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Case Report

Dorsal spinal epidural cavernous hemangioma

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Abstract

A 61-year-old female patient presented with diffuse pain in the dorsal region of the back of 3 months duration. The magnetic resonance imaging showed an extramedullary, extradural space occupative lesion on the right side of the spinal canal from D5 to D7 vertebral levels. The mass was well marginated and there was no bone involvement. Compression of the adjacent thecal sac was observed, with displacement to the left side. Radiological differential diagnosis included nerve sheath tumor and meningioma. The patient underwent D6 hemilaminectomy under general anesthesia. Intraoperatively, the tumor was purely extradural in location with mild extension into the right foramina. No attachment to the nerves or dura was found. Total excision of the extradural compressing mass was possible as there were preserved planes all around. Histopathology revealed cavernous hemangioma. As illustrated in our case, purely epidural hemangiomas, although uncommon, ought to be considered in the differential diagnosis of spinal epidural soft tissue masses. Findings that may help to differentiate this lesion from the ubiquitous disk prolapse, more common meningiomas and nerve sheath tumors are its ovoid shape, uniform T2 hyperintense signal and lack of anatomic connection with the neighboring intervertebral disk or the exiting nerve root. Entirely extradural lesions with no bone involvement are rare and represent about 12% of all intraspinal hemangiomas.

Key words: Epidural, hemangioma, spinal

INTRODUCTION

The aim of this case report is to describe an uncommon etiology of back pain caused by purely epidural spinal cavernous hemangioma, to review the literature and to highlight the diagnostic challenges associated with this lesion. The differential diagnosis of extra-axial soft tissue lesions of the spine has been briefly reviewed, including comparison of the imaging features of this lesion with other more common pathologies at this site.

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CASE REPORT

A 61-year-old female patient presented with diffuse pain in the dorsal region of the back of 3 months duration. There was no history of fall or trauma. There was no history of radicular pain in the chest. There were no associated sphincter disturbances. The tone, power and reflexes were normal in all four limbs. On local examination, there was no focal tenderness over the spine.

A plain and contrast-enhanced magnetic resonance imaging (MRI) study of the dorsal spine was performed. The MRI showed an extramedullary, extradural space occupative lesion on the right side of the spinal canal from D5 to D7 vertebral levels. The mass was well marginated and there was no bone involvement. The lesion was hypointense on T1-weighted images [Figure 1] and hyperintense on T2-weighted images, in comparison to the intervertebral disk. On post-contrast, fat suppressed, T1-weighted axial [Figure 2a] and sagittal [Figure 2b] images, the lesion displayed intense and

homogenous enhancement. Compression of the adjacent thecal sac was observed, with displacement to the left side. Radiological differential diagnosis included nerve sheath tumor and meningioma.

The patient underwent D6 hemilaminectomy under general anesthesia. Intraoperatively, the tumor was purely extradural in location with mild extension into the right foramina. No attachment to the nerves or dura was found. The mass was mostly soft and reddish with more bleeding noted than usual for a meningioma or a schwannoma. Total excision of the extradural compressing mass was possible as there were preserved planes all around. Back pain disappeared after total surgical excision.

Histopathology revealed cavernous hemangioma [Figure 3]. On histology, the lesion was composed of multiple, closely spaced, dilated and ectatic vascular channels containing blood. The spaces were lined by a single layer of benign endothelial cells. Nuclear pleomorphism and mitotic figures were not seen.

The patient followed up for 15 months after surgery and she had complete resolution of symptoms. Follow-up MRI at 15 months showed no residual or recurrent lesion [Figure 4].

DISCUSSION

The differential diagnosis of extra-axial soft tissue lesions of the spine has been briefly reviewed, including comparison of the imaging features of this lesion with other more common pathologies at this site.

