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Spinal Epidural Hematoma Caused by Pure Epidural Spinal Arteriovenous Malformation: Case Report and Literature Review

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Abstract

Spontaneous spinal epidural hematoma (SEH) represents an extremely rare cause of spinal cord compression. Symptomatic pure extradural spinal AVMs (E-sAVM), in the absence of cavernous hemangiomas, are very rare and have rarely been reported. The clinical presentation of SEH caused by E-sAVM is often nonspecific and may lead to delayed diagnosis and treatment. We report the case of a 16-year-old adolescent girl who presented with paraparesis that rapidly evolved in paraplegia. Emergent magnetic resonance imaging (MRI) of the whole spine showed a posterior SEH, extending from C7 to T2, highly suspicious for the presence of an underlying AVM. The patient underwent emergent C7–T2 laminoplasty. An E-sAVM was intraoperatively found and subsequently excised. The patient was discharged with no neurological defects. E-sAVMs are extremely rare pathologies; they represent an extremely rare cause of spinal cord compression. If immediately diagnosed and treated, most patients recover with good prognosis.

Keywords
- epidural spinal hematoma
- spinal epidural vascular malformation
- spinal digital subtraction angiography

Introduction

Spinal epidural hematoma (SEH) is usually a condition observed in traumatic or oncologic contexts, drug abuse, anticoagulation therapy, blood dyscrasia, or as a complication after a lumbar puncture. In the literature, the term "spontaneous spinal epidural hematoma" is used to describe SEH without clear traumatic etiology.[1,2] Spontaneous SEH (SSEH) represents an extremely rare cause of spinal cord compression with approximately an incidence of 0.1 per 100,000 per year.

Spinal arteriovenous malformation (sAVM) is a comprehensive term that groups different spinal vascular lesions located within the spinal canal.[3,4] sAVMs are identified annually in 1 in 1 million patients.[5,6] The majority of spinal AVMs are intradural (~70%)[7–9]; epidural AVMs are rare and usually are found with an intradural vascular component.[10,11]

Pure extradural spinal AVMs (E-sAVMs), in the absence of vertebral body hemangiomas, are uncommon with only few cases reported in the literature; they account for 20% of all the spinal vascular malformations and approximately 5 to 9% of all vascular malformations affecting the central nervous system. In E-sAVM, the shunt is exclusively in the spinal epidural space and drains into the epidural venous plexus (intervertebral veins).[12]

Most cases of E-sAVMs present as SSEH (65%); however, pure E-sAVMs as cause of SSEH is even more rare and there are only few case reports in the literature.[13,14]

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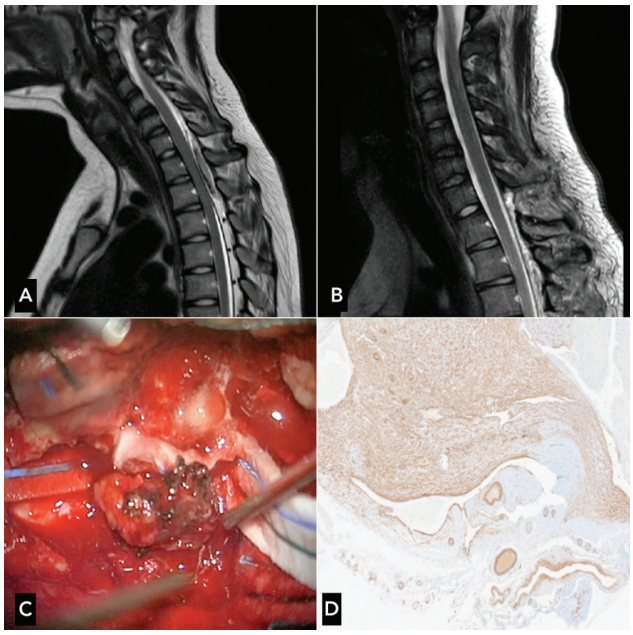

Fig. 1 (**A**) Preoperative magnetic resonance (MR) of the cervical spine, T2-weighted sequence, sagittal plane showing posterior epidural collection of blood at the C7–T2 level with cord compression. (**B**) Postoperative MR of the cervical spine, T2-weighted sequence, sagittal plane, showing full expansion of the dural sac after complete removal of the cervical epidural hematoma without cord abnormalities. (**C**) Intraoperative image of abnormal compressive and bleeding nidus tissue found after laminotomy and decompression at C7 and T2. (**D**) Scanning magnification shows irregularly dilated and proliferated vessels; the cells of the vessel wall were positive for smooth muscle actin (SMA; 4 ×).

