Capillary hemangioma of the spinal cord

Report of four cases

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The authors describe the clinicopathological features of four cases of capillary hemangioma of the spinal cord. All occurred in adult patients. The presenting symptoms were similar to those of more common intramedullary tumors. Radiologically, they resemble other vascular spinal cord tumors. All patients underwent surgery, and the outcomes varied. Histologically, the lesions resembled capillary hemangioma of skin or of soft tissue that is composed of lobules of small capillaries with associated feeding vessels, all enveloped by a delicate fibrous capsule. Capillary hemangiomas of the central and peripheral nervous system are extremely rare. Although examples of these lesions have been described as occurring in the dura mater and in peripheral nerve, including spinal nerve roots, none has occurred within the spinal cord. Knowledge of their existence may help practitioners to avoid misdiagnosis of tumor and resultant overtreatment of these benign lesions.

KEY WORDS • vascular malformation • hemangioma • capillary • spinal cord

Capillary hemangiomas are benign vascular lesions most often encountered in the skin and soft tissues of children. Histologically, they are characterized by lobular architecture, with each lobule fed by a large vessel and consisting of numerous capillaries lined by flattened endothelium. In the “juvenile” variant of capillary hemangioma, vascular lumens are narrowed by plump endothelial cells, resulting in a solid-appearing lesion. Capillary hemangiomas of the central and peripheral nervous system are very rare. Documented examples have been shown to arise in the dura and in spinal nerve roots. One vascular lesion, loosely termed “hemangioma” but not conforming to the morphological definition, was reported to have occurred in spinal cord parenchyma. In the present report, we describe the clinicopathological features of four classic capillary hemangiomas of the spinal cord. In both clinical and radiological terms, all mimicked neoplasms.

Case Reports

Clinical Findings

This report summarizes four cases, data on three of which were retrieved from the consultation files of one of the authors (B.W.S.). The patients underwent surgery between January 1980 and November 1998 at different institutions. The patient in the fourth case was treated at the Institute of Anatomic Pathology, Bellaria Hospital, in Bologna, Italy. Detailed clinical data, operative reports, and histological sections were available in all cases. Radiological studies or reports were also reviewed (Fig. 1).

The clinicopathological data obtained in the four cases are reported in Table 1.

Histological Findings

All specimens were fixed in 10% buffered formalin, routinely processed, and embedded in paraffin. Five-micron sections were stained with hematoxylin and eosin and by Gomori’s silver impregnation method for reticulin fibers. Immunohistochemical analysis was performed using the avidin-biotin-peroxidase complex method and polyclonal antisera directed against NSE (dilution 1:1000, Dako, Carpenteria, CA).

Histologically, all lesions were demarcated from spinal cord parenchyma by a fibrous pseudocapsule and featured a distinctly lobular architecture. Fed by sizeable, thick-walled arteries, each lobule was composed of numerous capillary-sized vessels lined by a single layer of cytologically benign endothelial cells. A minor component resembling “juvenile hemangioma” with its plump endothelial cells was seen in each case, but no cavernous elements were noted. In all lesions some

Abbreviation used in this paper: NSE = neuron-specific enolase.
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Stromal edema and occasional endothelial and stromal mitoses were encountered. No neural parenchyma was present within or at the periphery of the lesions. Scattered stromal lymphocytes were present in only one patient (Case 1). Indicators of bleeding, such as extravasated erythrocytes and hemosiderin deposits were absent. Gomori’s silver impregnation stain highlighted the delicate network of reticulin fibers surrounding the vessels (Fig. 2 lower). Neither foamy stromal cells nor NSE immunoreactivity was observed.

Discussion

The purpose of this study was to assess the clinicopathological features of four capillary hemangiomas of the spinal cord, which is an exceedingly rare lesion.

Presenting symptoms were those common to all patients with intramedullary tumors. The major points of distinction between patients with these lesions and other spinal cord neoplasms were the absence of severe neurological deficits at diagnosis and the discrete, noninfiltrative nature of the lesions. In all cases, neuroimaging studies were suggestive of a highly vascular neoplasm arising from the posterior surface of the spinal cord or conus medullaris. Understandably, a preoperative diagnosis of hemangioma was not made in any of our cases.

Capillary hemangioma can be distinguished from cavernous angioma. Three of our patients were men older than the age of 50 years, and cavernous angiomas are more often seen in young adult women. Clinically, whereas capillary hemangiomas are associated with slowly progressive worsening of symptoms, cavernomas often present acutely with symptoms referable to bleeding. Magnetic resonance–depicted features of spinal cord cavernous angiomas include heterogeneous high and low signal intensity consistent with subacute and chronic hemorrhage, as well as a peripheral T1 rim related to hemosiderin deposition, a feature lacking in our cases. The distinction of capillary hemangioma from such highly vascular spinal cord tumors as hemangioblastoma and hemangiopericytoma requires histological examination.