As illustrated in our case, purely epidural hemangiomas, although uncommon, ought to be considered in the differential diagnosis of spinal epidural soft tissue masses. Findings that may help to differentiate this lesion from the ubiquitous disk prolapse, more common meningiomas and nerve sheath tumors are its ovoid shape, uniform T2 hyperintense signal and lack of anatomic connection with the neighboring intervertebral disk or the exiting nerve root.[1] In our case, differential diagnosis offered, based on preoperative MRI features, was nerve sheath tumor and meningioma. The presence of intense, homogenous post-contrast enhancement and lack of anatomic connection with the disk ruled out a prolapsed intervertebral disk. Retrospectively, we realized, the shape of the lesion was unusual for a nerve sheath tumor. The mass did not have the linear or dumbbell shaped configuration characteristic of a nerve sheath tumor; it appeared instead to enclose or wrap around the adjacent thecal sac. The high T2 signal was unusual for a meningioma.

Other than meningiomas and nerve sheath tumors, less common differential diagnoses for this lesion would have included metastasis, round cell tumor, sarcoidosis, histiocytosis and tuberculosis. The absence of any bony changes at all in our case made metastasis, round cell tumor and eosinophilic granuloma unlikely. Epidural lymphoma shows characteristic low T2 signal. Although spinal epidural granulomatous pathologies such as tuberculosis and sarcoidosis with no origin in the vertebral body are known entities, they are uncommon.

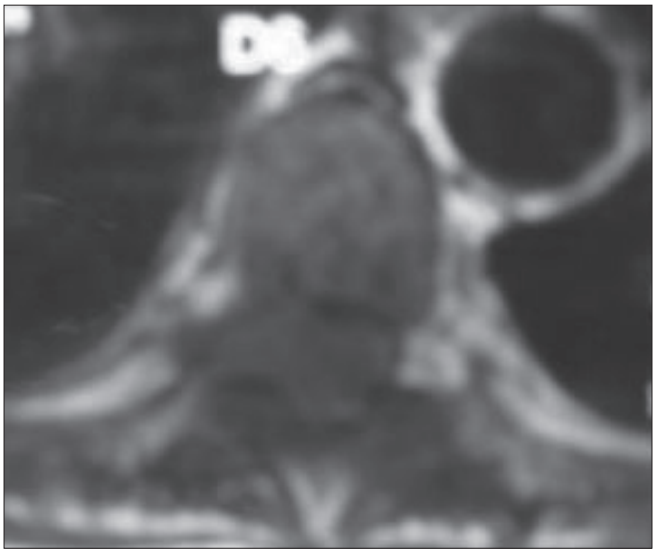

Figure 1: Axial T1-weighted image shows an epidural soft tissue mass in the right side of the dorsal spinal canal at D6 vertebral level, causing thecal sac compression

