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Case Report

# Intracranial subarachnoid hemorrhage as part of spinal arteriovenous metameric syndrome ☆,☆☆

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ABSTRACT

Spinal Arteriovenous Metameric Syndrome is a rare and complex nonhereditary genetic vascular disorder, affecting multiple layers of tissues at the same metamere, including the spinal cord. We present a case of a 20-year-old man who presented to the emergency department with sudden headache and transient loss of consciousness. Cranial computed tomography scan revealed subarachnoid hemorrhage predominantly in the cerebellar cisterns, fourth ventricle, extending to the basal cisterns. Cerebral angiography showed no abnormalities. Cervical angiographic acquisitions demonstrated a spinal metameric arteriovenous malformation (AVM) at the C3 and C4 levels. Cervical magnetic resonance imaging also confirmed the metameric AVM, revealing both intradural intramedullary and extradural vascular lesions in the vertebrae and adjacent soft tissues. The patient was referred for endovascular treatment. Although quite rare, the association between cervical spinal arteriovenous shunt diseases and intracranial hemorrhage has been reported. The bleeding in this case may be attributed to venous reflux into intracranial veins.



## Introduction

Vascular malformations of the spinal cord, including arteriovenous fistulas and arteriovenous malformations (AVMs), are rare lesions, accounting for approximately 3% to 4% of intradural spinal cord lesions [1]. They pose challenges both in terms of imaging and diagnosis as well as treatment, dating back to the initial classification scheme proposed by Di Chiro in 1971 [2].

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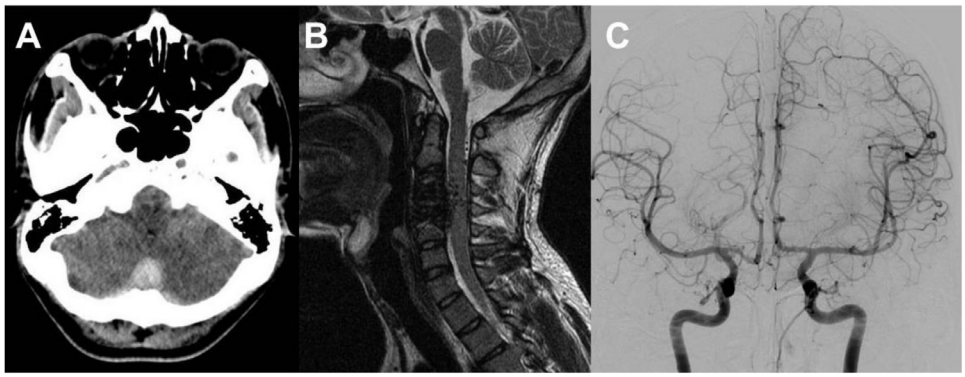


**Fig. 1 – Neuroradiological findings. (A) Axial noncontrast CT scan shows diffuse subarachnoid hemorrhage throughout the posterior fossa, clearly visible in the cisterna magna and peribulbar cistern. (B) Cervical sagittal T2-weighted MRI shows an abnormal vascular network in the C3 and C4 vertebrae with extension into the vertebral canal, involving the spinal cord. (C) Posteroanterior view of internal carotid artery angiograms shows no abnormality.**

Spinal Arteriovenous Metameric Syndrome (SAMS), also known as Di Chiro type 3, comprises high-flow lesions that involve both intradural and extradural tissues within the same metamere. There is a scarcity of publications in the medical literature regarding this subtype of vascular malformation, with reported cases not exceeding a hundred, given the rarity of this condition [1,3]. We present a case of a young patient with this rare type of malformation exhibiting atypical clinical manifestations.

### *Case report*

A 20-year-old previously healthy man presented to the emergency department with sudden headache, transient loss of consciousness, and neck stiffness. Upon initial medical evaluation, the patient was awake, Glasgow coma scale at 15 points, and without focal neurological deficits. Cranial computed tomography (CT) scan revealed subarachnoid hemorrhage (SAH) predominantly in the cerebellar cisterns, fourth ventricle, extending to the basal cisterns and Sylvian fissures (Fig. 1A). Cerebral angiography did not show any intracranial aneurysms or vascular malformations (Fig. 1C). Cervical angiographic acquisitions demonstrated a metameric AVM at the C3 and C4 levels (Fig. 2A-C), with an intra and extradural nidus.

