CASE REPORT

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Spinal epidural cavernous angioma: two case reports and review of the literature

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Cavernous angiomas are vascular malformations that occur most frequently in the supratentorial area of the central nervous system (CNS). Spinal epidural occurrence is rare. This article describes 2 cases of spinal epidural cavernous angioma. The lesions were hypo- to isointense on T1-weighted magnetic resonance images (MRIs) and hyperintense on T2-weighted images. Both were enhanced homogenously with intravenous contrast. Total resection was achieved in both cases, and the lesions were histopathologically diagnosed as cavernous angiomas. The patients' symptoms regressed postsurgery. Although the MRI features of cavernous angiomas are well known, spinal epidural occurrence is rare and many differential diagnoses have similar clinical and imaging findings. It is important to definitively diagnose these lesions prior to surgery in order to prevent possible intraoperative complications such as massive bleeding and to maximize chances for complete resection. In addition to case descriptions, this article includes a thorough literature review to raise clinical awareness about this well-known but rare spinal entity.

Keywords: Cavernous angioma; epidural; intraspinal extradural tumor.

Cavernous angiomas, also known as cavernomas, are vascular malformations that occur primarily in the central nervous system (CNS). Thirteen percent of cavernous angiomas occur at extra-axial body sites, including the orbit, cavernous sinus, middle cranial fossa, vertebrae, spinal epidural space, and other extradural intracranial sites.[1] The histological features[2,3] of these lesions are identical regardless of their location. Spinal epidural cavernous angiomas (SECAs) are well-known pathological entities that typically originate from vertebral bone. Other sites of origin are rare. Spinal epidural cavernous angiomas account for 12% of all intraspinal angiomas and 4% of all spinal epidural[1–7] masses.

We present 2 cases of SECA in which the malformation did not originate from vertebral bone.

Case reports

Case 1 – A 50-year-old woman was admitted to our outpatient clinic in July 2009, complaining of neck and left arm pain. She had been experiencing these symptoms for 1 year. Her family and medical histories were unremarkable. Neurological examination revealed no motor deficits, but there were sensory deficits in the T2 and

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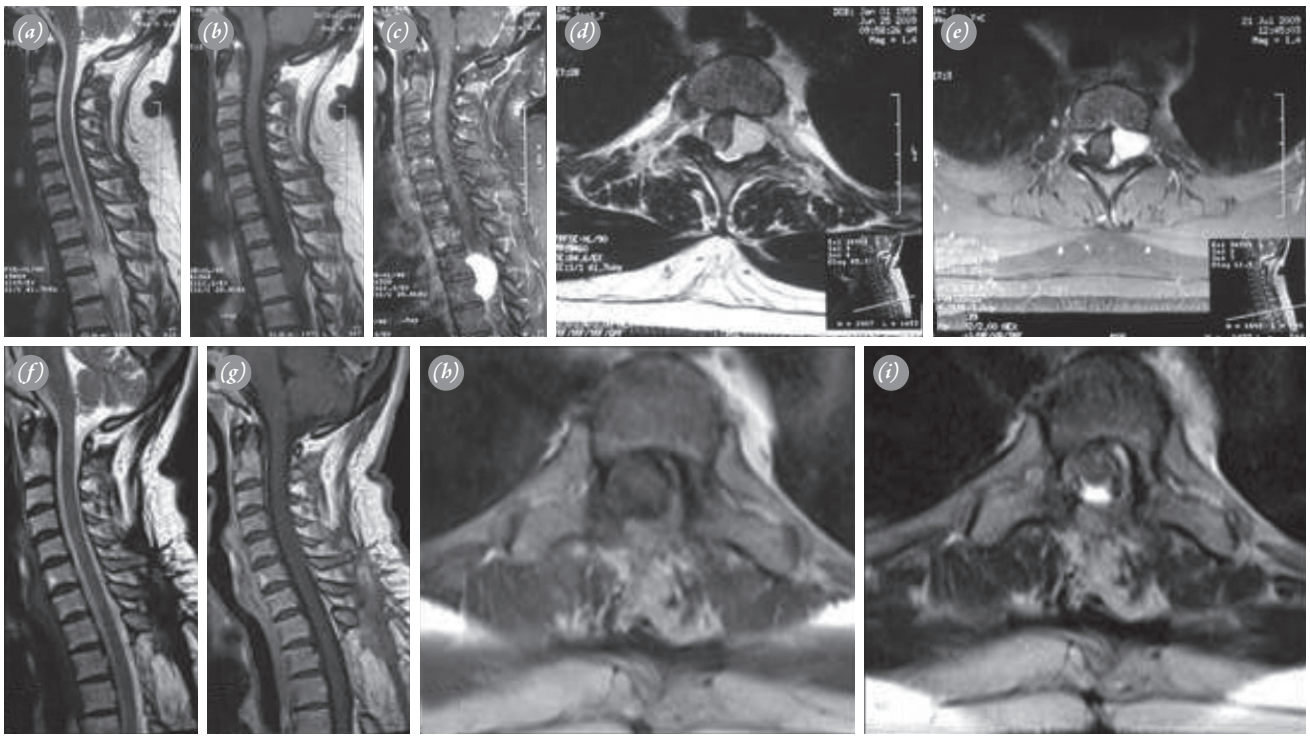