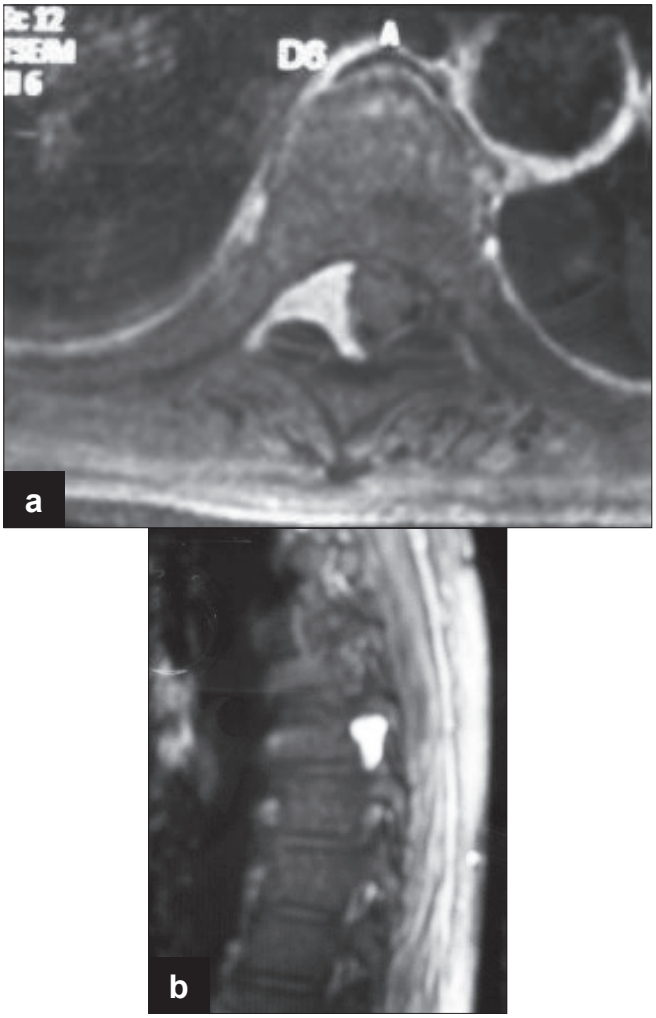

Figure 2: (a) Axial and (b) sagittal post-contrast, fat saturated, T1-weighted images show intense enhancement of the epidural lesion

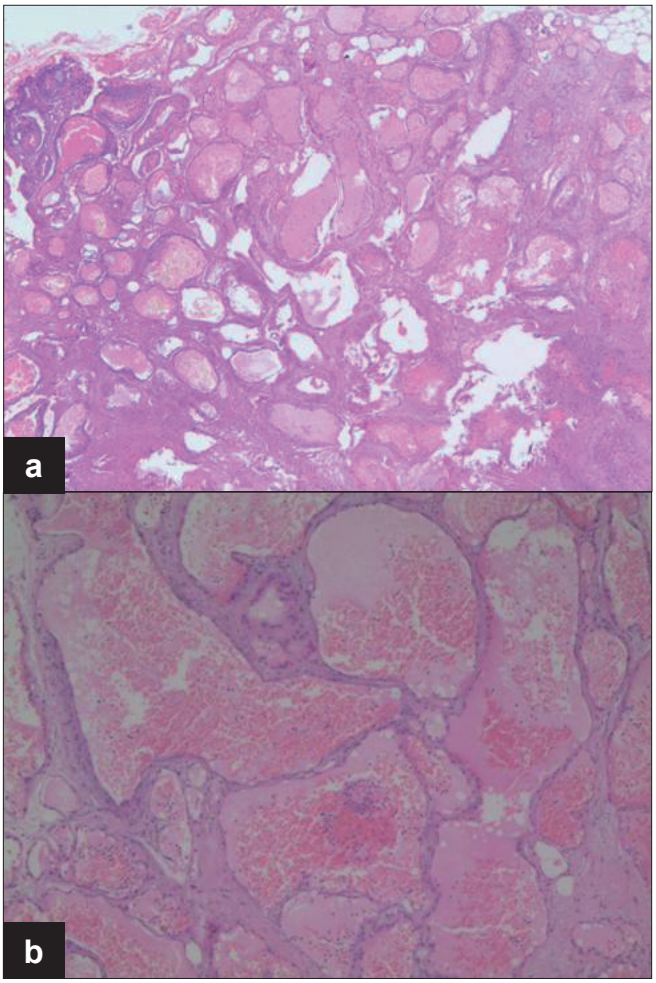

Figure 3: Histopathology showed cavernous hemangioma. (a) ×4 and (b) ×10 showing large dilated vascular spaces containing blood, lined by a single layer of endothelium

Leptomeningeal sarcoidosis and tuberculosis are likely to be more linear in morphology with craniocaudal extension along more than one vertebral body. They are also associated with other systemic signs and symptoms as well as with mediastinal lymphadenopathy in most instances.

