Review of Spinal Epidural Cavernous Hemangioma

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Abstract

The characteristics of spinal epidural cavernous hemangioma without primary origin in the vertebral bone were evaluated in 54 patients including our new case. The 36 male and 18 female patients were aged 5 to 78 years (mean 47 years). Most lesions were in the thoracic spine (80%) and on the dorsal side of the spinal cord (93%). The clinical course was mostly slowly progressive, with myelopathy in 33% at onset and 83% at admission. The lesion appeared isointense to the spinal cord on T1-weighted imaging, and isointense or slightly hypointense to the cerebrospinal fluid on T2-weighted imaging. Lesion without hemorrhage showed prominent homogeneous enhancement after administration of gadolinium-diethylenetriaminepenta-acetic acid because of the sinusoidal channel structure. Heterogeneous enhancement was caused by hematoma and/or post-hemorrhagic degeneration. The differential diagnosis of this disease includes metastatic tumor, Ewing’s sarcoma, chordoma, eosinophilic granuloma, sarcoidosis, lipoma, hypertrophy of the posterior longitudinal ligament or the ligamentum flavum, meningioma, and neurinoma. The relationships between clinical course and surgery or outcome suggest that early diagnosis and total removal of the lesion before massive lesional bleeding occurs are necessary for a good outcome.

Key words: spine, cavernous hemangioma, hemorrhage, early operation

Introduction

Spinal epidural cavernous hemangioma accounts for 4% of all spinal epidural tumors, mostly occurring as a primary lesion in the vertebral bone. Spinal epidural cavernous hemangioma without a primary origin in the vertebral bone is rare, with only 53 reported cases.1–6,8,9,11–17,19,20,23–29,31,32,34–39,42,43,45 Here we describe a typical case, as well as the clinical characteristics of this disease, especially the clinical course, differential diagnosis between this disease and other spinal extra-axial diseases, and surgical treatment.

Materials and Methods

Fifty-four patients, including our case, were selected between 1932 and 2001. Eleven characteristics were selected, age, sex, lesion site, onset symptom, course, neurology at admission, neuroradiological findings, treatment, findings at operation, outcome, and course after discharge, to clarify the clinical characteristics of this disease. To identify the most common site of the lesion, the spine was divided into the cervical, thoracic, and lumbosacral parts, and lesions extending into two parts were counted one lesion in each part, so the 54 cases contained 65 lesions. The symptoms were classified into local disorders, radiculopathy, and myelopathy, and the most severe symptom was selected for assessing the clinical changes. Surgical treatment was classified into three categories, partial, subtotal, and total removal. Subtotal removal was defined as total removal of the intracanal lesion and residual mass in the intervertebral foramen and/or thoracic area. The clinical outcome was classified into five grades according to the Glasgow Outcome Scale.22

Results and Discussion

The 54 patients with spinal epidural cavernous hemangioma not originating in the vertebral bone consisted of 36 men and 18 women aged from 5 to 78 years (mean 48 years in males and 45 years in females). Associated skin findings of telangiectasia and/or pigmentation were found in only two patients.23,24 The thoracic spine contained 52 (80%) of 65 lesions. The lesions were located in dorsal side in 50 (93%) of 54 cases with/without lateral extension.

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The symptom at onset was classified as local disorder in 17 of the 54 cases, radiculopathy in 19, and myelopathy in 18, whereas the symptom at admission was no neurological deficits in three of the 54 cases, radiculopathy in six, and myelopathy in 45. Such progressive clinical deterioration can be explained by the fact that cavernous hemangioma is a benign hamartomatous vascular anomaly and increases in volume gradually. Sudden or intermittent clinical deterioration in the course of this disease might be caused by a more rapid increase of lesional volume due to thrombotic venous occlusion, neovascularization by estrogenic factor, or drainer compression by an enlarged pregnant uterus.