Fig. 1. Case 1: MRIs of the patient's cervicothoracic region showed an ovoid epidural lesion between T2 and T3 levels **(a–c)**. Axial images revealed the lesion in a left lateral location **(d,e)**. The lesion enhanced homogenously with intravenous gadolinium injection **(c,e)**. During surgery, total laminectomies were performed at T2 and T3, and the lesion was totally excised **(f–i)**.

T3 dermatomes. The patient also exhibited hyperactive deep tendon reflexes in her left lower extremity. Urinary and anal sphincter control was normal. Magnetic resonance imaging (MRI) demonstrated an oval mass in the epidural space between T2 and T3. The lesion was isointense on T1- and hyperintense on T2-weighted images, and it exhibited homogeneity with intravenous contrast injection. The mass was entirely epidural; there was no involvement of vertebral bodies or vertebral foramina (Figures 1a–e). The patient's spinal cord was compressed to the right side of the spinal canal.

Intraoperatively, total laminectomies at T2 and T3 revealed a highly vascularized purple mass. Total resection was performed using microsurgical techniques. Microscopic examination identified the lesion as a cavernous angioma encased in adipose tissue (Figure 2a).

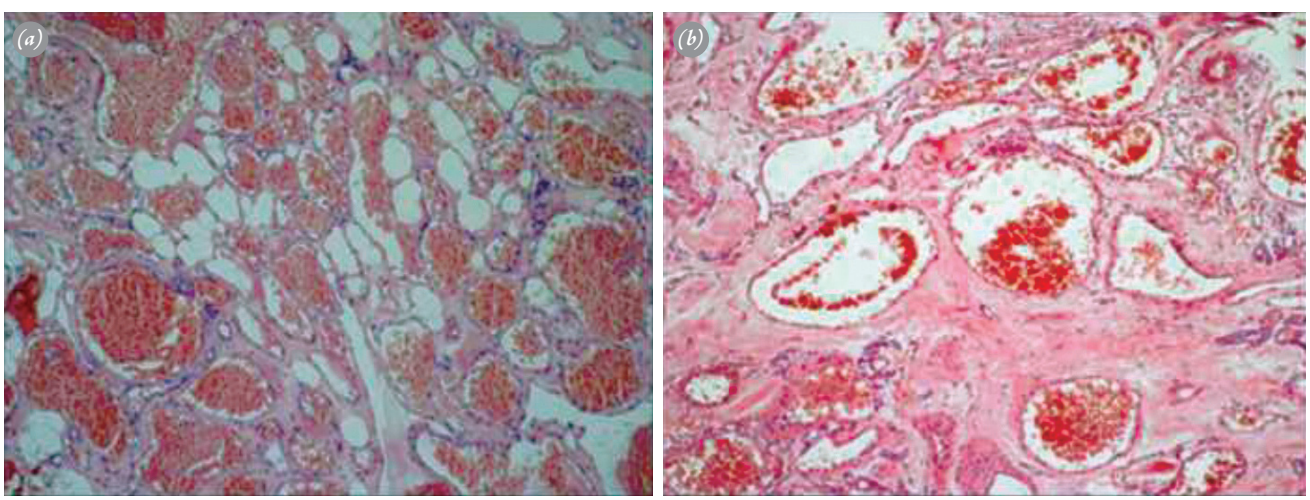

Fig. 2. **(a, b)** Sections of the cavernous angiomas from both cases show the typical histological features of closely apposed, hyalinized vessels of various sizes (h,e; left side: Case 1 x100; right side: Case 2 x200). [Color figures can be viewed in the online issue, which is available at www.aott.org.tr]

