Exclusively Epidural Spinal Arteriovenous Malformation: A Short Review

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Abstract
The incidence of exclusively epidural spinal arteriovenous malformation (EESAVM) is extremely low and there are only a few case-reports in literature. Early, correct recognition of the pathology is mandatory to halt the progression of the disease and minimize permanent spinal cord injury. This review depicts EESAVM’s characteristics, from the aspect of pathophysiology, clinical presentation, treatment strategies, and outcome. EESAVMs are located entirely in the epidural space and fed by radicular vessels or segmental arteries. The nidus of EESAVM purely locates in the spinal epidural space. The drainage of nidus flows into the epidural venous plexus or intradural vein. The retrograde blood flow from the AVM results in a diminished intramedullary blood flow and symptoms due to spinal cord ischemia and myelopathy. There is no gender predilection (male 52.9%), age distribution by the time of diagnosis shows most cases are younger than 20-year (64.7%), most cases present as spontaneous epidural hematoma (64.7%), and the majority of those lesions are located on cervicothoracic junction or upper thoracic segment (75%). Generally, the clinical presentation of EESAVM is slighter than intradural/intradural AVMs. Unruptured EESAVM may symptomless, or present as protracted, progressive neurological decline. Ruptured EESAVM may present acute back or thoracic pain subsequently with paraplegia. Epidural haemorrhage is common and urgent condition for EESAVM. Up to now, spinal angiography remains the gold standard and the first choice for diagnosis and characterization of spinal vascular lesion. The best management for EESAVM is still surgical operation. Prompt diagnosis and emergency surgical treatment are crucial. Long-term functional prognosis of EESAVM is good, but delayed surgical operation leave residual symptoms.

Keywords: Epidural, Spinal arteriovenous malformation, Hematoma, Angiography

Introduction
Spinal vascular malformations are rare lesions; they can be localized in the spinal cord, intradural extra medullary space, epidural space, bone, Para spinal soft tissues or muscles [1-3]. Spinal vascular malformations comprise only approximately 5% to 9% of all vascular malformations affecting the central nervous system [4]. The incidence of brain arteriovenous malformations (AVMs) is 1.34 per 100,000 persons, and spinal AVMs are identified annually in approximately 1 in 1 million patients [5,6]. Although the pathology and biology of spinal AVMs are similar to their counterparts within the brain and cranial meninges, their natural history and response to treatment have been comparatively worse. More than 48% of patients with untreated AVMs of the spinal cord were confined to bed or wheelchair within 3 years of symptom onset, and complications of
chronic paraplegia were directly responsible for a mortality rate of 15% [7,8].

However, the incidence of exclusively epidural spinal arteriovenous malformation (EESAVM) is extremely low and there are only a few case-reports in literature. Whether the EESAVM is different from other spinal AVM in its clinical presentation, natural history, and vascular anatomy has never been revised. Early, correct recognition of the pathology is mandatory to halt the progression of the disease and minimize permanent spinal cord injury. The goal of this review is to determine EESAVM’s characteristics, from the aspect of pathophysiology, clinical presentation, treatment strategies, and outcomes.

Spinal vascular anatomy

The arterial supply to the spinal column, dura, nerve roots and spinal cord derives from the segmental arteries. However, the blood supply of cervical spinal cord varies among individuals; they may derive from the vertebral arteries, the deep and ascending cervical arteries, the ascending pharyngeal and occipital arteries. In the thoracic spine, several segmental arteries may arise from a common origin (the supreme intercostal artery) above T2. In the rest levels of the thoracic spine, ten pairs of the segmental arteries (nine pairs of posterior intercostal arteries and one pair of subcostal arteries) originate from the descending aorta. In upper lumbar spine, there are four pairs of lumbar segmental arteries arising from the descending aorta. In the lower lumbar and sacral region, segmental arteries originate from branches of the internal iliac artery and the median sacral artery, providing blood supply to the L5 vertebra and the sacrum. There are extensive anastomoses between segmental arteries with important connections both above and below a given level, as well as contralateral [9]. The segmental arteries divide into three major trunks: lateral or ventral trunk, middle or dorsal trunk, medial or spinal trunk. The spinal trunks of each segmental artery come into the spinal canal through the intervertebral foramen and divide into: radicular artery, anterior and posterior spinal canal arteries. The majority of the radicular arteries regress during development, with an average number of six still present in adult life in an unpredictable pattern. At several levels, radiculomedullary arteries derived from the radicular artery follow the anterior and/ or posterior nerve roots to supply the spinal cord. The radiculomedullary arteries are further divided into anterior radiculomedullary arteries (average number is 7-8, range from 2 to 15), and posterior radiculomedullary arteries (average number is 15, range from 10 to 25) [10].

