CASE REPORT

# Acute Presentation of Solitary Spinal Epidural Cavernous Angioma in a Child

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## ABSTRACT

Solitary spinal epidural cavernous angiomas are rare lesions, especially in paediatric age group. They are infrequently considered in the differential diagnosis of spinal epidural masses in children. We report a case of solitary epidural cavernous angioma of the thoracic spine in a child presenting with acute onset of back pain and myelopathy. Magnetic resonance imaging of the thoracic spine demonstrated a posterior epidural mass at T6-T8 levels with compression of the spinal cord. Using microsurgical technique and bipolar coagulation, total excision of the lesion was achieved. Histopathological examination confirmed the diagnosis of cavernous angioma. At the five-year follow-up, there was no recurrence of the tumour.

**Key words:** *Cavernous angioma. Cavernoma. Epidural thoracic spine. Child.*

## INTRODUCTION

Cavernous angiomas or cavernomas are rare vascular malformations which may localize in all locations within the central nervous system, but seen quite rare in the spine, where they account for 4% of all spinal epidural tumours and 5 – 12% of all spinal vascular lesions.[1,2] In the spine, they are mainly located in the vertebral body with occasional epidural involvement.[3] Pure epidural cavernous angiomas of the spinal canal are extremely rare in children and are infrequently considered in the differential diagnosis of spinal epidural masses.[1,3-5]

We report a case of a focal epidural cavernous angioma of the thoracic spine in a child presenting with acute onset back pain and myelopathy. Clinical aspect, imaging, surgical findings, differential diagnosis and pathology of these lesions are discussed and the relevant literature is reviewed.

## CASE REPORT

A 13-year-old girl was admitted with acute onset of severe mid-thoracic pain and progressive paraparesis that occurred 20 hours prior to presentation. Three months before admission, she began to have complaints of mild mid-thoracic pain. She had no prior history of trauma.

On admission, neurological examination revealed spastic paraparesis, Babinski reflex, and hyperreflexia of the lower extremities. Hypoesthesia upto the transverse umbilical line on both sides was present. She also had urinary retention.

Radiography of the thoracic spine was normal; however, magnetic resonance imaging (MRI) demonstrated a posterior epidural mass at T6-T8 levels with compression of the spinal cord. The lesion was slightly hyperintense relative to the spinal cord on T1-weighted, and hyperintense on T2-weighted images with marked enhancement after gadolinium administration (Figure 1a and 1b). The T7-vertebral body showed low signal intensity on T1-weighted and high signal intensity on T2-weighted images but there was no enhancement in vertebral body after gadolinium administration.

The pre-operative diagnosis was Ewing sarcoma. Bilateral T7 and partial T6 and T8 laminectomy was performed, and no bony erosion was noted. Intra-operatively, a well-defined lobulated and dark red mass was encountered with central intralesional hematoma, located in the posterior and bilateral surface of the T7 epidural space; the mass displaced the spinal cord anteriorly. The lesion was highly vascular but was easily separable from the dura. Using microsurgical technique and bipolar coagulation, total excision of the lesion was achieved. Intraoperative bleeding was about 200 cc.

Histopathological examination of the excised specimen revealed compact sinusoidal-type vessels without intervening brain tissue. This confirmed the diagnosis of cavernous angioma (Figure 2).

Postoperatively, the patient's condition improved gradually after a period of rehabilitation and started walking after 3 months. At the 5-year follow-up, there was no recurrence of the tumour.

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*Received April 23, 2011; accepted August 01, 2012.*

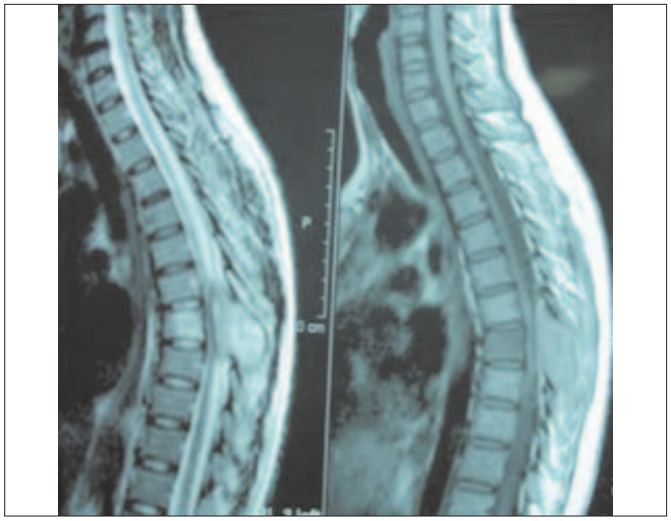

**Figure 1(a):** T1-weighted (right) and T2-weighted (left) sagittal MRI of thoracic spine demonstrated a posterior thoracic epidural lesion at T7 level that is hyperintense on both T1- and T2-weighted images.