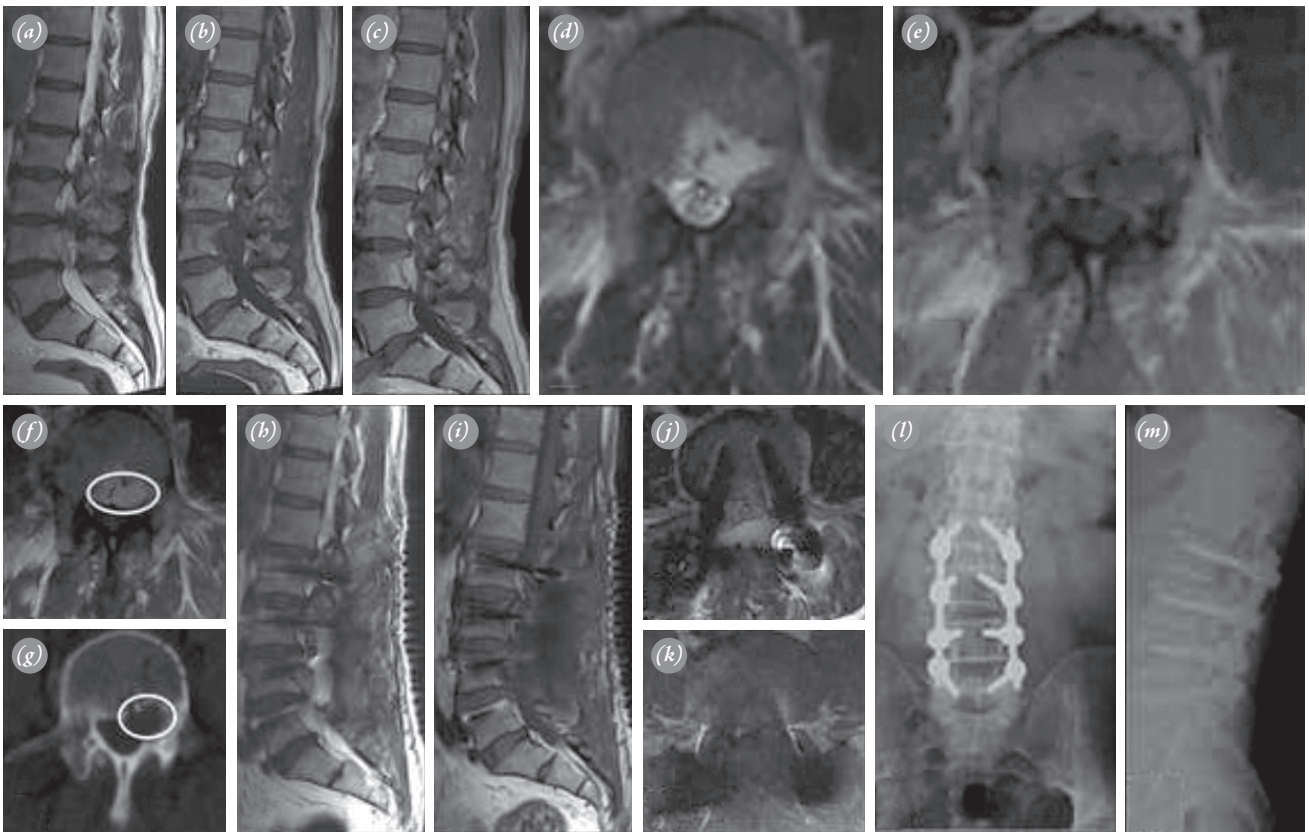

Fig. 3. Case 2: MRIs of the patient's lumbar region revealed spinal stenosis. An ovoid mass posterior to the L4 vertebral body enhanced homogenously with intravenous gadolinium **(a–f)**. CT revealed the lesion infiltrating the posterior portion of the vertebral body **(g)**. During surgery, total laminectomies were performed at L3 and L4, the lesion was resected, and instrumentation was performed between levels L2–L5 **(h–k)**. Anterior-posterior and lateral lumbar films at 10 months postsurgery showed screws in place **(l,m)**.