There are two arterial systems on the surface of the spinal cord, the longitudinal arterial trunks that extend along the long axis of the spinal cord and the pial plexus that covers the periphery of the spinal cord. The longitudinal arterial trunks consist of the anterior spinal artery (ASA) and posterior spinal artery (PSA). The ASA arises from the vertebral arteries and extends from the level of the foramen magnum to the tip of the conus medullaris. Along its long course, additional blood supply to the ASA originates from anterior radiculomedullary arteries. Blood supply to the PSA via radiculomedullary arteries originates from three major regions: cervicothoracic, midthoracic and thoracolumbar [9-13]. ASA supplies the ventral surface of the medulla and the anterior two-thirds of the spinal cord [14]. The two PSAs originate from the ipsilateral posterior inferior cerebellar arteries or vertebral arteries at the level of the foramen magnum. The PSAs travel along the right and left posterolateral surface of the spinal cord. Along its long course, additional blood supply to the PSA originates from posterior radiculomedullary arteries at various levels. The PSAs supply the posterior one-third of the spinal cord, including the dorsal surface of the medulla, posterior columns and dorsal gray matter [9,13]. Pial plexus formed by effective anastomoses, which directly connects the ASA and PSA at the conus medullaris, is an extensive arterial network on the entire surface of the spinal cord.

The spinal cord intrinsic arterial system can be divided into the central system and the peripheral system. The central arteries derived from the ASAs constitute the central system, which travel into the anterior median fissure, penetrate into the spinal cord, branch within the gray matter centrifugally and vascularize the majority of the gray matter. The peripheral system consists of small perforators that originate from the pial plexus. These small arteries penetrate into the spinal cord centripetally and vascularize the periphery of the spinal cord [9,13].

The venous system of the spinal cord consists of intrinsic and extrinsic systems. The variety of the venous system is even greater than the arterial system. The intrinsic venous system consists of central and peripheral veins. Generally, the
central veins collect blood from the center of the spinal cord (gray matter) and the peripheral veins collect blood from the periphery of the spinal cord (white matter) [9].

At the level of the spinal pia mater, blood is accumulated in the anterior and posterior spinal veins. The anterior median spinal vein accompanies the ASA, receiving blood from the central veins and the veins of the ventral fissure. There may be as many as three posterior spinal veins, one posterior median vein located in the posterior median sulcus, and a pair of posterolateral spinal veins accompanying each PSA. The posterior spinal veins receive blood supply from the veins of the dorsal spinal cord [9,15]. The superficial veins surrounding the spinal cord are drained by radiculomedullary vein. In contrast to the arteries, these veins do not always exit the dural tube with the nerve roots. In the intervertebral foramen, the radiculomedullary veins join the epidural veins (the internal vertebral plexus), which have the segmental communications with external vertebral plexus. A functional valve at the level of the dura that consists of an oblique, zigzag course of the vein coupled with a narrowed lumen prevents reflux from the epidural veins into the intradural veins [9,15]. The vertebral venous plexus is a valveless system along the length of the spinal cord. The external vertebral plexus eventually joins the caval system; mainly the innominate veins at the cervical level, the azygos vein at thoracic level and the ascending lumbar vein at lumbar level [15,16].

