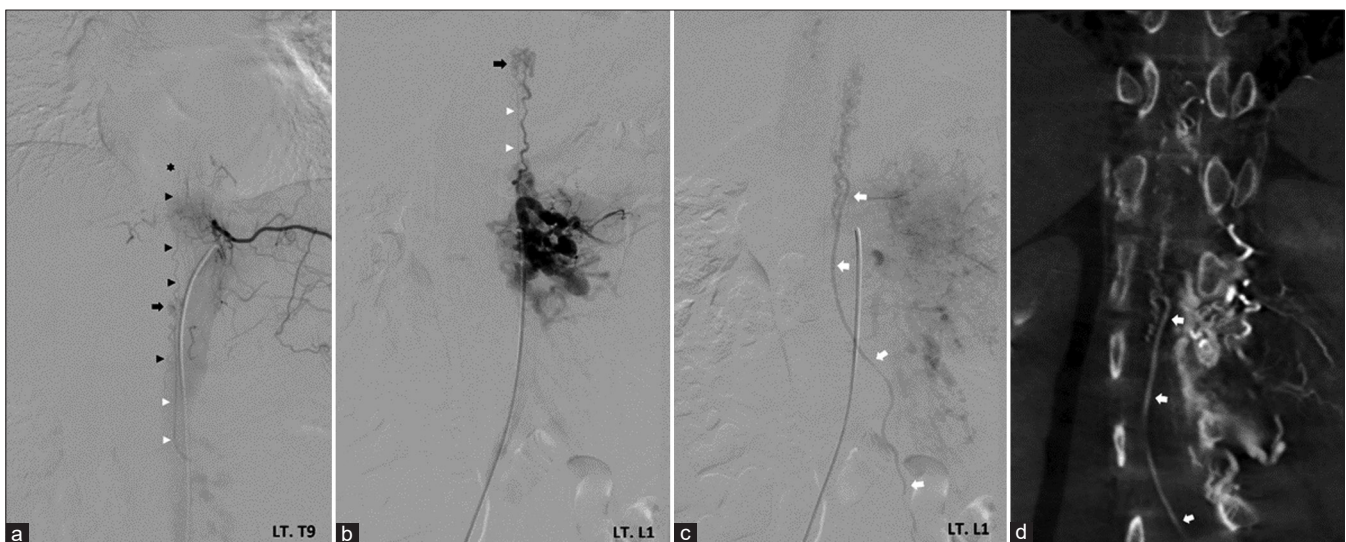


Figure 8: (a) Anteroposterior view of the left T9 intercostal artery angiography demonstrates spinal cord arteriovenous malformation (SCAVM) (black arrow) at the T10-11 level supplied by an anterior spinalartery (black arrowheads) with a characteristic hairpin turn (asterisk). Another feeding artery from the radiculomedullary branch (white arrowheads) from the left L1 segmental artery is identified. Anteroposterior views of the left L1 segmental artery angiography in (b) arterial and (c) venous phases reveal the same SCAVM (black arrow) fed by the radiculomedullary branch (white arrowheads) and dilated radiculopial artery. There is a high-flow spinal osseous arteriovenous fistula supplied by enlarged radicular arteries from the left L1 segmental artery with caudal drainage into perimedullary veins and subsequent drainage to the dilated left sacral radicular vein (white arrows). (d) Coronal multiplanar reconstruction image of angiographic computerized tomography clearly illustrates the enlarged left radicular vein (white arrows).

Spontaneous regression of high-flow spinal extradural AVFs remains unclear and has rarely been reported.^[10,12] Spontaneous obliteration or thrombosis of high-flow AVFs or AVMs may be induced by exposure of nonionic contrast agent during angiography, sudden postpartum changes in the circulatory system, changes in the concentration of circulating estrogen and progesterone, symptomatic hemorrhage resulting in compression of the draining vein, hypotension, severe dehydration, vascular

wall injury of draining veins, or the interaction of genetic and hemodynamic factors.^[8,10,12,14,15,18] In our case, the mechanism of spontaneous regression remains unknown.

Because the natural history of SAMs remains poorly understood. Therefore, further long-term clinical assessment and radiological follow-up in our patient are required to monitor the progression of intradural SCAVM and recanalization of extradural SOEAVF.

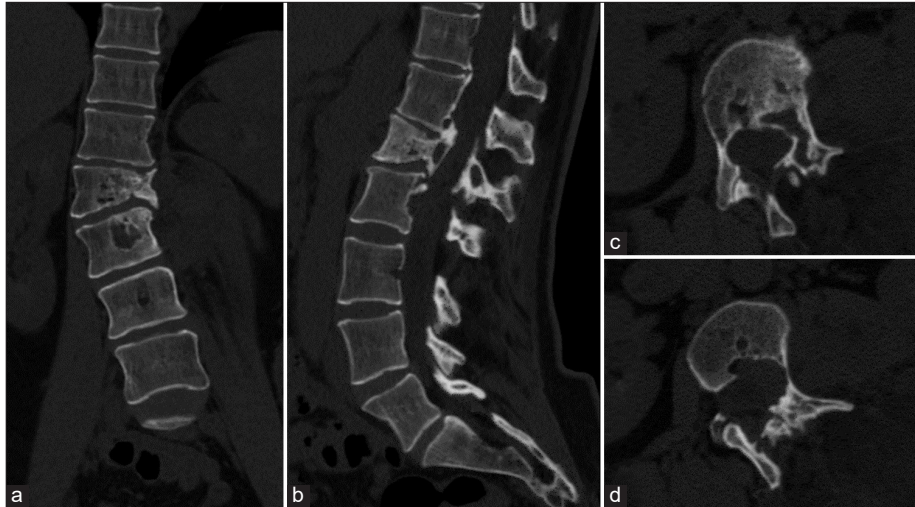


Figure 9: Computed tomography scan of the lumbar spine. (a) Coronal, (b) sagittal, and axial views at the level of (c) L1 and (d) L2 reveal dextroscoliosis and decrease of left sided L1 vertebral height with associated sclerotic change. There are multiple well-defined tubular and round-shaped osteolytic lesions and erosion with sclerotic rims involving particularly left-sided vertebral bodies and posterior elements of L1 and L2, corresponding with the dilated venous lake.

