

Primary Dumbbell-Shaped Epidural Cavernous Hemangioma Mimicking a Thoracic Nerve Sheath Tumor: A Case Report

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Abstract

The author presents a rare case of dumbbell-shaped epidural cavernous epidural hemangioma in the thoracic spine mimicking nerve sheath tumor. A 57-year-old female presented with progressive weakness and hypoesthesia in both lower limbs. MRI revealed a dumbbell-shaped mass at vertebral levels T4 to T7 traversing the right neural foramen at T6. The lesion was isointense to the spinal cord on T1-weighted and hyperintense on T2-weighted images and showed intense homogeneous enhancement after contrast administration. The patient underwent total laminectomy from T4 to T6. Intraoperatively, a dorsal epidural reddish mass with good cleavage from the dura was detected. No dural attachment was noted. Total excision was achieved. The pathological diagnosis confirmed a cavernous hemangioma. The patient progressively improved and was ambulant at the time of discharge. Subsequently, the patient recovered completely. A repeat MRI 17 months after surgery revealed no residual or recurrence of the lesion.

Keywords: *Cavernous Hemangioma; Spinal Cord Tumor; Epidural Tumor; Spine Surgery*

Abbreviations

CNS: Central Nervous System; MRI: Magnetic Resonance Imaging

Background

Cavernous hemangioma is a benign vascular malformation that occurs primarily in the central nervous system (CNS). It accounts for 8 - 15% of all developmental vascular hamartoma in the CNS. Most of these lesions occur intracranially with supratentorial sites being most commonly affected [1].

Primary epidural cavernous hemangiomas presenting as a purely epidural lesion are rare. In particular, dumbbell-shaped epidural cavernous hemangiomas are extremely rare, and they are easily misdiagnosed as spinal nerve sheath tumors. To the best of our knowledge, only a few cases of dumbbell-shaped epidural cavernous hemangioma in the thoracic spine have been reported. In the current study, the authors describe a case of dumbbell-shaped epidural cavernous hemangioma involving spinal cord compression.

Case Presentation

A 57-year-old female presented with a 7-month history of progressive spastic weakness and hypoesthesia in both lower limbs. Neurological examination revealed grade 3 - 4 power in the lower extremities. All modalities of sensation (pin prick, touch, vibration, temperature) were diminished below level T8. The patient also exhibited hyperactive deep tendon reflexes in both lower limbs. Urinary and anal sphincter control was normal.

All routine blood investigations were within normal limits. Plain radiographs of the thoracic spine did not show any bony abnormality. Magnetic resonance imaging (MRI) revealed an intraspinal extradural tumor suggestive of a spinal nerve sheath tumor (a schwannoma or neurofibroma). It had grown into the thoracic spinal cord displacing it anteriorly. The tumor extended from the fourth to the upper margin of the seventh thoracic lamina traversing the right neural foramen at T6. This dumbbell-shaped lesion was isointense to the spinal cord on T1-weighted images, was hyperintense on T2-weighted images and showed intense homogeneous enhancement after contrast administration (Figure 1).



Figure 1: Thoracic T2-weighted sagittal MRI showing an epidural cavernous hemangioma at vertebral levels T4-T6 (a). Axial images reveal a dumbbell-shaped mass that is hyper-intense on T2-weighted images (b) images with homogeneous contrast enhancement (c).

Total laminectomies were performed from T4 to T6. Intraoperatively, no bony erosion was noted. An extradural reddish mass of 5 x 2 cm was detected dorsally extending to the right side and displacing the dura to the left. The right T6 nerve root was identified within the ventral part of the tumor. No dural attachment was observed. Using microsurgical technique, total excision of the mass was achieved. The histopathological diagnosis was a cavernous hemangioma. On histology, the lesion was composed of multiple closely spaced, dilated vascular channels containing blood (Figure 2).