Historical perspective

Before the advent of modern angiographic techniques, it is nearly impossible to detect or treat an EESAVM. With the introduction of the Seldinger technique in 1953, the procedure of angiography became markedly safer and wider use. In this setting, Newquist reported the first case report of an EESAVM in 1960; the surgeon operated on a patient with a spinal vascular abnormality, subsequently pathologist described the epidural lesion as: The vascular tissue shows a significant change in the form of degeneration of the walls. A number of vessels show only basophilic degeneration of the adventitia and muscularis. In other areas the entire wall of the vessel is involved in such a degenerative process. At one end of the specimen the vascular channels can be identified as such only because of their characteristic shape and are arranged in a noncommunicating network. Both arterial and venous channels appear to be involved in this process, impression: Arterial angioma [17]. Although it was recognized as arterial angioma, in the setting of 1960’s, various vascular malformations were categorized as angioma, actually the pathological description were consistent with the diagnosis of AVM.

In 1966, Hoffman was the first to report EESAVM confirmed by angiography [18]. The so called modern era in the treatment of spinal vascular malformation began in 1969 with Krayenbuhl and Yasargil and the publication of their microsurgical techniques, based heavily on the use of the operating microscope and bipolar cauterity [19-21]. After that time, a few cases were reported. In 1977, Kendall reported the largest case series of EESAVM, 10 cases of EESAVM. But all the reported cases in his series were older than 50 years, and some cases were difficult to find the nidus on angiography, when it was fed by a single artery drained by a single vein and when the vessels constituting the nidus itself were not clearly defined [22]. AVM is congenital lesion presenting symptoms at early age, and limited to the understanding of the difference between arteriovenous fistula and AVM at the time of 1970’s, some cases reported by Kendall might actually be arteriovenous fistula.

Because of the rarity of this entity, the widely-used classification of spinal AVMs has not categorized it separately [3]. But it is widely accepted that the nidus of EESAVM should exactly located in the epidural space, venous drainage of the nidus may be drained into spinal cord or not [23-29]. The EESAVM should not involve bone, muscle, skin, spinal cord, because complete involvement of an AVM along an entire somite level has been described as Cobb’s syndrome [3].

Pathophysiology

EESAVMs are located entirely in the epidural space and fed by radicular vessels or segmental arteries [27,28]. This concept is supported by the observations that spontaneous epidural hematomas commonly occur in the lower cervical and thoracic spine, where radicular arteries are prominent [17,23-25,27,29-33]. The nidus of EESAVM purely locates in the spinal epidural space. The drainage of nidus flows into the epidural venous plexus (intervertebral veins).
These intervertebral veins empty into segmental veins which drain into the ascending lumbar and azygos venous systems [9]. Generally, the venous drainage of the radicular vein pierce the dura into epidural venous plexus, and there is no retrograde reflux from the epidural vein to the intradural vein by a functional valve at the level of the dura that consists of an oblique, zigzag course of the vein coupled with a narrowed lumen [9]. However, in pathological situations demonstrated experimentally and by phlebography, such reflex occurs because of the destruction of the venous valve [34]. The retrograde blood flow from the AVM results in spinal cord coronal venous plexus hypertension and increased pressure in the part of the coronal plexus receiving blood from the AVM, and causes venous dilatation, congestion and stagnation in the radial veins draining from the spinal cord into it. This in turn reduces the arteriovenous pressure gradient between the normal arteries supplying this region of the spinal cord and the affected veins and capillaries, resulting in a diminished intramedullary blood flow and symptoms due to spinal cord ischemia and myelopathy [22,35]. The increased venous pressure and hypoxia could, in addition, cause edema which may also produce symptoms. This hypothesis was supported by spinal cord ischemia of a 13-year old boy with EESAVM [23]. Three other physiological mechanisms have been proposed to explain the neurological deterioration: hemorrhage, vascular steal, and mass effect [21]. Epidural hemorrhage is common and urgent condition for EESAVM, which is supported by the observations that spontaneous epidural hematomas commonly reported in the EESAVM patients [23-29,31,32,36]. Mass effect can occur with massively dilated vein, especially when the arterialized vein occluded. Arterial ischemia due to “steal” has been a popular explanation but is unlikely to be the cause in the great majority of cases because the blood supply to the spinal cord itself is entirely separate from that of the AVM [22].