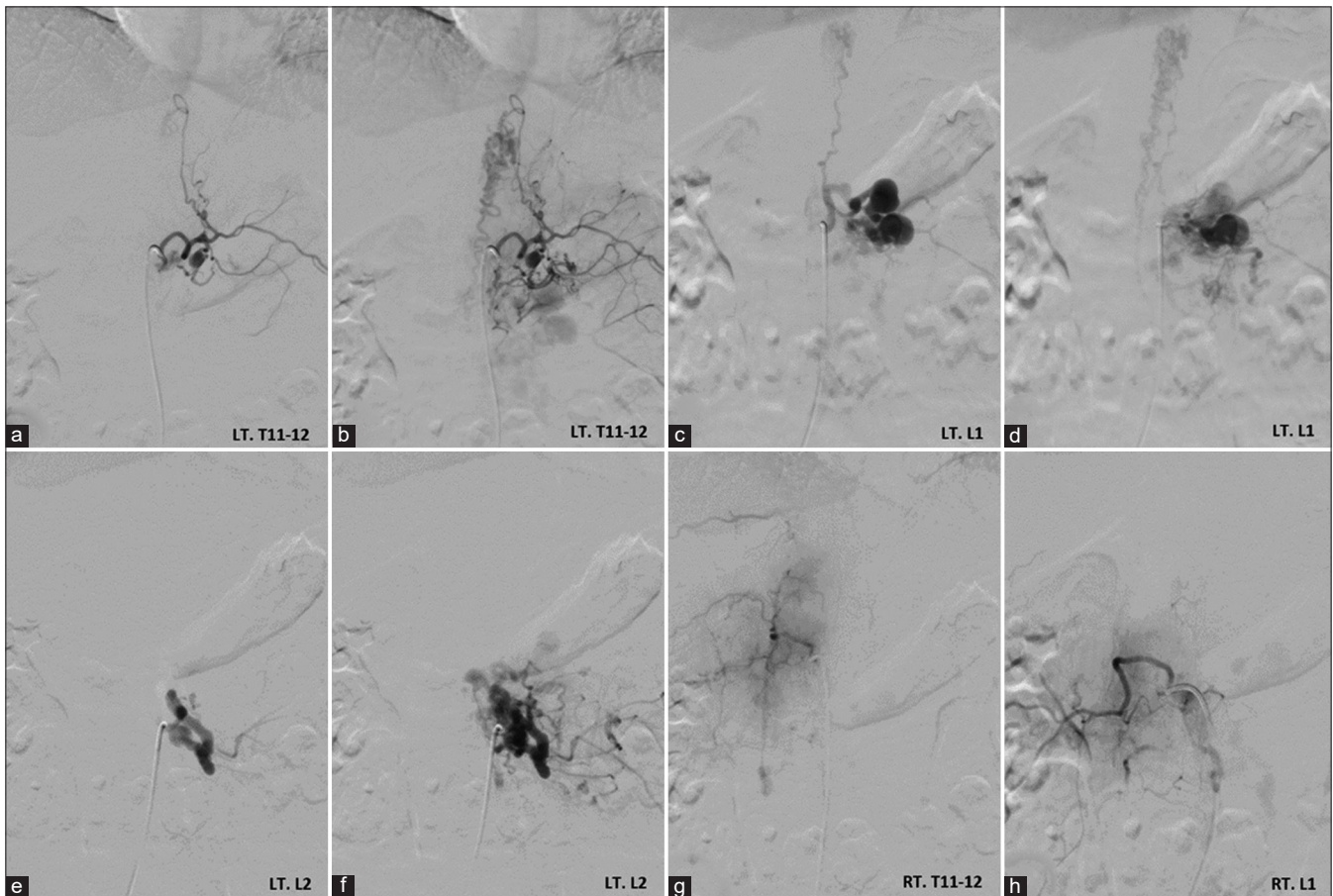


Figure 10: Spinal angiography obtained 8 years after initial angiography. Anteroposterior views of the (a and b) left T11-12, (c and d) left L1 segmental, (e and f) left L2 segmental, (g) right T11-12 intercostal, and (h) right L1 segmental arteries angiography demonstrate significant regression of the spinal osseous epidural arteriovenous fistula at the level of L1 and L2. Spinal cord arteriovenous malformation at the level of T 10-11 remains stable in size and angioarchitecture.

CONCLUSION

SAMS is a rare subtype of SCAVM. This syndrome occurs from a somatic mutation developing in the region of the neural crest or adjacent mesoderm before migration and differentiation, affecting the corresponding metameric distribution. We describe a unique case of SAMS with spontaneous regression of extradural component during a long-term observation period.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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How to cite this article: Iampreechakul P, Chuntaroj S, Wattanasen Y, Hangsapruet S, Lertbutsayanukul P, Siriwinonmas S. Spontaneous regression of extradural high-flow vascular malformation in spinal arteriovenous metameric syndrome (SAMS): A unique case report. *Surg Neurol Int* 2023;14:163.

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