Magnetic resonance (MR) imaging showed the lesion nature more clearly than computed tomography. Our typical case is shown in Fig. 1. Table 1 summarizes the results of MR imaging in 17 patients. Lesions appeared isointense to the spinal cord on T1-weighted imaging in 12 of 17 cases, and isointense or slightly hypointense to cerebrospinal fluid on T2-weighted image in 16. An exceptional finding of hyperintensity on both T1- and T2-weighted imaging was explained by the coexistence of hematoma in the subacute phase. The lesions

<table>
<thead>
<tr>
<th>Signal intensity</th>
<th>MR sequence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypointense to T1-weighted</td>
<td>4</td>
</tr>
<tr>
<td>Isointense to T2-weighted</td>
<td>12</td>
</tr>
<tr>
<td>Hyperintense to spinal cord</td>
<td>1</td>
</tr>
<tr>
<td>Not reported</td>
<td>3</td>
</tr>
</tbody>
</table>

*Of 15 cases, 11 cases were or might be homogeneously enhanced, three cases were heterogeneously enhanced, and another one was not enhanced. Gd-DTPA: gadolinium-diethylenetriaminepenta-acetic acid.
Table 2  Relationship between course and outcome

<table>
<thead>
<tr>
<th>Course</th>
<th>Glasgow Outcome Scale</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Good recovery</td>
</tr>
<tr>
<td>Sudden onset</td>
<td>2 (1)</td>
</tr>
<tr>
<td>Progressive, ≤ 1 yr</td>
<td>18 (1)</td>
</tr>
<tr>
<td>Progressive, &gt; 1 yr</td>
<td>4</td>
</tr>
<tr>
<td>Progressive, unclear term (not sudden onset)</td>
<td>5 (1)</td>
</tr>
<tr>
<td>Intermittent</td>
<td>9 (1)</td>
</tr>
<tr>
<td>Total</td>
<td>38 (4)</td>
</tr>
</tbody>
</table>

Six of the 54 cases were excluded because course or outcome was not reported. Numerals in parentheses indicate number of hemorrhage from the lesion. *Patient who suffered sudden onset of myelopathy and died after 3 years without surgery. **Patient who died of pulmonary embolism.

were homogeneously enhanced by gadolinium-diethylenetriaminepenta-acid (Gd-DTPA) administration because of the sinusoidal channel structure. One case with peripheral enhancement revealed the degenerated part at the center of the lesion. 9 Another case with no enhancement was certified as sclerohyaline degeneration of the lesion. 35 Angiography is not useful for the diagnosis since epidural cavernous hemangioma has no communication with the spinal cord arteries. 21

MR imaging can provide indications for the differential diagnosis of spinal epidural cavernous hemangioma. Various diseases should be considered as follows. Metastatic tumor, which is associated with bone erosion of the adjacent vertebral bone, tends to enclose the cord epidurally. Ewing’s sarcoma often appears heterogeneous because of hemorrhage, calcification, and necrosis. Enhancement by Gd-DTPA occurs in the lesion located both in the epidural space and in the adjacent vertebral bone. 22 Chordoma occurs anywhere in the spine, 10,18 but predominantly in the sacrum and clivus. Destruction of the vertebral bone and the disc and surrounding osteosclerosis can be detected. 30 Chordoma is less enhanced by Gd-DTPA than cavernous hemangioma. Eosinophilic granuloma of the spine is a well-defined osteolytic lesion of the vertebral bone, and is associated with epidural mass. 7 Other osteolytic changes in the long bone, rib, pelvis, and skull are useful for the differential diagnosis. Sarcoidosis of the leptomeningeal type may resemble spinal epidural cavernous hemangioma on MR imaging with Gd-DTPA, but may appear thinner and/or with wider extension in the subdural space. 1,33 Prominent lymph nodes, especially at the hilus, and Kveim’s reaction are useful findings for differential diagnosis. Lipoma is distinguished by hyperintensity on T1-weighted and hypointensity on T2-weighted imaging. Dural-based lymphoma may show similar findings to epidural cavernous hemangioma. This type of lymphoma extends along the Virchow-Robin space, so may be identified as an intradural extra-axial lesion by fluid-attenuated inversion recovery imaging. 41 Hypertrophy of the posterior longitudinal ligament, which is located on the ventral side of the spinal cord, appears isointense to the cord on T1-weighted imaging and is only slightly enhanced by Gd-DTPA administration as a thin irregular venous plexus in the lesion. Hypertrophy of the ligamentum flavum, which is located on the dorsal side of the spinal cord, shows similar findings to hypertrophy of the posterior longitudinal ligament without enhancement. Neurinoma and meningioma may show similar MR imaging findings to spinal epidural cavernous hemangioma, even on T1-weighted imaging with Gd-DTPA. Enlarged intervertebral foramen is less meaningful for the differential diagnosis. Homogeneous enhancement may be more prominent in epidural cavernous hemangioma than neurinoma because of the sinusoidal channel structure. Osteolytic or osteoblastic changes in the adjacent bone and signal voids due to calcification are distinctive findings of meningioma, especially in the psammomatous type. 3

