Hemangiopericytoma of the spinal canal

Report of three cases

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Hemangiopericytoma is a vascular neoplasm consisting of capillaries outlined by an intact basement membrane that separates the endothelial cells of the capillaries from the spindle-shaped tumor cells in the extravascular area. These neoplasms are found in soft tissues but have rarely been shown to involve the spinal canal. This is a report of three such cases. Surgical removal of the tumor from the spinal canal was technically difficult. A high risk of recurrence has been reported but in these three cases adjunctive radiotherapy appeared to be of benefit in controlling the progression of the disease. These cases, added to the six cases in the literature, confirm the existence of hemangiopericytoma involving the vertebral column with extension into the spinal canal. This entity should be included in the differential diagnosis of lesions of the spinal canal. The risk of intraoperative hemorrhage should be anticipated.

KEY WORDS  •  hemangiopericytoma  •  spine  •  spinal canal  •  radiotherapy  •  surgery  •  survival

Hemangiopericytoma, first described by Stout and Murray in 1942, is a vascular neoplasm associated with numerous capillaries. The capillaries are outlined by an intact basement membrane that separates the endothelial cells of the capillaries from the ovoid or spindle-shaped tumor cells in the extravascular area. The tumor does not have an organoid structure, is often without encapsulation, and lacks the neural component often found in glomus tumors. The neoplastic cells are thought to arise from the Zimmerman pericyte. The cells, which have contractile powers, are thought possibly to be related to smooth-muscle cells. Studies have shown the presence of bundles of filamentous structures within the cytoplasm of the tumor cells. The neoplasms are found in the soft tissues of the body; they are uncommon in the cranial cavity and have rarely been shown to involve the spinal canal. In this report we add three new cases with spinal canal involvement to the six already reported in the literature in an attempt to better characterize this uncommon lesion.

Case Reports

Case 1

This 28-year-old man presented in May, 1973, with gradual onset of weakness of grip in his right hand and paresthesia down the inner aspect of his right arm. The remainder of his history was non-contributory.
Fig. 1. Case I. Upper: Electron micrograph showing intact endothelium, a prominent basement membrane with a sprinkling of collagen fibrils, and the neoplastic cells beyond the basement membrane. × 5180. Lower: Photomicrograph showing intact capillaries with prominent endothelial cells that are distinct from the surrounding tumor. H & E, × 440.

Examination. He had weakness of the right finger flexors and interossei. Touch and pain sensation were decreased in the C-5 and C-6 distribution on the right. All reflexes were equal bilaterally, but abnormally brisk. His abdominal reflexes were absent in all four quadrants and clonus was elicited at both ankles. Cerebellar function tests revealed dysmetria with past pointing on the right. His gait was normal and no other abnormal physical signs were found.

Cervical spine films demonstrated a congenital fusion between the C-2 and C-3 vertebrae. The intervertebral foramina on the
right side of C2-4 were enlarged. Cervical myelography revealed a complete obstruction of the extradural space at C-4, with displacement of the spinal cord to the left. The cerebrospinal fluid protein was 124 mg/100 ml. All other investigations were normal.

**Operation.** A cervical laminectomy was performed from C1-7. The tumor was exposed lying in the lateral epidural space, displacing the cord to the left and extending anteriorly to the cord from C2-6, involving the bone of C-2. The tumor was lobulated, red-brown in color, and highly vascular. It was adherent to the underlying dura. Resection of the tumor was extensive but clearly subtotal.

**Postoperative Course.** Postoperatively, 5000 rads were delivered to the area through anterior and posterior cervical fields in 25 treatments over 4 weeks. The patient’s neurological status returned to normal. Subsequently, he required an extensive anterior cervical fusion from C3-7 for instability. At last follow-up examination in December, 1977, there was no evidence of tumor recurrence.

**Pathological Findings.** The tumor was composed of masses of closely packed spindle-shaped cells with densely staining nuclei (Fig. 1). There were many vascular channels with a distinct endothelial lining and a well demarcated basement membrane. The tumor cells were arranged on the external aspect of this vascular meshwork. The fact that the capillary basement membrane was intact was confirmed by silver impregnation. In the tumor cells there were occasional mitotic figures, and a moderate degree of pleomorphism. These histological features were characteristic of hemangiopericytoma. Electron microscopic examination further substantiated this diagnosis (Fig. 1 upper).