Imaging

Before the introduction of spinal angiography, myelography aided significantly in the diagnosis of spinal AVM. Unfortunately, myelography was unable to provide insights into the angioarchitecture of these lesions and therefore little knowledge was added to their pathophysiology [37]. The development of spinal angiography in the 1960s revolutionized the understanding of the spinal vascular malformations; spinal angiography became a critical diagnostic and research tool to identify EEAVMs [37]. In 1965, Doppman and his college emphasized the importance of the subtraction technique in highlighting the details of the spinal vascular lesions [38]. From 1960s, spinal angiography has been the gold standard for diagnosing and analysing spinal vascular lesions.

Up to now, spinal angiography remains the gold standard and the first choice for diagnosis and characterization of spinal vascular lesion. Spinal digital subtraction angiography is superior to magnetic resonance angiography (MRA) and computerized tomography angiography (CTA) for precisely depicting involved vessels and the characteristics of AVM vascular architecture. Spinal angiography (even the super selective angiography) should be recommended when the EESAVM is suspected. Some lesions can be identified during the hematoma resection even though the preoperative angiography is negative. At our institution, the procedure is performed under general anaesthesia, with femoral access by 4-French sheath. Selective injections are crucial for an accurate assessment of the feeding artery, angioarchitecture of the AVM, and venous drainage. Three-dimensional rotational spinal angiography can be useful in mapping the morphology of the AVM. In particular, 3-dimension spinal DSA is helpful in identifying intranidal pseudoaneurysms and in evaluating the feasibility of embolization [5,39]. CTA maybe the second choice to identify the EEAVMs. CTA has lower spatial resolution than DSA, and has difficulty in diagnosing complicated vascular malformations [40].

Magnetic resonance imaging (MRI), typically, displays a mass of dilated epidural vessels visualized as flow voids on T2-weighted sequences, serpiginous flow voids extending through several levels. Hyperintense signal within the spinal cord may be seen on T2-weighted images at the vertebral levels where the spinal cord coronal plexus receiving blood from the AVM. In advance stage of the lesion, spinal cord atrophy may be present, MRI of the involved spinal cord would be narrow and high signal intensity on T2-weighted images [23]. Most of the EEAVMs are too small to be detected by MRA, even though high-resolution contrast-enhanced MRA of the spinal cord/column hold further promise to increase both the sensitivity and precision in the diagnosis of spinal vascular lesions.
malformations. AVMs that hemorrhage may demonstrate varying signal intensities consistent with acute or subacute blood products. MRI is considered to be the gold standard in diagnosing spinal epidural hematomas, particularly accurate in demonstrating the extent of the hematoma and its effect on the spinal cord [24,25,29]. The presence of a vascular malformation might be anticipated in a child presenting with a spontaneous epidural hematoma even without overt evidence of flow voids on MRI scan [24].

**Clinical presentation and diagnosis**

Generally, the clinical presentations of EESAVM are slightly more intradural/intramedullary AVMs. Unruptured EESAVM may present as protracted, progressive neurological decline [18,23,33,34]. Ruptured EESAVM may present acute back or thoracic pain subsequently with paraplegia [17,24-29,31,32,36]. There was no large clinical study of EESAVM because of the rarity of this entity. We reviewed the reported cases of EESAVM in the published medical literature. Note: Kendall reported case series of EESAVMs were excluded since they were not quite clear whether they were AVM or arteriovenous fistula. In the English published medical literature, EESAVM is reported in only a few cases (Table 1). There is no gender predilection (male 52.9%), age distribution by the time of diagnosis shows most cases are younger than 20-year (64.7%), most cases present as spontaneous epidural hematoma (64.7%), and the majority of those lesions are located on cervicothoracic junction or upper thoracic segment (75%), which helps explain why upper extremities involvement is less common than lower extremities.

