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Case Report

Pure Spinal Epidural Cavernous Hemangioma: A Rare Cause of Thoracic Myelopathy

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Astract:

Cavernous hemangiomas mostly occur in intracranial structures but they also may occur in the spine very rarely. Even if found, spinal hemangiomas are commonly vertebral origin and solely epidural hemangiomas not originating from the vertebral bone are very rare. Our case is a 65-year-old man presented to us with progressive paraparesis from six months with motor power 3/5 in both lower extremities. The pathology was total epidural spinal hemangioma at D9-10 vertebral level with typical myelopathy like presentation.

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Introduction:

Cavernous hemangiomas are vascular malformation, occur usually in the intracranial structures and rare in the spine[1,2,3,6,10]. About 80% of the cavernous hemangiomas are supratentorial, 15% infratentorial and only 5% are located in the spine[2,3,5,12,13] . Pure spinal epidural cavernous hemangiomas (PSECHs) are very rare[4,7,10]. In most cases, epidural hemangiomas are secondary extensions from the vertebral pathology and the "pure" epidural hemangiomas not originating from the vertebral bone are very rare[1,5,9,13].

Case Report:

A 65-year-old man presented to us with progressive paraparesis from six months ago and made him bed bound for last 2 months. On neurological examination, his motor weakness was rated as grade 3/5 in bilateral lower extremities. There was hypertonia and exaggerated knee and ankle jerk bilaterally. He also complained of decreased sensation below the T11 sensory dermatome. Magnetic resonance imaging showed dorsal spinal tumor at D9 level. The tumor was in epidural space compressing spinal cord ventrally and totally free from thoracic vertebral bone.

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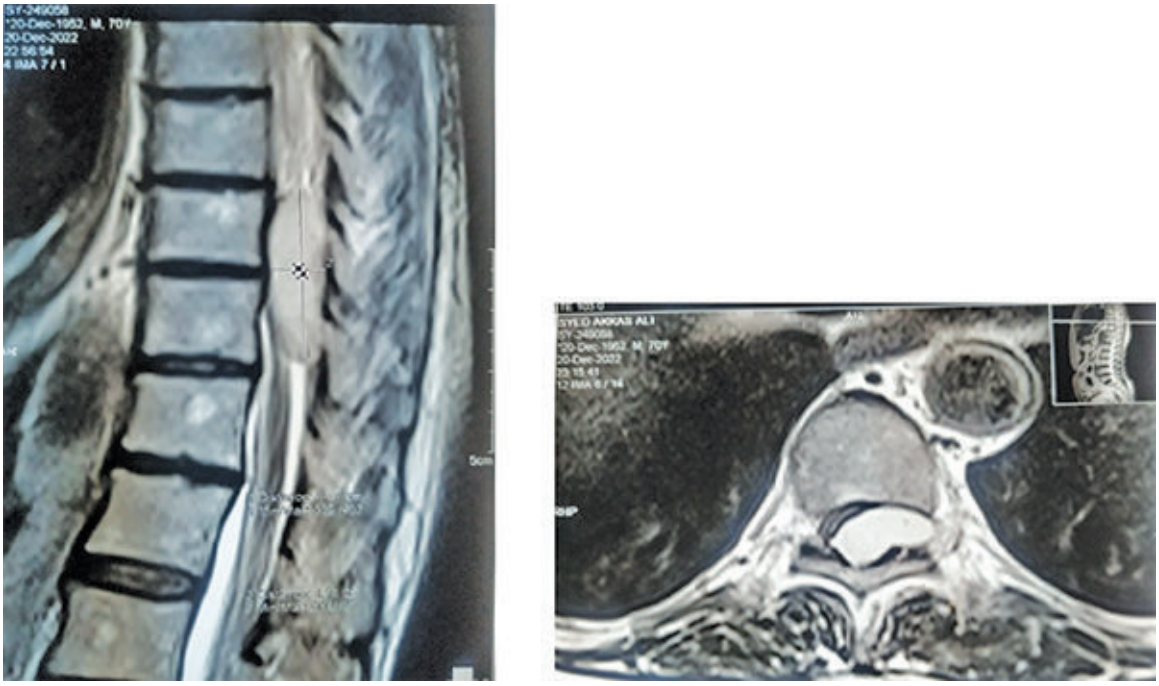

(A) (B)

Figure 1: *Pre-operative MRI of thoracic spine: T2-weighted image sagittal section (A) and T1-weighted image coronal section (B) showing epidural lesion located dorsally occupying most part of canal at D9-10.*

We performed D9 laminectomy and removed the tumor completely. The tumor was not infiltrating into intradural space or vertebral bone. The histopathologic study confirmed the epidural tumor as cavernous hemangioma. Postoperatively, his weakness improved gradually. Here, we present a case of pure spinal epidural cavernous hemangioma. We think cavernous hemangioma should be in mind in the differential diagnosis of the spinal epidural tumors.