**Case 2**

This 65-year-old Caucasian woman had a known history of adenocarcinoma of the uterus, which was diagnosed in 1970 and had been treated with external Betatron therapy (5000 rads) and internal application of caesium (3673 rads). Since 1972, she had been given intermittent intramuscular courses of Depo-Provera (medroxyprogesterone acetate). In May, 1973, she presented with pain radiating down her right leg from the hip to ankle on the lateral aspect.

**Examination.** No abnormal neurological signs were elicited. Roentgenograms demonstrated a soft-tissue lesion lying posterior to the L-2 vertebral body. It had a well defined anterior margin eroding the posterior surface of the vertebral body, with erosion of the pedicle and left lamina. A lumbar myelogram revealed multiple serpiginous filling defects in the subdural space, with a large filling defect in the region of the eroded body of the L-2 vertebra.

**Operation.** Laminectomy was performed at the L1-3 vertebrae. The tumor measured $3 \times 2 \times 0.5$ cm, and was lying in the extradural space, adherent to the underlying dura. It involved the L-2 vertebra, causing erosion of the pedicle, facet joints, and lamina on the left. It was red-brown in color and bled profusely. Total resection was not possible, and decompression was carried out above and below the remaining tumor.

**Postoperative Course.** The patient received 3000 rads to the thoracolumbar spine in 10 treatments over 2 weeks, and her symptoms were completely relieved. She died in December, 1976, from complications of the adenocarcinoma of the uterus. There had been no clinical evidence of tumor recurrence in her lumbar region. An autopsy was not performed.

**Pathological Findings.** Examination of the uterine tumor revealed a typical adenocarcinoma. The tumor tissue removed from the extradural space was different (Fig. 2); it was a vascular tumor composed of closely packed spindle-shaped cells. The cells were regular in pattern, and mitotic figures were scarce. The vascular channels were lined by endothelial cells. Silver impregnation demonstrated an intact basement membrane surrounding the endothelium of the capillaries, and separating it from the spindle-shaped cells of the tumor. An abundant reticulin meshwork permeated the entire tumor tissue. These microscopic features established a diagnosis of hemangiopericytoma.

**Case 3**

**History.** This 46-year-old Caucasian man presented in January, 1972, with a nodule 2 cm in diameter under the skin of his groin. Pathological examination of the tissue after excision revealed the characteristic appearance of a hemangiopericytoma. Postoperatively he was irradiated with 2500 rads.
Spinal hemangiopericytoma

Fig. 2. Case 2. Photomicrographs of excised tumor tissue. Left: The neoplasm has a more diffuse pattern than that in Case 1, while the capillaries show an intact endothelial lining. H & E, × 400. Right: Reticulin stain confirms the presence of intact basement membrane separating the endothelial cells from the surrounding tissue. An abundance of reticulin fibrils is noted in the intercellular compartment of the neoplasm. Reticulin stain, × 400.

to his right groin and 1600 rads to his right thigh. The only notable features in his history was that he had been undergoing treatment for gout with allopurinol since 1966.

In December, 1972, he presented again with weakness of both legs. Muscle atrophy was present bilaterally in his lower limbs, pectoralis majori, and infraspinatii. Deep tendon reflexes were absent bilaterally at the ankle, but normal elsewhere. Bilateral ankle clonus was elicited, but sensation was normal. Roentgenograms revealed narrowing of the C-5 intervertebral space, and minimal cervical spondylitis. The only abnormality in the electromyogram was defibrillation potentials in the left gastrocnemius. Over the next 3 years his neurological status remained constant.

In July, 1975, lung metastases were found and biopsied. They had the characteristic pattern of hemangiopericytoma. A course of irradiation was given as follows: 1500 rads to the left thorax, 1200 rads to the mediastinum, and 800 rads to a painful nodule on his left forearm. In July, 1976, he was started on courses of adriamycin, cyclophosphamide, and DTIC (dacarbazine) for widespread and enlarging metastases. In addition, a large tumor deposit on the right side of his neck, posterior to the mid-portion of the sternomastoid, was irradiated with a dose of 2500 rads.