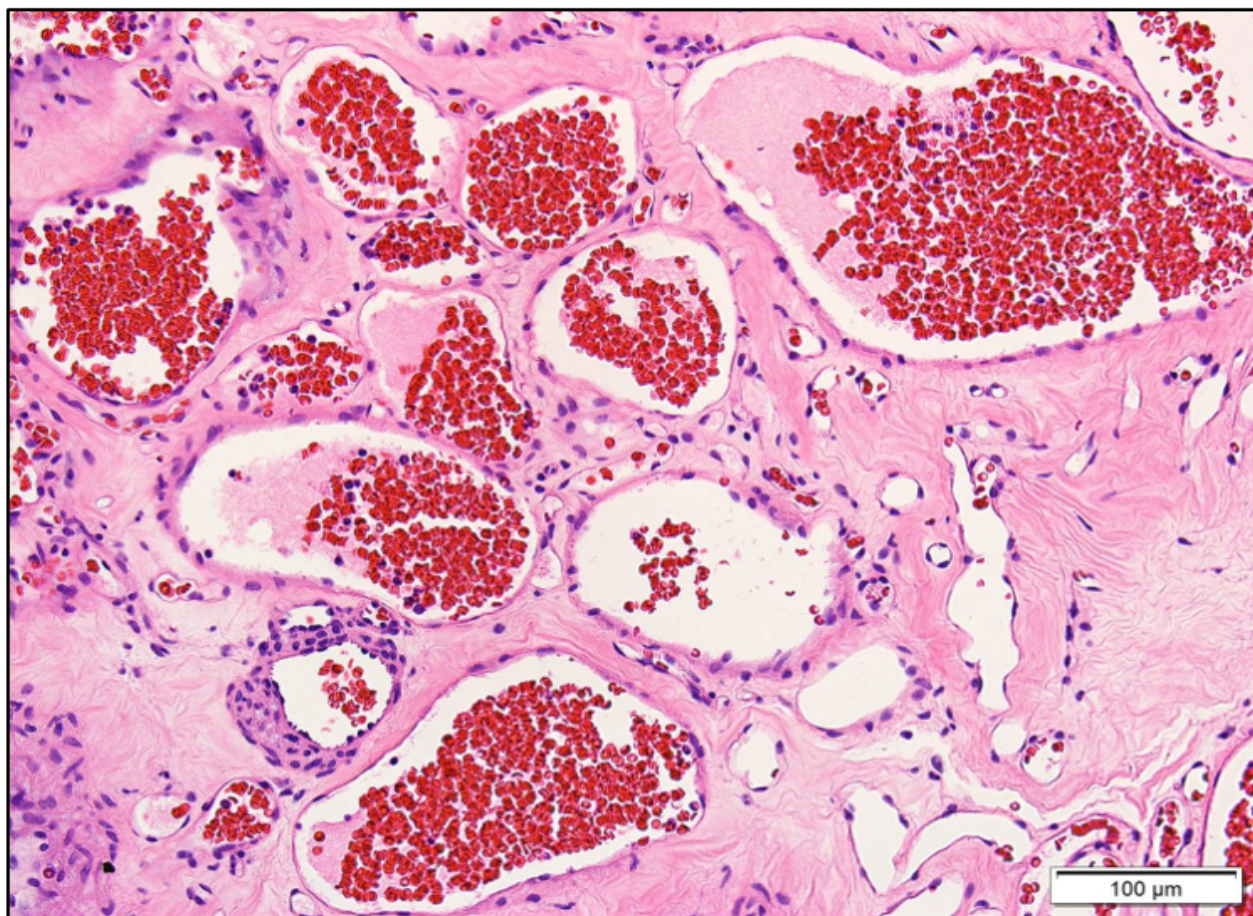


Figure 2: Histopathology showed large number of vascular channels in collagenous tissue containing blood, lined by a single layer of endothelium.

The patient progressively improved and was ambulant at the time of discharge. Subsequently, the patient recovered completely. A repeat MRI 17 months after surgery revealed no residual or recurrence of the lesion.

Discussion

Cavernous hemangioma is a developmental vascular malformation. It can occur anywhere in the central nervous system. Cavernous hemangiomas represent about 8 - 15% of all vascular malformations, and spinal vascular malformations comprise 3 - 12% of spinal space-occupying lesions [2,3]. Among them, pure spinal epidural cavernous hemangiomas without bone involvement are very rare. Histologically, cavernous hemangiomas are composed of mature thin-walled blood vessels and sinuses containing blood. The vessel walls are lined with a single layer of flattened endothelial cells in collagenous tissue without interposed neural tissue [4]. Residues of previous hemorrhages with fibrous scar tissue and hemosiderin calcifications are common, particularly in larger lesions [5].

Generally, about 80% of spinal epidural cavernous hemangiomas affect the thoracic spine, and most of them, in 93% of the cases, are located posteriorly within the spinal canal [1]. Two hypotheses try to explain the higher incidence of thoracic spinal cavernous hemangiomas. The first hypothesis attributes it to the larger size of the thoracic epidural space. The other one associates the higher incidence with the lower resistance of the posterior thoracic spinal canal [6,7]. The hemangioma location influences the clinical presentation; dumbbell-shaped hemangiomas in the thoracic spine mainly lead to myelopathy, whereas those in the lumbar spine predominantly present with radicular pain [8].

Spinal cavernous hemangiomas may present with local symptoms such as back pain or with radiculopathy or myelopathy. Typically, the clinical presentation of thoracic cavernous hemangiomas shows a slowly progressive spinal cord compression. Acute compression symptoms due to associated extradural hemorrhage are rarely observed [9]. Our case showed progressive para-paresis, and thoracic myelopathy was clinically suspected.

MRI is the diagnostic tool of choice. MRI shows T1 isointense and T2 iso- to hyperintense signal changes. Contrast enhancement may be minimal or absent. A plain computed tomography scan may show a high density or calcified lesion [10]. Digital subtraction angiography plays no role in the diagnosis of epidural cavernous hemangiomas as these lesions do not communicate with the spinal arterial circulation and are angiographically occult.

Spinal epidural cavernous hemangiomas are prone to bleeding and enlargement. Therefore, early surgical removal is strongly recommended. Total resection is the best treatment for epidural cavernous hemangiomas. Their potential for massive intraoperative bleeding, though rarely occurring, should be taken into consideration by the surgeon. In addition, the surgeon should be prepared for the possibility of a spinal nerve root compression or a hemangioma extension outside the spinal region [6].

Spinal dumbbell tumors were defined as a group of tumors arising along the spine. They are constricted at the point, where they penetrate the intervertebral foramen or dura mater, thereby acquiring a dumbbell shape [11]. Because a dumbbell shape is typically found in spinal schwannoma, we initially assumed this to be the diagnosis of the patient's thoracic tumor. However, it was pathologically confirmed as an epidural cavernous hemangioma. The spinal cavernous hemangioma's extension could occur tumor growth through the intervertebral foramen, but usually to a minor extent. Though rarely occurring, the extra-vertebral component may be enlarged and give rise to a dumbbell-shaped tumor, as seen in this patient.

Conclusions

The radiological presentation can be misleading in primary epidural cavernous hemangiomas. It is important to consider hemangiomas in the differential diagnosis of spinal epidural lesions with a dumbbell-shaped appearance in the MRI, especially if they present at the thoracic level. Awareness of the characteristics of epidural cavernous hemangiomas facilitates the diagnosis and treatment of these lesions.

Acknowledgments

None.

Conflict of Interest

There are no conflicts of interest.

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