Common to Unruptured EESAVM are symptoms of myelopathy and pain, such as lower or upper extremity weakness, abnormal sensory, disturbance of gait, back pain, and bladder and/or bowel incontinence. MRI may show hyperintense signal within the spinal cord on the T2-weighted images in the involved vertebral levels. Noncontrast MRI is insensitive for the early detection of AVMs. Furthermore, some abnormal scans were read as normal in real-world practice (41). A misdiagnosis of transverse myelitis or Guillain–Barre syndrome may be made at this time, but the patients respond poorly to the treatment of intravenous or oral medicine. A lumbar puncture may be performed, generally, results of cerebrospinal fluid analysis (white blood cell count, red blood cell count, chloride, glucose, bacteria, antibodies, oligoclonal bands) are normal. These symptoms may be intermittent and progressively worse over time. The correct diagnosis may be established until the EESAVM rupture or patients already have certain degree motor and sensory deficits [23,24,33]. The myelopathy syndrome following spinal cord congestion caused by AVMs is rare and nonspecific, when spinal AVM is highly suspected, although the MRI of spine is read as normal; angiography should be performed as soon as possible, because EESAVM may cause spinal epidural hematoma.

Acute symptoms caused by spinal epidural hematoma are typical presentation of ruptured EESAVM, patients may be without any previous syndromes or secondary to unrecognized previous neurologic symptoms caused by EESAVM [23-29,32,33]. Typically, initial symptoms of spinal epidural hematoma are rapid development of excruciating back pain, with or without neurological deficit, followed by rapidly progressive severity of myelopathy, such as abnormal sensory, disturbance of gait, bladder and/or bowel incontinence, lower or upper extremity weakness/paralysis due to the location of hematoma [41]. But in children, the initial presentation may be even more nonspecific and misleading, with the only symptoms being irritability and excessive cry [42]. The diagnosis may be delayed by several hours to days until objective signs of neurologic dysfunction appear [42,43]. Delay in diagnosis may reflect a lower index of suspicion, nonlocalizing clinical findings at presentation, and a small or slower progression of the hematoma [42]. The differential diagnosis of such presentation includes spinal abscess, intrinsic or extrinsic cord tumour, trauma, spinal cord ischemia, disk disease, Guillan-Barré syndrome, transverse myelitis, acute coronary syndrome, and congenital abnormality of the spinal cord [44,45]. CT scanning has a high sensitivity for detecting acute blood products within the spinal canal but has limited capability to accurately assess the degree of spinal cord compression [46]. Spinal MRI has replaced CT myelography as the diagnostic tool of choice for the evaluation of a suspected epidural hematoma, not only by displaying high sensitivity in detecting blood products and defining the age of haemorrhage but also being particularly accurate in demonstrating the extent of the hematoma and its effect on
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Table 1: Exclusively epidural spinal arteriovenous malformation.