Since 1975, he had had progressively increasing weakness in both right arm and right leg, but refused surgical therapy. He had developed atrophy and fasciculation of the right deltoid and biceps brachii muscles. On the right side, the deep tendon reflexes were increased at the triceps, but absent at the biceps brachialis. Hoffmann’s sign was positive on the right but absent on the left. In his lower limbs, bilaterally absent jerks with ankle clonus were the only abnormal findings. Sensation and bladder functions were normal.

Roentgenography in 1975 revealed an erosion of the C-5 vertebra. Myelography
FIG. 3. Case 3. Photomicrographs of tissue from the same neoplasm. Left: The intact capillary and the pleomorphism of the surrounding tumor are evident. H & E, × 400. Right: The reticulin stain shows the characteristically intact basement membrane surrounding the capillaries and a very prominent reticulin meshwork within the neoplasm reflecting the hyalinization of the intercellular matrix. Reticulin stain, × 400.

demonstrated a marked narrowing of the mid-cervical canal at the C2–6 level. By January, 1977, his neurological status had markedly deteriorated, and a myelogram showed an incomplete extradural block at the C-5 vertebra.

Operation. A cervical decompression laminectomy was performed with concurrent stabilization. At operation, an extradural tumor, 2 cm in diameter, was found adjacent to and involving the body of the C-5 vertebra. It was firm in consistency and not vascular. The resection was subtotal.

Postoperative Course. There was little improvement in the neurological status, but no further progression was noted at the last follow-up examination.

Pathological Findings

Histological examination of the original tissue (January, 1972) and lung metastases (July, 1975) revealed a spindle-celled tumor with a prominent vascular pattern (Fig. 3). The endothelial lining of the vessels had a well marked reticulin network separating it from the surrounding spindle cells. There were occasional large bizarre spindle-shaped cells with hyperchromatic nuclei. The tissue satisfied all the criteria for a diagnosis of hemangiopericytoma.

The tissue from the extradural space was primarily dense collagenous sclerotic fibrous tissue (Fig. 3 left). In the fibrous tissue there were separate pleomorphic, bizarre, and often large neoplastic cells. They were irregular in outline with vacuolated cytoplasm. Nuclei were lobulated and hyperchromatic. A rich vascular network consisting of thin-walled capillaries with intact endothelium and basement membrane coursed throughout this fibrous tissue. The reticulin framework of the basement membrane blended with the surrounding hyalinized fibrous tissue; nevertheless, the cellular component of the tumor resembled the bizarre malignant tumor cells of earlier specimens. The modification in the
Spinal hemangiopericytoma

TABLE 1
Summary of seven cases of primary spinal column hemangiopericytoma

<table>
<thead>
<tr>
<th>Authors, Year</th>
<th>Age, Sex</th>
<th>Site</th>
<th>Original Therapy</th>
<th>Time to Recurrence (yrs)</th>
<th>Treatment of Recurrence</th>
<th>Follow-Up Period</th>
</tr>
</thead>
<tbody>
<tr>
<td>Schirger, et al., 1958</td>
<td>33, F</td>
<td>T-2</td>
<td>surgery</td>
<td>26</td>
<td>surgery &amp; radiotherapy surgery</td>
<td>1 yr</td>
</tr>
<tr>
<td>Kruse, 1961</td>
<td>53, M</td>
<td>C-3</td>
<td>surgery &amp; irradiation</td>
<td>1</td>
<td>surgery</td>
<td>2 yrs</td>
</tr>
<tr>
<td>Pitlyk, et al., 1965</td>
<td>60, M</td>
<td>C-4</td>
<td>surgery</td>
<td>none</td>
<td>surgery</td>
<td>10 yrs</td>
</tr>
<tr>
<td></td>
<td>39, M</td>
<td>T-8</td>
<td>surgery</td>
<td>9, 15, 17</td>
<td>surgery (3)</td>
<td>4 mos</td>
</tr>
<tr>
<td>Harris, et al., 1978</td>
<td>28, M</td>
<td>C-3</td>
<td>surgery &amp; irradiation</td>
<td>none</td>
<td>surgery</td>
<td>5 yrs</td>
</tr>
<tr>
<td></td>
<td>65, F</td>
<td>C-5</td>
<td>surgery &amp; irradiation</td>
<td>none</td>
<td></td>
<td>4 yrs</td>
</tr>
</tbody>
</table>