Table 2 shows the relationship between the clinical course, leional hemorrhage, and outcome. All nine patients with intermittent neurological deficit had a good recovery, and leional hemorrhage was identified in only one patient. Thirty-two of the 34 patients with a progressive course had a good recovery or moderate disability, and leional bleeding was observed in two patients. Two of the five patients with sudden onset had a good recovery, whereas three had severe disability or died, and leional
## Table 3 Treatment and outcome

<table>
<thead>
<tr>
<th>Treatment</th>
<th>Good recovery</th>
<th>Moderate disability</th>
<th>Severe disability</th>
<th>Vegetative state</th>
<th>Died</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total or subtotal removal</td>
<td>39 (9)</td>
<td>5</td>
<td>2</td>
<td>0</td>
<td>1*</td>
</tr>
<tr>
<td>Partial removal</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Conservative</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>39 (9)</td>
<td>5</td>
<td>3</td>
<td>0</td>
<td>2</td>
</tr>
</tbody>
</table>

Five of the 54 cases were excluded because course or outcome was not reported. Numerals in parentheses indicate the number of cases of brisk bleeding during surgery. *Patient died of pulmonary embolism.

bleeding was found in four patients. These results suggest that the spinal cord is fragile to the sudden onset of massive compression.

Table 3 shows the relationship between treatment and outcome. Forty-four of the 46 patients who underwent total or subtotal removal had a good recovery or moderate disability. One patient with partial removal had severe disability. One patient treated conservatively died of rebleeding 15 months after the initial hemorrhage. Brisk hemorrhage from the lesion occurred in nine cases during surgery but had no influence as all patients had a good recovery. One patient suffered recurrence after partial and subtotal removal respectively, but he had a good recovery finally after the third operation with total removal of the lesion.\(^{12}\)

Follow-up duration was from several weeks to 7 years, so the long-term outcome for patients with subtotal removal remains to be proved. However, early diagnosis and total removal of spinal epidural cavernous hemangioma are recommended before massive bleeding from the lesion, since such bleeding causes severe spinal cord damage.

## References


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Commentary on this paper appears on the next page.
Commentary

This review article describes 54 cases of spinal epidural cavernous hemangioma including one case of the authors. They discuss the characteristic MRI findings of epidural cavernous hemangioma and the exact differential diagnosis. This is an informative article, in particular for young neurosurgeons. Another interesting description of this paper is the relationship between surgical treatment and outcome. Forty-four of 46 patients with total or subtotal removal had good recovery, in contrast to one patient with partial removal who suffered severe disability and one patient with conservative treatment who died of bleeding later. This is not a surprising finding, but is instructive, as this promotes early diagnosis and total removal before catastrophic bleeding as the authors point out.

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The authors review the previously reported cases of spinal epidural hemangioma including a single case of their own. The value of this paper is that it brings to our attention a rare cause of epidural spinal compression that most neurosurgeons never see or treat. The clinical features, treatment results, and differential diagnosis are discussed. One entity, not mentioned by the authors, is the equally rare angiolipoma, which can have identical radiographic features and an indistinguishable clinical presentation. In fact there may be some overlap between these two types of tumors. It would have been nice if they had given more details regarding their personal case with pathologic information and surgical description. Similar to spinal dural arteriovenous fistulae, these lesions have a predilection for the thoracic spine location in men. The common dorsal location and benign nature make them amenable to low risk surgical extirpation as pointed out in this paper. Typically, the presentation is an indolent one, which allows the spinal cord to adapt over many years to external compression. Only rarely, when the patient has a more apoplectic presentation due to intratumoral hemorrhage or swelling, is a less than favorable outcome more likely. This is not surprising given the sensitive nature of the spinal cord to rapid changes in its local environment.

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