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs), Sex</th>
<th>Presentation</th>
<th>Angiographic/Pathologic Findings</th>
<th>Treatment</th>
<th>FU (mos)</th>
<th>Radiologic outcome</th>
<th>Clinical outcome</th>
<th>SSEH</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Newquist, et al. 1960 [17]</td>
<td>18, F</td>
<td>Paralysis of the right leg, below T3 abnormal sensory</td>
<td>C7-T4 Arterial angioma</td>
<td>SO</td>
<td>12</td>
<td>UK</td>
<td>Good improvement</td>
<td>No</td>
<td>During pregnancy</td>
</tr>
<tr>
<td>Harry, et al. 1967 [18]</td>
<td>17, M</td>
<td>Weakness of the right arm and pain in the neck and right shoulder</td>
<td>C3-6 AVM</td>
<td>SO</td>
<td>UK</td>
<td>Residual AVM</td>
<td>Died of induction of anesthesia</td>
<td>No</td>
<td>NF</td>
</tr>
<tr>
<td>Bradac, et al. 1977 [30]</td>
<td>41, F</td>
<td>Disturbance of gait and weakness of fingers</td>
<td>C6-T1 AVM</td>
<td>SO</td>
<td>UK</td>
<td>Completely removed</td>
<td>Neurologically free</td>
<td>No</td>
<td></td>
</tr>
<tr>
<td>Packer, et al. 1978 [36]</td>
<td>17, M</td>
<td>Back pain, Paraplegia</td>
<td>AVM</td>
<td>SO</td>
<td>6</td>
<td>UK</td>
<td>Improved but sever spasticity in legs</td>
<td>Yes</td>
<td></td>
</tr>
<tr>
<td>Muller, et al. 1982 [31]</td>
<td>59, F</td>
<td>Back pain, Paraplegia</td>
<td>T9-10, vascular malformation</td>
<td>SO</td>
<td>6</td>
<td>UK</td>
<td>Improved but weakness of legs</td>
<td>Yes</td>
<td></td>
</tr>
<tr>
<td>Matsumura, et al. 1986 [34]</td>
<td>50, F</td>
<td>Weakness of the legs, sensations disturbed below right L2 and left L5 level</td>
<td>S1 AVM</td>
<td>SO</td>
<td>4</td>
<td>UK</td>
<td>Improved but ambulating with cane</td>
<td>No</td>
<td></td>
</tr>
<tr>
<td>Emery, et al. 1988 [32]</td>
<td>61, M</td>
<td>Chest pain and paraplegia</td>
<td>C7 AVM</td>
<td>SO</td>
<td>UK</td>
<td>UK</td>
<td>Neurologically free</td>
<td>Yes</td>
<td></td>
</tr>
<tr>
<td>D’Angelo, et al. 1990 [29]</td>
<td>51, M</td>
<td>Paraparesis, absence of sensory below T5</td>
<td>T2-4 AVM</td>
<td>SO</td>
<td>UK</td>
<td>UK</td>
<td>Neurologically free</td>
<td>Yes</td>
<td></td>
</tr>
<tr>
<td>Olivero, et al. 1993 [28]</td>
<td>16, M</td>
<td>Acute midline thoracic pain, paraplegia</td>
<td>T5 AVM</td>
<td>SO</td>
<td>6</td>
<td>Completely removed</td>
<td>Neurologically free</td>
<td>Yes</td>
<td></td>
</tr>
<tr>
<td>Muhonen, et al. 1995 [27]</td>
<td>2, M</td>
<td>Paraparesis in the legs</td>
<td>C7-T2 vascular malformation</td>
<td>SO</td>
<td>2</td>
<td>UK</td>
<td>Neurologically free</td>
<td>Yes</td>
<td>Infant</td>
</tr>
<tr>
<td>Miyagi, et al. 1998 [26]</td>
<td>16, F</td>
<td>Neck pain, complete quadriaparesis, hypesthesia below both shoulders</td>
<td>C2 AVM</td>
<td>SO</td>
<td>1</td>
<td>UK</td>
<td>Neurologically free</td>
<td>Yes</td>
<td></td>
</tr>
<tr>
<td>Nadig, et al. 2000 [31]</td>
<td>10, F</td>
<td>Abnormal posture</td>
<td>L3-5 AVM</td>
<td>SO</td>
<td>12</td>
<td>UK</td>
<td>Neurologically free</td>
<td>No</td>
<td>NF</td>
</tr>
<tr>
<td>Rohany, et al. 2007 [33]</td>
<td>29, F</td>
<td>Right upper extremity weakness and numbness</td>
<td>C6-T1 AVM</td>
<td>Embolization and SO</td>
<td>UK</td>
<td>UK</td>
<td>No</td>
<td>KTWS</td>
<td></td>
</tr>
<tr>
<td>Paraskevopoulos, et al. 2013 [25]</td>
<td>8, M</td>
<td>Weakness of lower limbs</td>
<td>C7-T2 AVM</td>
<td>SO</td>
<td>7</td>
<td>Completely removed</td>
<td>Ambulating with support</td>
<td>Yes</td>
<td></td>
</tr>
<tr>
<td>13, M</td>
<td>13, M</td>
<td>Interscapular pain, paraplegia</td>
<td>T1-T5 AVM</td>
<td>SO</td>
<td>3</td>
<td>Completely removed</td>
<td>Improved but intermittent urinary continence</td>
<td>Yes</td>
<td></td>
</tr>
<tr>
<td>Wang, et al. 2016 [23]</td>
<td></td>
<td>Paraplegia</td>
<td>C7-T3 AVM</td>
<td>SO</td>
<td>6</td>
<td>Completely removed</td>
<td>Neurologically free</td>
<td>Yes</td>
<td></td>
</tr>
</tbody>
</table>

FU = follow-up; UK = unknown; SO= Surgical operation; SSEH=spontaneous spinal epidural hematoma; NF=neurofibromatosis; KTWS =Klippel-Trenaunay-Weber syndrome
the spinal cord [25,47]. Therefore, in our opinion, an MRI should be routinely obtained, even in cases that have spontaneous resolution of symptoms. Post-operation histological and spinal angiography should be routinely performed. Although some spinal epidural hematoma resolved spontaneously with conservative treatment [48-50], an angiogram should be routinely obtained for the possibility of repetitive hemorrhage from AVM.