Postoperatively, the patient reported complete pain relief and had no additional neurological deficits. Follow-up MRI of her thoracic spine 2 years postsurgery showed no residual or new lesions (Figures 1f–i).

Case 2 – A 68-year-old man presented to our outpatient clinic in January 2011 with gait impairment and progressive back pain and bilateral leg pain. He had experienced the symptoms for 6 years, and the back pain and leg pain were greater on the left side. Neurological examination revealed normal muscle strength and deep tendon reflexes in the lower extremities, with no sensory deficits. Sphincter control was normal. After walking approximately 20 meters, the patient exhibited intermittent neurogenic claudication in both lower limbs. Lumbar spinal stenosis was suspected. MRI confirmed severe spinal stenosis at L3–L4 and L4–L5 (Figures 3a–c). The images also showed an oval mass in the epidural space at L4. The lesion was hypointense on T1- and hyperintense on T2-weighted images and enhanced homogenously with intravenous gadolinium injection (Figures 3a–f). Axial T2-weighted images revealed that the mass was impinging on the anterior and left lateral aspects of the spinal cord (Figure 3f). Computed tomography (CT) showed erosion of the posterior portion of the L4 vertebral body (Figure 3g).

Surgery exposed a highly vascularized purple lesion in the anterior epidural space of the lumbar spine. The mass had invaded the L4 vertebral body but did not originate from it. The lesion was totally resected. To treat the coexisting spinal stenosis, total laminectomies were performed at L3 and L4, combined with instrumentation from L2–L5 (Figures 3h–k). The histopathologic diagnosis was cavernous angioma (Figure 2b). Postoperatively, the patient's back pain improved; however, he developed neuropathic pain in his lower limbs. A low dose of a pregabalin analogue was prescribed. At 10 months postsurgery, the neurogenic claudication and neuropathic pain had regressed, and plain radiographs of the lumbar spine showed screws in place (Figures 3l, m).

Discussion

Cavernous angiomas can develop anywhere in the body,[2] but occur most frequently in the supratento-

Table 1. Differential diagnoses for spinal epidural lesions.

Lesion in spinal epidural space	Radiological and/or clinical findings, and comparisons to cavernous angioma
Cavernous angioma	Hypo-to isointense on T1-, hyperintense on T2-weighted MR images. Homogenous enhancement with IV contrast. Oval shape but may flatten over spinal cord. Lobular contour. May feature erosion of adjacent bone, foraminal extension, and dural tail.
Metastasis	Erosion of adjacent bone and wrapping of spinal cord. Usually in posterior epidural space. May feature multiple lesions and bone involvement.
Meningioma	Similar MR findings as CA but isointense on T2-weighted images. Osteoblastic or osteolytic changes in adjacent bone and calcification of lesion on CT scan. Dural tail on MR imaging, as seen with some CAs.
Neurinoma	Similar MR findings as CA but less intense contrast enhancement. Intervertebral foraminal extension is common, as seen with some CAs.
Angiolipoma	Hyperintense with or without hypointense foci (vascular regions) on T1-weighted MRIs. Heterogenous enhancement with IV gadolinium. Generally non-infiltrating, fusiform shape. Lobular contour. Typically features calcifications and hemorrhagic areas.
Lymphoma	Dura-based types have similar MR findings to CA but can be identified as intradural extra-axial lesions via fluid-attenuated inversion recovery MR scan. Hypointense on T1-, hyperintense on T2-weighted MR images. May feature multiple lesions and bone involvement.
Eosinophilic granuloma	Osteolytic accompanying lesion in adjacent bone, long bone, pelvis, rib, skull.
Disc herniation	Low intensity on T1- and T2-weighted MRIs. Hyperintense posterior annular tear on T2-weighted MRIs. Usually anterior and lateral lumbar locations. Peripherally enhancing cyst-like mass. Thick and irregular rim. Accompanying degenerative intervertebral disc.
Pure hematoma	History of previous trauma, coagulopathy, intervention. In extremely acute stage, hyperintense on T1-, very low intensity on T2-weighted MRIs. Spontaneous resolution on follow-up imaging.
Abscess or phlegmon	Typically observed bone marrow and intervertebral disc abnormalities, but may occur without disc pathology.
Multiple myeloma	Isointense to spinal cord on T1 and heterogenously hyperintense on T2-weighted MR images. MR changes in vertebrae at multiple spinal levels.