Case Presentation

We present the case of a 16-year-old Caucasian adolescent girl who was referred to our Department of Neurosurgery with sudden onset of back pain associated with rapid paraplegia. She had no oncological history or previous trauma, and the coagulation profile was normal. Neurological examination revealed paraplegia with sensory level below T1 and neurological bladder, Babinsky's sign, and hyper-elicitability of deep reflexes of the lower extremities.

Magnetic resonance imaging (MRI) scan showed a posterior epidural hematoma (►**Fig. 1A**), extending from C7 to T2, displacing the spinal cord. The patient underwent emergent C7–T2 decompression. After the laminotomy, a wide epidural blood clot was encountered; once removed, we found an active bleeding from a congested and tortuous venous drainage (►**Fig. 1C**). The arterial feedings were deriving from multiple posterior dural branches. After the coagulation of the major arterial feeding, the bleeding stopped and subsequent dissection and removal of the AVM nidus was uneventful. At the end of the nidus dissection, the dural sac was normally expanded, with physiological pulsation and with no evidence of dural involvement.

The postoperative MRI showed complete evacuation of hematoma and no residual vascular malformation (►**Fig. 1B**); the spinal angiography confirmed complete resection of the AVM. The patient rapidly improved, recovering motor and sensory abilities, and the neurogenic bladder completely recovered in 3 days. She was discharged 1 week after the operation without neurological defects. Histological examination confirmed the diagnosis of pure E-sAVM (►**Fig. 1D**).

Discussion

The etiopathogenesis of SSEH is not always clear and approximately 40% of cases remain undiagnosed; bleeding predisposition is the most common risk factor.[7,8] The sources of bleeding can be venous, when the epidural plexus is involved, arterial, or mixed, in case of vascular malformation or neoplasms.

The most frequent location of an SSEH is where radicular arteries are more prominent: lower cervical region in children and adolescents and thoracic or thoracolumbar regions in adults. Age distribution shows two peaks: between 15 and 20 years and 65 and 70 years.[3,9]

From a clinical point of view, SEH can be misdiagnosed and, even when adequately diagnosed, its origin is often misunderstood.[5,11] The clinical presentation is often nonspecific and may lead to delayed diagnosis and treatment. SEH usually manifests with acute onset of back pain and radiculopathy, followed within hours by myelopathy with paraparesis/paraplegia, although nonspecific or even deceiving clinical signs and symptoms have also been described such as irritability and excessive crying in children.

Differential diagnosis includes intrinsic or extrinsic cord tumor, minor trauma, spinal abscess, spinal cord ischemia, disk disease, Guillain–Barré syndrome, transverse myelitis, and congenital abnormalities such as a syringomyelia, especially in children.

Pure E-sAVMs had been described by Spetzler et al.[13] The authors proposed a reclassification of spinal cord vascular lesions to include extradural variants. These extradural lesions may cause myelopathy through a combination of different potential mechanisms including compression by dilated venous channels, venous congestion, "vascular steal," and conceivably hemorrhage.

In ►**Table 1**, we collected the cases of ruptured E-sAVMs published in the medical literature. There is slight male predilection (57%), and age distribution by the clinical onset shows most patients are younger than 30 years (92,8%). As in our case, the majority of those lesions are located on the cervicothoracic junction or upper thoracic segment (71.4%).

MRI is fundamental in the diagnostic workup of epidural compression because it can detect tortuous or dilated vessels, giving rise to the suspicion of an underlying sAVM. Finding a hemangioma in the adjacent vertebral body could be helpful in suspecting sAVMs.

