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%), epidural AVMs are rare and usually are found with an intradural vascular component.7,10

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

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.

**References**