Discussion:

Cavernous hemangiomas are composed of dilated sinusoidal capillaries lined by a single layer of benign endothelial cells[6,13,] which are separated by variably thin fibrous adventitia but not by neural parenchyma[14].

According to International Society for the Study of Vascular Anomalies (ISSVA) classification, vascular anomalies were classified into two subgroups: neoplasms and malformations[14]. “Hemangioma” is characterized by tumor-like endothelial proliferation and spontaneous involution; it refers to infantile or congenital hemangioma in this classification[4]. Vascular malformations are classified by the predominant type of vascular channel (i.e. capillary, cavernous, arteriovenous, or venous)[3,7,11]. Cavernous hemangioma is composed of dilated sinusoidal vascular channels, differential from capillary-sized vessels in capillary hemangioma[13].

On histopathologic examination, our case was composed of a large number of dilated vascular channels lined by thin endothelial cells. The prominent venous lakes and dilated vascular channels, mainly of the cavernous type, were filled with abundant of red blood cells. Although the tumor showed strong positive in cluster of differentiation (CD) 34 staining, it was negative on CD31 staining, which implies that there was no lymphatic component in our tumor. Thus, histopathologically it was confirmed as cavernous hemangioma.

Although PSECHs do not represent true neoplasms, they are dynamic lesions, and intralesional hemorrhage, thrombosis, organization, cyst formation and involution of the cavernous contribute to the changes in size and nature of these lesions with subsequent spinal cord compression, necessitating complete surgical removal as the treatment of choice.

Common clinical presentation of PSECH is a progressive myelopathy or radiculopathy in some patients back pain is the only clinical sign of the

lesion[6,9,13]. Acute onset of symptoms, especially with a very significant neurological deficit is rare[2,4,5,9,10] And it can be explained by a sudden increase in volume of the lesion, mainly due to hemorrhage or thrombosis of the draining veins[5,6,8,9]. Although intramedullary cavernomas show progressive myelopathy followed by acute deterioration, the protective effect of the dura and cerebrospinal fluid may play a role in preventing spinal cord injury in PSECH[2].

According to literature, PSECHs showed high T2 signal intensity and homogeneous, strong contrast enhancement on MRI[3,5,6,7,9]. This pattern is different from the intra axial cavernomas, which do not enhance homogeneously[2]. Although the intramedullary or intracerebral cavernous hemangiomas frequently show a peripheral low signal intensity rim, representing hemosiderin deposition from recurrent intra-lesional hemorrhage, PSECHs usually do not show a low signal rim like our case[2,5,13].

Despite the advancement of MRI technique, PSECHs are usually misdiagnosed as other lesions. The PSECH should be differentiated from other epidural, neoplastic or inflammatory lesions, such as meningioma, lymphoma, metastasis, and hemorrhagic vascular lesions[5,13]. Although they are less frequently found in the foraminal and extraforaminal regions, schwannoma or neurinoma should also be considered as differential diagnosis[6,12].

Because of the high vascularization of hemangiomas, preoperative misinterpretation may result in unexpected intraoperative hemorrhage and incomplete resection, which results in the persistence of clinical symptoms or recurrence[3,7]. Some authors recommended en-bloc removal to avoid intraoperative blood loss, which is possible only after accurate preoperative diagnosis[2,13]. Although bleeding is seldom a problem with intra-medullary cavernous hemangiomas during surgery, severe intraoperative bleeding may occur in spinal epidural lesions[6]. Reoperation for remnant or recurrent epidural hemangioma is very difficult because of peridural or periradicular adhesion and unclear tumor margins. As a result, complete resection cannot be guaranteed in reoperation. Therefore, proper preoperative planning and complete resection in the first operation is essential[7].

In summary, we believe that the accurate preoperative diagnosis and complete resection is indispensable to favorable treatment outcome and PSECHs should be considered as differential diagnosis for spinal epidural lesions.

Conclusion:

Pure epidural cavernous hemangiomas are very rare finding in spinal region. However, the hemangioma should be included in the differential diagnosis of spinal epidural tumors.

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