Spine digital subtraction angiography (DSA) remains the gold standard to diagnose and characterize spinal vascular lesions and should be recommended in the diagnostic workup when E-sAVM is suspected. However, as in our case, it is not always performed before surgery. Moreover,

Table 1 Exclusively epidural spinal arteriovenous malformations

Study	Age/sex	Level	Clinical presentation	Comorbidity	Treatment	Imaging outcome	Clinical outcome
Sharma et al[7]	50/F	T1–T2	Paraplegia	–	AVM resection	No residual	Complete recovery
Muhonen et al 1995[15]	2/M	C7–T2	Paraparesis in the legs	–	SSEH evacuation and AVM resection	No residual	Complete recovery
Miyagi et al[8]	16/F	C2	Neck pain, complete quadriparesis, and hypesthesia below both shoulders	–	SSEH evacuation and AVM resection	No residual	Complete recovery
Nadig et al 2000[16]	10/F	L3–L5	Abnormal posture	–	AVM resection	No residual	Complete recovery
Rohany et al 2007[17]	29/F	C6–T1	Right upper extremity weakness and numbness	–	Embolization and AVM resection	Unknown	Unknown
Rispoli et al[6]	14/F	C4–C6	Intractable cervical neck pain	–	SSEH evacuation and AVM resection	No residual	Complete recovery
Fairhall et al 2010[18]	22/M	T6–T8	spastic paraparesis	–	Embolization and AVM resection	No residual	Complete recovery
Cabral et al[9]	9/M	C7–T4	Motor weakness in the lower limbs; absent reflexes, cervicodorsal pain	–	SSEH evacuation and AVM resection	No residual	Complete recovery
Paraskevopoulos et al[5]	8/M	C6–C7/T2	Mimicking GBS; weakness in the lower limbs	–	SSEH evacuation and AVM resection	No residual	Motor partial recovery; ambulating with support
Elkordy et al[2]	15/M	T1–T7	weakness in the lower limbs; difficulty in walking and urinating	–	Embolization and AVM resection	No residual	Complete recovery
Sivakumaran et al 2016[19]	8/F	C5–C7	Paraplegia	–	SSEH evacuation and AVM resection	No residual	Complete recovery
Wang et al[3]	13/M	T1–T5	Interscapular pain and paraplegia	–	SSEH evacuation and AVM resection	No residual	Improved but intermittent urinary continence
Wang et al[3]	13/M	C7–T2	Complete paraplegia and mimicking transverse myelitis; Babinski's positive	–	SSEH evacuation and AVM resection	No residual	Transitory bladder disfunction; then complete recovery
Yakar et al[11]	29/M	T12–L1	Left leg pain	–	AVM resection	No residual	Complete recovery
This study	16/F	C7–T2	Paraplegia with sensory level below T1	–	SSEH evacuation and AVM resection	No residual	Complete recovery

Abbreviations: AVM, arteriovenous; GBS, Guillain–Barré syndrome; SSEH, spontaneous spinal epidural hematoma.

some lesions can be first identified during surgery, even though the preoperative angiography was found negative.

Because of E-sAVMs' extreme rarity, there is no standardized treatment and complete surgical resection can be difficult with potential risk of neurologic morbidity. However, E-sAVM niduses are located in the epidural space without spinal cord involvement; this is the reason why the effect of surgical/intravascular treatment and the outcome are better than spinal cord AVMs.

Elective treatment of unruptured E-sAVMs is debatable and includes surgical, endovascular or conservative management. Embolization could have a crucial role for devascularization of the feeding arteries to get a safer surgical removal.

Most lesions are accessible through a posterior laminectomy or laminotomy and partial facetectomy since they are mostly located in the posterolateral aspect of the spinal epidural space.

SEH caused by E-sAVM usually requires emergent surgery because of neurological deficits on onset. Long term functional outcome is generally good with complete recovery in 78.6% of cases, but is correlated with the rapidity of decompression and severity of the preoperative neurological deficits. Postoperative spinal DSA is mandatory to discover residual AVMs or other associated vascular lesions. Although small SEHs could resolve spontaneously with conservative treatment, DSA should be obtained at follow-up to rule out the possibility of repetitive hemorrhage from misdiagnosed AVMs.

Conclusion

E-sAVM is a rare, disabling, or even fatal entity that has to be suspected in case of SSEH. Spinal DSA is the preoperative gold standard examination.

If promptly treated, patients with E-sAVM can achieve good neurological and radiological outcomes, which can be equally good as in other patients with SEH.

Conflict of Interest
None declared.

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