CA: Cavernous angioma; CT: Computed tomography; MR: Magnetic resonance; MRI: Magnetic resonance imaging; IV: Intravenous.

rial area[2–4,6,7] of the CNS. In 1959, Russell and Rubinstein[8] classified cavernous angiomas as vascular malformations, along with arteriovenous malformations, venous angiomas, and capillary telangiectasia. Cavernous angiomas of the spine are very uncommon, but in such cases the most frequent site of origin is the vertebral body.[2,3,7] Spinal epidural cavernous angiomas, a rare type of lesion,[2] were first described by Globus and Doshay[9] in 1929. They can originate from vertebral bone or can be purely epidural (i.e., not originating from vertebrae).[3] Spinal epidural cavernous angiomas may be sporadic or familial.[6] The most common site of occurrence is the thoracic posterior epidural space;[4,5] some reports describe SECAs involving vertebral foramina or even paravertebral structures.[6] Since the first description of SECA, 128 cases, including ours, have been reported.[4,10–23] These patients ranged from 23 months to 81 years of age, with a mean age of 47.7 years (51 years and 42.6 years for males and females, respectively). The male to female ratio for the 128 cases is 1.5:1, which suggests a male bias for SECA.

Most SECAs increase in volume gradually; thus, patients tend to present with progressive sensorimotor deficits.[1,4,6] The main signs and symptoms are related to radiculopathy or myelopathy.[5,7] Lesions in anterior and lateral lumbar epidural locations may cause radicular symptoms that mimic disc herniation.[6] In rare cases, patients may exhibit rapid deterioration of motor strength due to sudden enlargement of a SECA. Such enlargement can result from thrombotic or mechanical venous occlusion, increased vascularization caused by hormonal effects, or acute intralesional hemorrhage.[1,2,6,7,24,25] However, acute presentation is more common in patients with intra-axial cavernous angiomas than in cases of SECA.[1]

The advent of MRI has increased the frequency of SECA diagnosis from 0.8 to 1.9 cases per year.[1] MRI is superior to CT for diagnosing SECA; however, CT

is useful for delineating connections with surrounding bone or extension into vertebral foramina.[3,4,6] In contrast to intraparenchymal-type cavernomas, SECAs are more homogenous in appearance on CT and MRI.[3] These lesions appear as solid ovular hypervascular lesions with lobular contours, and they are enhanced markedly with contrast injection.[1,3,5–7] They often span multiple vertebral segments, in contrast to arteriovenous malformations or venous angiomas, which typically occur at 1 vertebral segment.[5] Spinal epidural cavernous angiomas tend to be hypo- to isointense compared to spinal cord on T1-weighted MRIs due to slow blood flow and hyperintense on T2-weighted scans due to high content of inert blood.[1–4,6,7] However, hemorrhagic SECAs appear hyperintense on T1- and T2-weighted images in the acute stage.[2,6] Spinal epidural cavernomas typically do not exhibit a hemosiderin rim on MRI.[1–3,6,7] It is not clear whether angiography can help delineate the vascularity of these lesions.[1,6]

The differential diagnosis for SECA includes a broad spectrum of disease processes.[1–7, 26–28] Table 1 summarizes the radiological and clinical properties of lesions that occur in the spinal epidural space.

Histopathologically, cavernous angiomas are composed of packed dilated blood-filled vessels with intervening loose connective and adipose tissue.[2,3,5,7,8] They are devoid of elastic and muscular tissue and contain no neurons.[2,3,6–8] It has been theorized that these malformations develop when embryonic primordial vessels do not differentiate fully,[2,29] thus producing only partially-differentiated adult-type vascular structures.

Total resection of a SECA typically leads to a satisfactory recovery.[1,3,4,6] One consideration regarding surgery is the potential for massive intraoperative bleeding, though this is rare. Furthermore, the surgeon should be prepared for potential extension of the lesion outside the spinal region or for possible compression of spinal nerve roots. Currently, the long-term outcome for cases of subtotal resection of SECA is unclear, but radiotherapy is reportedly beneficial for these patients.[1,6] Spinal epidural cavernomas do not regress with time, and SECAs that are managed conservatively may hemorrhage. As the spinal cord is very sensitive to sudden compression, hemorrhage generally results in poor clinical outcomes.[4]

A preoperative diagnosis of SECA is significant, as these vascular malformations carry risk of massive bleeding and may not be amenable to total resection without thorough preoperative assessment and surgical planning.

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