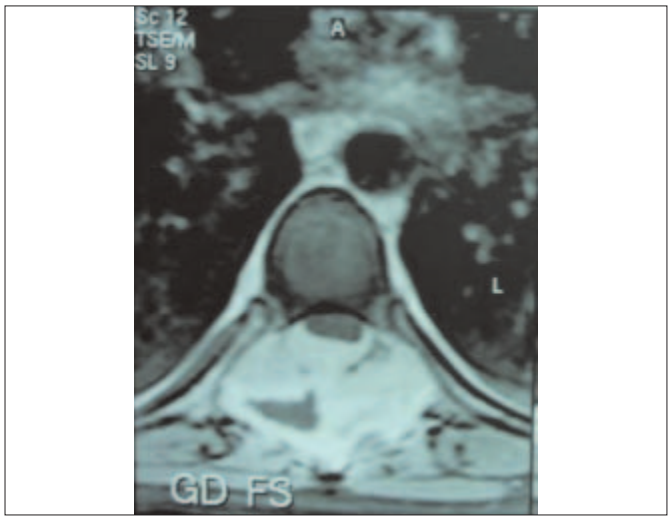


**Figure 1(b):** Axial T1-weighted gadolinium-enhanced MRI showing a strongly enhanced epidural mass at T7 level compressing the spinal cord anteriorly.

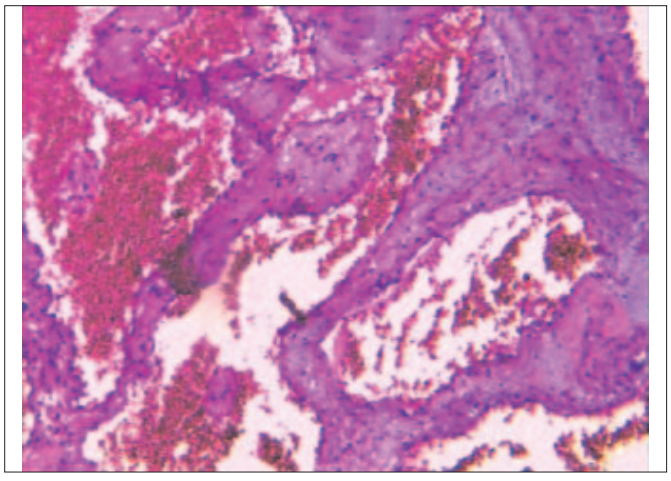

**Figure 2:** Histopathology of lesion showed compact sinusoidal-type vessels without intervening brain tissue.consistent with cavernous angioma.

## DISCUSSION

Spinal epidural cavernous angioma is a rare but well-known developmental pathology that causes epidural compression of the spinal cord in children.[1-5] Cavernous angiomas are well-circumscribed, mulberry-like lesions, consisting of closely packed, capillary-like vessels, without intervening neural tissue with a simple endothelial lining.[6,7] They are usually located in the posterior part of the epidural space at the thoracic spine.[6,8-10] They are usually confined to two or more spinal levels, and have an oval shape, sometimes flattened on the spinal cord rather than distorting it.[6] Solitary spinal epidural cavernous angioma is very rare in children. Only 7 cases were found to have been occurred in paediatric age group (age under 18 years).[1-5]

The spinal epidural cavernous angiomas are mainly situated posterolateral to the spinal cord, with less frequent extension into the neural foramen and extra spinal components,[2,10] as in the presented case. The larger available space and the lower resistance in the posterior portion of the spinal canal compared with the anterior portion may be an explanation for the mainly posterolateral localization of the lesions.[2,5,6]

The clinical presentations of spinal epidural cavernous angioma are determined by the anatomic level of the spine in which they are located and the pathophysiologic progression of the lesion and can be divided into three major patterns[5]: acute haemorrhage into the lesion; acute haemorrhage surrounding the lesion; and slow seeping haemorrhage.

The clinical course of spinal epidural cavernous angioma is commonly insidious,[5,6,8] however, a sudden increase in the size of the lesions following haemorrhage may cause acute clinical symptoms.[2] In the presented case, intralesional haemorrhage, as observed intra-operatively, caused acute cord compression and resultant paraparesis.

On MRI, the lesion is isointense with the spinal cord on T1-weighted images because of the slow blood flow and is hyperintense on T2-weighted images owing to the content of stagnant blood and shows homogeneous strong enhancement because of the high vascularization and sinusoidal channel structure.[2,5-7,9] In the presence of haemorrhage inside the lesion, it can be hyperintense on both T1 and T2-weighted MR images,[6,7] as was shown in this patient.

The main differential diagnoses of spinal epidural cavernous angioma would include extraosseous Ewing sarcoma, lymphoma, metastasis, eosinophilic granuloma, neurogenic tumour, multiple myeloma, meningioma, extruded disc, and rarely angiolipoma.[2,3,6,9]

Given the benign nature of cavernous angioma and the good neurological outcome reported in the literature,

their management should be a complete microsurgical removal. This should not only obtain neurological improvement, but also achieve a definitive diagnosis of a lesion not always easy to interpret radiologically pre-operatively.

*En-bloc* resection, which can avoid intraoperative bleeding, is recommended,[2,4-6] radiosurgery of these lesions alone has been reported with good result.[10] Sub-total removal may lead to re-appearance of symptoms and continued progressive deterioration as a result of bleeding from residual malformation.[2] Radiotherapy after surgery has been suggested for spinal epidural cavernous angiomas; when their extension into or out of the spinal canal makes their total removal impossible.[5]

The result of surgical resection appears to be related largely to the patient's pre-operative neurological status. As the chance for complete recovery diminishes with worsening pre-operative neurological status, it is important to operate on these patients early in their course before they develop severe or long-standing neurological deficits.[1] The potential for delayed spinal deformities in the paediatric age group and the need for regular, radiologic follow-up should be recognized. This is particularly important when the surgical approach requires an extensive degree of bone resection.

## REFERENCES


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