Cervical magnetic resonance imaging confirmed the metameric pattern of the vascular malformation (Fig. 1B), involving the vertebrae and soft tissues at these levels. Additionally, a flow aneurysm was observed in a feeding segmental artery at the foraminal level of C4 (Fig. 2B).

Although the flow aneurysm was not the cause of the patient's intracranial hemorrhage, it was treated with microcoils and excluded from circulation. Two embolization sessions using ethylene vinyl alcohol (EVOH-18) embolic liquid were performed (Fig. 3A and B). There was significant devascularization of the lesion, but complete exclusion of the arteriovenous shunt had not yet been achieved. In 1 session, vascular access was through a segmental feeding branch of the right vertebral artery, and in the other session, it was through the left costocervical arterial trunk.

The arteriovenous shunt persists, fed by the anterior spinal artery originating from the intracranial segment of the left vertebral artery. The safe and minimally viable caliber arterial feeder for a subsequent embolization session was not found. The patient has been experiencing mild paresis in the upper limbs since the last session, with slow and insidious progression. Rebleeding has not been experienced since the initial event.

## Discussion

In 1971, Di Chiro and colleagues [2] proposed a classification that brings together angioarchitectural and epidemiological characteristics similar in each group of spinal cord arteriovenous shunt diseases, classified as follows:

- Type 1: Dural arteriovenous fistula.
- Type 2: Intramedullary AVM.
- Type 3: Metameric AVM.
- Type 4: Intradural perimedullary fistula.

SAMS, also referred to as Di Chiro's type 3, juvenile, extra-intradural, or Cobb Syndrome, encompasses all spinal vascular malformations of metameric, genetic, and nonhereditary origin, affecting not only the spinal cord but also other tissues derived from the same metamere, such as nerves, skin, bones, and blood vessels [4,3].

During the early stages of embryogenesis, cell proliferation occurs in the central portion of the neural plate. These cells then migrate laterally between the embryonic ectoderm and endoderm to form the mesoderm. The mesoderm further differentiates into segments called somites, which give rise to the sclerotome, myotome, and dermatome. Mutations in the mother cells at this stage can cause malformations in various tissues in the same distribution pattern [3,4].

Affected patients are typically adolescents or young adults, with a slightly higher occurrence in males. Approximately one-third present with progressive neurological deterioration, another third with hemorrhage, roughly one-fifth with acute deficits not attributable to hemorrhage, and the remaining

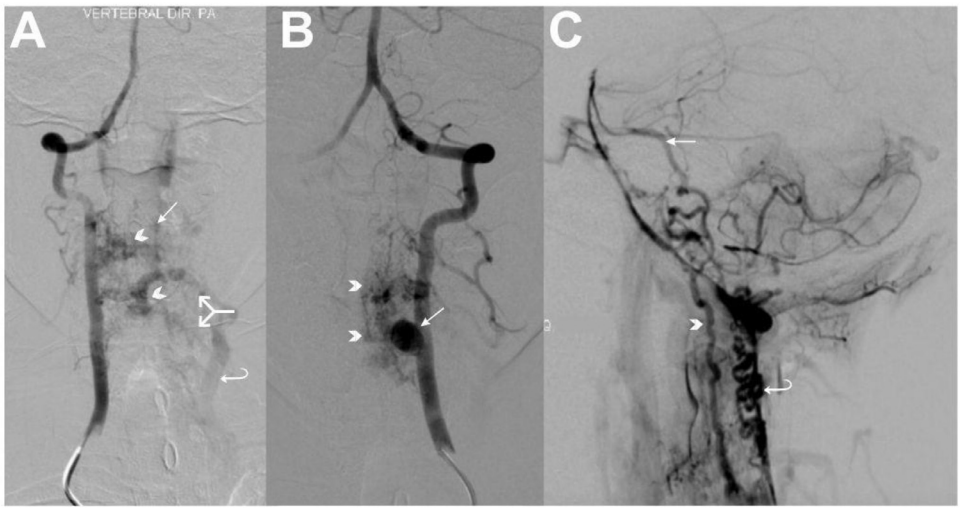


**Fig 2 – Digital subtraction angiography (DSA) of the vertebral arteries. (A) Posteroanterior view of right vertebral artery DSA shows opacification at the C3 and C4 levels of intramedullary and extramedullary nidal elements (arrowheads) and early filling of a left epidural venous plexus (arrow), foraminal veins (double arrow), and left vertebral vein (curved arrow). (B) Posteroanterior view of left vertebral artery DSA shows opacification at the C3 and C4 levels of intramedullary and extramedullary nidal elements (arrowheads). A flow aneurysm is observed at foraminal level C4 (arrow). (C) Profile view of left vertebral artery DSA, in the late arterial phase, shows early filling of tortuous intracranial and spinal veins with ectasia: anterior spinal vein (arrowhead), posterior spinal veins (curved arrow), lateral mesencephalic vein (arrow).**