Treatment and outcomes

Due to the extreme rarity of this lesion, there is no reliable and valid data to determine a standardized approach to the treatment of EESAVMs. EEAVMs tend to produce slowly progressive clinical syndromes of myelopathy, and most cases present acute onset of hemorrhage. Intervention, either by microsurgical operation or endovascular treatment, aims to interrupt and reverse this progression by minimizing blood flow through the abnormal connection of AVM, restore normal spinal cord perfusion, as well as remove the pressing against spinal cord which is caused by nidus and thick tortuous arterialized drainage veins. The goals of intervention also include prevention of future acute hemorrhage, removal of hemorrhage product-hematoma. Because the niduses of EESAVM are located in the region of epidural space without spinal cord involvement, the effect of surgical/intravascular treatment is better than spinal cord AVM. Most reported cases (62.5%) treated by surgical operation or combined with embolization have no residual neurological symptoms (Table 1). Harry reported a seventeen years old male with EESAVM in 1967, he was died of introduction of anaesthesia rather than the EESAVM itself [18], except this case all cases postoperative neurological syndromes had improvement. After 1990, all the reported cases treated by surgical operation can walk post-operation with or without support; no patient neurological symptoms get worse than pre-operation. The outcome is better than spinal cord AVM [21].

From the aspect of microsurgical operation, the location of the AVM determines the surgical approach and operative difficulty. Luckily, most EESAVMs are located in the posterior or posterolateral aspect of the spinal epidural space (94.1%) [17,18,22-33,36,51], only one case reported in the literature is located in the anterior aspect of the epidural space (5.9%) [34]. So, most EESAVMs are accessible through a posterior laminectomy and partial facetectomy. Because the niduses are out the thecal sac, the operation are relatively safe than spinal cord AVMs. In the literature after 1990, all reported outcomes of EESAVM were totally removed [23-25]. As the nature of vascular lesion, during the procedure venous and arterial bleeding may be profuse, pre-operation careful assessment of the EESAVM is valuable. Spinal epidural hematoma caused by EESAVM usually requires emergency surgery as soon as it is discovered. The postoperative recovery is correlated with the rapidity of surgical operation and severity of the preoperative neurological deficit [52].

Endovascular treatment of spinal vascular malformation was first reported by Doppman in 1968 [53]. Subsequently, angiography, in addition to its position as the gold standard of diagnosis, has experienced a continued technical and technological evolution allowing for its increasing utility in the treatment of spinal vascular malformations, either alone or in combination with surgical intervention [54]. However, there was only one EESAVM treated by endovascular embolization followed by surgical resection [33]. The reason for this phenomenon may be that most correct diagnosis establish until the EESAVM rupture. At the emergent setting, surgical operation is the first choice to evacuate the hematoma and decompress spinal cord. Given their small size and characteristics of blood supply, especially the usual locations of nidus are the posterior or posterolateral aspect of the spinal canal and totally out of the thecal sac, the best management is still surgical operation.

Conclusion

EESAVM is an extremely rare lesion, which is located entirely in the epidural space and fed by radicular vessels or segmental arteries. Generally, clinical presentations of EESAVM are slighter than intradural/intramedullary AVMs. EESAVM has a high tendency to cause hemorrhage. Symptoms are mostly caused by the epidural hematoma from ruptured AVM. Retrograde blood flow from the AVM can cause myelopathy. Prompt diagnosis and emergency surgical treatment are crucial. Long-term functional prognosis of EESAVM is good, but delayed surgical operation can leave residual symptoms.
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