The MRI signal intensity character of a cavernous hemangioma reflects its histopathology. The intermediate T1 signal correlates with the composition of the lesion, its marked hyperintense T2 signal corresponds to high water content and finally intense enhancement reflects the flow into the numerous vascular channels of the lesion (sinusoidal vascular structure).[2] In a few cases reported in literature, the signal is heterogenous on all pulse sequences with ring enhancement on post-contrast images,[3] and this is likely due to intralesional hemorrhage and related degeneration. In one instance,[4] hyperintense signal has been reported on both T1- and T2-weighted images, related to the presence of hemorrhage in the subacute phase. Sclerohyaline degeneration has been purported to be the cause of complete lack of enhancement in another reported case.[5] In a few cases, the T2 and gradient echo sequences show profoundly

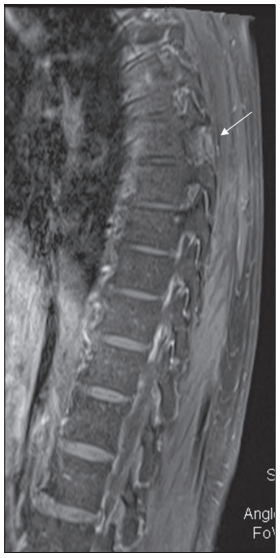

Figure 4: Follow-up postoperative sagittal post-contrast T1 image shows complete excision of the lesion

hypointense signal areas due to acute hemorrhage containing deoxyhemoglobin or as a result of hemosiderin deposition related to recurrent hemorrhages.[6]

These tumors are most frequently found in the thoracic and lumbar spine. A cervical location for an extradural cavernous hemangioma is less common. 80% of the reported cases were in the thoracic spine, with posterior location within the spinal canal in 93% of cases.[7] Extension into the lateral recesses may or may not be present. Spinal epidural hemangiomas have similar signal intensity and enhancement characteristics as hemangiomas of the cavernous sinus, with high T2 signal and intense homogenous enhancement. On the other hand, the MR appearance of parenchymal cavernous angiomas of the brain and spinal cord is vastly different with heterogenous T1 and T2 signals and significant susceptibility correlating to repeated episodes of bleeding and hemosiderin deposition. The lack of a low-signal hemosiderin ring on both T1- and T2-weighted images in epidural lesions is likely to be related to more rapid removal of blood degradation products outside the blood–brain barrier.[8-10]

Clinical presentation of these lesions may be with local symptoms such as back pain or with radiculopathy and/or myelopathy. Commonly, these lesions present as lumbar radiculopathy that clinically and radiologically mimics the presence of disk herniation.[11] In our case, the patient did not complain of radiculopathy or myelopathy. The clinical course is usually slowly progressive. Acute clinical deterioration may be observed in a few instances – related to significant growth of lesion, intralesional hemorrhage or thrombotic venous occlusion.[12] The propensity to bleed is explained by the thin-walled vessels and by stasis of blood flow in the lesion.[13] Other causes of acute symptoms include estrogen mediated neoangiogenesis in the lesion or drainer compression by a gravid uterus. Of the approximately 84 cases of this entity reported in

literature, four cases were seen in the pediatric population;[14-16] rest of the patients were adults.

The clinical presentation of back pain, radiculopathy and myelopathy usually leads to an MRI examination in these patients. A digital subtraction angiography (DSA) has no role in diagnosis as these lesions have no communication with the spinal arterial circulation and are angiographically occult.

Purely extradural benign vascular lesions of the spine are rare and comprise less than 6% of all spinal neoplasms.[1] They include cavernous hemangioma, arteriovenous hemangioma and also angiolipoma. Cavernous hemangiomas are commonly encountered as solitary or multiple intracranial lesions. Spinal cavernous hemangiomas most often originate from the vertebral bodies, sometimes with secondary extension into the extradural space.[17] Entirely extradural lesions with no bone involvement represent about 12% of all intraspinal hemangiomas.[18-20] To the knowledge of the authors, about 82 cases of purely epidural spinal hemangiomas, without primary origin in the vertebral body, have been reported in literature to date.

As in our case, a review of literature shows that the diagnosis is almost always missed on preoperative imaging. Therefore, although the lesion is rare and a high index of suspicion is required for the diagnosis based on pre operative imaging, cavernous hemangioma should be included in the differential diagnosis of purely extradural soft tissue lesions of the spine. It has been suggested[21] that early and accurate diagnosis followed by complete excision of the lesion prior to any possible episodes of massive intralesional bleeding is related to a favorable outcome. Sudden intralesional hemorrhage can lead to spinal cord compression and subsequent severe disability.

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