Tumor cells and the sclerosis of the supporting stroma were, in all probability, the result of changes caused by the irradiation, and cytotoxic chemotherapy administered to the patient. On this basis, it was concluded that this was a deposit of metastatic hemangiopericytoma.

Discussion

The term hemangiopericytoma was coined to describe a soft-tissue neoplasm in which two distinct elements were present. One is a vascular component in which capillaries were lined by intact normal endothelial cells and were distinctly circumscribed by an intact and complete basement membrane as defined by reticulin stain. The second component is the true neoplastic element consisting of ovoid to spindle-shaped cells forming cords and sheets in between the vascular components. Ultrastructural investigation supports identification of the tumor cells as pericytic in origin with features of smooth-muscle cells. While initially the tumor was described in soft tissues, it was later found to involve all tissues including bone. In the central nervous system (CNS) hemangiopericytoma is unusual and much controversy exists in its differentiation from angioblastic meningioma. There is now good evidence that hemangiopericytoma exists as a definite entity in the CNS. These tumors, nevertheless, are particularly rare in the spinal canal, but Pitlyk, et al., suggested that 1% of meningiomas occurring in the spinal canal might be hemangiopericytomas.

Enzinger and Smith’s review indicates that hemangiopericytoma occurs principally in middle age and affects both sexes equally. While the tumor has a marked propensity to recur, the 10-year survival rate is of the order of 70%. These clinical patterns also apply to lesions in the spinal column. Five of the nine cases we reviewed occurred in the cervical spine (Tables 1 and 2).

These lesions are difficult to assess histologically. Microscopically, they can be

TABLE 2
Summary of two cases of metastatic hemangiopericytoma to the spinal column

<table>
<thead>
<tr>
<th>Authors, Year</th>
<th>Age, Sex</th>
<th>Original Tumor</th>
<th>Site</th>
<th>Therapy</th>
<th>Metastases</th>
<th>Follow-Up Period</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kruse, 1961</td>
<td>22, F</td>
<td>intracranial</td>
<td>surgery</td>
<td>L?</td>
<td>8 yrs</td>
<td>surgery</td>
</tr>
<tr>
<td>Harris, et al., 1978</td>
<td>46, M</td>
<td>thigh</td>
<td>surgery, x-ray, chemotherapy</td>
<td>C-5</td>
<td>6 yrs</td>
<td>surgery, radiotherapy</td>
</tr>
</tbody>
</table>
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categorized as either benign or malignant; however, the clinical correlation of this histological classification has generally not been good. While there is no general agreement, some authors maintain that local control of hemangiopericytoma can be obtained, and also maintained by irradiation alone.8,11 By contrast, tumor treated by surgery alone appears to have a high recurrence rate.7,12 The combination of surgery with irradiation therapy or chemotherapy has not been considered to have any greater beneficial effects.4,5,14 All three of our patients were operated on by the same surgeon. Removal of the tumors from the canal was technically difficult because of adherence to dura and irregular invasion of bone, in addition to the marked vascularity encountered in Cases 1 and 2. Having taken into account the high risk of recurrence (Table 1 and 2), adjunctive radiotherapy appeared to be of benefit in controlling the progression of the disease in our three cases.

Our cases, when added to the experience recorded in the literature, confirmed the existence of hemangiopericytoma involving the vertebral column, with extension into the spinal canal. While this remains a rare occurrence, its existence must be included in the differential diagnosis of lesions of the spinal canal. The high risk of intraoperative hemorrhage should be anticipated and potentially serious problems avoided at surgery. Its recognition will lead to improved outlook for the patient since the use of radiotherapy appeared to produce an improvement in the outlook of our cases.

References

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