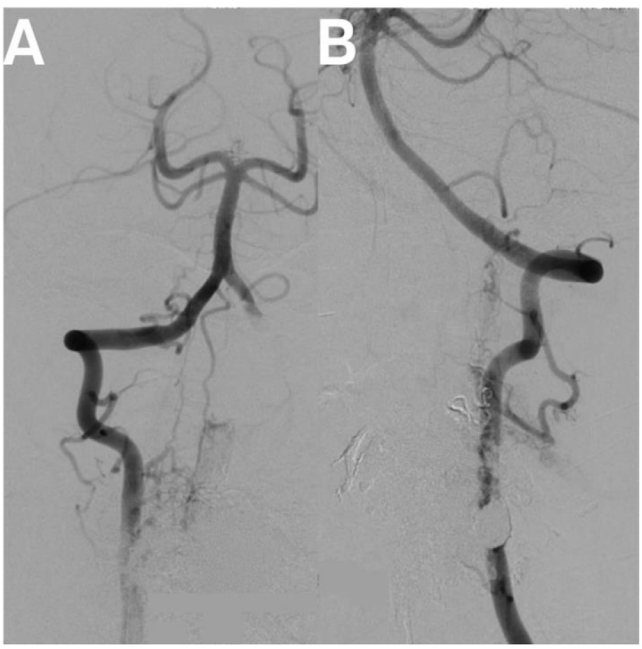


**Fig. 3 – Angiographic follow-ups after the last embolization session. Note the embolization material from the previous session filling and excluding most of the malformation, but with the persistence of the malformative nidus. (A) Posteroanterior view of right vertebral artery DSA. (B) Oblique view of left vertebral artery DSA.**

with incidental lesions [5,6]. In Niimi's series, [7] the most common manifestation was intradural hemorrhage (64%- 18 out of 28 patients).

The manifestation of intracranial SAH in our patient's case is indeed very uncommon and may be attributed to intense venous reflux into intracranial veins clearly visible on angiography (Fig. 2C). Intracranial SAHs have been previously described in cases of perimedullary spinal dural fistulas [8].

Vascular investigation of the cervical spinal cord should be performed in cases of intracranial SAH and absence of intracranial vascular lesions.

SAMS lesions are high-flow and typically have multiple feeding vessels originating from various sources, such as vertebral arteries, radicular arteries, and other cervical vessels. They are extensive lesions, and thus, catheter angiography of a metameric AVM shows only a portion of the lesion in each studied area, as depicted in Fig. 2 [6].

Another interesting aspect in these cases is the association with flow aneurysms, as evident in the presented case (Fig. 2B). Niimi and colleagues [7] reported intradural medullary aneurysms in 46.4% (13 out of 28) of SAMS patients, while Gross and colleagues [6] reported 48% (11 out of 23 SAMS patients) having such association.

These aneurysms may account for the high rates of hemorrhage observed in SAMS patients. Although the flow aneurysm in our patient was not the cause of the hemorrhage as it was foraminal in projection. Flow aneurysms, whether intra- or extradural, are not commonly observed in other types of spinal vascular malformations.

Metameric spinal AVMs are extremely challenging to treat. Endovascular treatment through the injection of embolic agents appears to be the most adopted approach in the re-

ported cases. Complete surgical resection of the lesion following staged embolization has also been reported. The treatment goals involve disconnecting feeding pedicles, thereby minimizing vascular steal, venous hypertension, and aiding in decompression of mass effect along nerves, roots, and spinal cord. A cure is not expected through current methods due to the complexity of the lesions [5–7,9].

While the patient presented here did not have skin hemangiomas, as seen in the classic Cobb Syndrome [10], the recognition of metameric cutaneous lesions is essential. Further vascular investigation at the dermatomal level should be conducted with the aim of diagnosing and treating this rare and aggressive subtype of spinal vascular malformation in young patients at an early stage.

## Patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given him consent for his images and other clinical information to be reported in the journal.

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