In all our patients a one- or two-level laminectomy was performed using standard microsurgical techniques to achieve a resection. Review of the operative notes indicated that the lesions presented at the spinal cord surface, thus facilitating their exposure and dissection. On gross inspection, the hemangiomas appeared as red or blue subpial nodules. Soft in texture, they were well demarcated from the spinal cord parenchyma. A subtotal removal was performed in one case because an erroneous preliminary diagnosis of hemangiopericytoma was made.

Surgical outcomes were comparable with those associated with more common intramedul lary tumors. Specifically, the patients experienced a slight worsening of neurological status immediately after surgery. Deficits in proprioception, presumably due to the surgical manipulation of the posterior column, were marked in two cases. Bladder function worsened in one case and was stable in the others. In three cases, motor deficits improved substantially over a 6-month postoperative interval. On balance, sensory disturbances improved to a lesser degree. No improvement in sphincter function was observed.

As with other architecturally solid lesions, complete surgical excision is the goal. In the two cases in which this was achieved, no clinical recurrence has been demonstrated during an average follow-up period of 10 years. In cases in which a complete resection is not possible, postoperative serial magnetic resonance imaging seems warranted.

Histologically, our four lesions were identical to capillary hemangiomas of skin or soft tissues and differed significantly from both intradural arteriovenous malformation and capillary telangiectasia. Both of the latter tumors invariably feature a racemose architecture with parenchyma between vessels, and they lack lobularity as well as demarcation from the surrounding parenchyma. Unlike capillary hemangiomas, cavernous hemangiomas consist of dilated hyaline vessels and often show thrombosis, perivascular hemosiderin deposition, and calcification. Rare tumorlike, unclassifiable vascular malformations have also been described as occurring in the spinal cord. These have been described as featuring clusters of capillary-sized blood vessels with prominent mural hyalination and spinal cord tissue between the vessels.

Capillary hemangiomas must be distinguished from highly vascular neoplasms of the spinal cord. The solid appearance and large feeding vessels of capillary hemangiomas may mimic the cellularity and stag-horn vasculature of hemangiopericytoma. Indeed, two of the lesions were originally diagnosed as hemangiopericytomas. Unlike the latter, capillary hemangiomas feature lobularity and lack pericellular deposition of reticulin fibers. Despite their mimicry of capillary hemangioma, the reticular variant of hemangioblastoma contains foamy NSE-reactive stromal cells. Hematoxylin and eosin staining can be performed to exclude other spinal cord tumors such as angiolipoma.
spindle cell hemangioendothelioma, meningioma, schwannoma, solitary fibrous tumor, and metastases. Although a capillary component is occasionally observed in cavernous angiomas of the spinal cord, no pure capillary hemangiomas have been described to date. We are aware of the case report of Hida, et al., in which the authors describe an intramedullary vascular lesion in the cervical cord of a 50-year-old man. Despite its designation as a “capillary hemangioma,” the authors described the coexistence of capillary-like and telangiectatic vessels in addition to venules within edematous, partially necrotic parenchyma. Their lesion was diffuse, lacked lobular architecture, and featured spinal cord tissue between blood vessels. These are not the features of capillary hemangioma.

In conclusion, it is important for surgeons to be aware of the existence of capillary hemangiomas of the spinal cord. Such lesions must be distinguished from true tumors with aggressive potential, particularly hemangiopericytoma, so that overtreatment can be avoided.

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References


TABLE 1
Clinicopathological findings obtained in four cases of spinal cord capillary hemangioma

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Symptom</th>
<th>Duration (yrs)</th>
<th>Neurological Deficits</th>
<th>Site</th>
<th>Original Diagnosis</th>
<th>Therapy</th>
<th>Follow Up (yrs)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>42, F</td>
<td>leg weakness, abdominal pain</td>
<td>1.5</td>
<td>mild leg weakness</td>
<td>T-11</td>
<td>hemangioma</td>
<td>surgery</td>
<td>recovery (11)</td>
</tr>
<tr>
<td>2</td>
<td>50, M</td>
<td>lower-back pain, weakness</td>
<td>1</td>
<td>bilat decrease in proximal leg strength, absent tendon knee reflexes, sensory level at L-1</td>
<td>T-11</td>
<td>HPC</td>
<td>surgery &amp; radiotherapy</td>
<td>little improvement (2)</td>
</tr>
<tr>
<td>3</td>
<td>53, M</td>
<td>back, left calf pain</td>
<td>2</td>
<td>upper, lower motor neuron signs in both legs</td>
<td>conus</td>
<td>HPC</td>
<td>surgery</td>
<td>leg weakness (1.5)</td>
</tr>
<tr>
<td>4</td>
<td>64, M</td>
<td>bilat leg pain &amp; weakness</td>
<td>2</td>
<td>mild leg weakness</td>
<td>T-10</td>
<td>capillary hemangioma</td>
<td>surgery</td>
<td>recovery (9)</td>
</tr>
</tbody>
</table>

* bilat = bilateral; HPC = hemangiopericytoma.

Fig. 2. Case 4. Photomicrographs. Upper: The lesion features a conspicuous lobular architecture. H & E, original magnification × 4. Lower: The delicate network of reticulin fibers surrounding the delicate vessels of capillary hemangiomas can be seen. Gomori’s silver impregnation stain, original magnification × 100. 
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