Primary epidural Ewing’s sarcoma presenting as a lumbar disc protrusion

Case report

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Primary neoplasms of the spinal epidural space are uncommon. One of the rarest of these is a soft tissue sarcoma indistinguishable from Ewing’s sarcoma of the bone. Only 39 such cases have been reported previously, of which only three arose within the epidural space. The authors report an additional case, which presented in an atypical manner, and review the pertinent literature.

KEY WORDS • Ewing’s sarcoma • epidural neoplasm • lumbar disc

Tumors of the spinal epidural space are usually malignant and are commonly presumed to be metastatic. Primary neoplasms arising wholly within the spinal epidural space are considerably less common and tend to fall largely within the lymphoma-lymphosarcoma category. Other types of primary spinal epidural neoplasms are rare and the occurrence of some may be virtually unrecognized. One of these is the “extraskeletal neoplasm resembling Ewing’s sarcoma,” of which only 39 cases have been previously reported, and of which only three can truly be considered as neoplasms arising in the spinal epidural space. The obvious rarity of this tumor warrants further exposition. We are, therefore, reporting an additional case, which presented in a manner initially suggestive of a lumbar disc protrusion.

Case Report

This 19-year-old man was in good health until June, 1977, when he developed low-back pain and stiffness after lifting heavy weights. This was relieved by bedrest and mild analgesics, but recurred in mid-August, followed by the gradual onset of radicular pain radiating down the right buttock and thigh. A trial of conservative management at a local hospital was successful in alleviating the pain. Neurological examination was reportedly normal at the time.

The patient was re-evaluated in December, 1977, following the recurrence of pain, accompanied by paresthesias involving the right medial thigh and a feeling of right leg “weakness.” These symptoms were aggravated by lumbar flexion, by coughing or sneezing, and by prolonged standing or sitting, and were relieved by bedrest. Examination disclosed atrophy of the right calf, and the patient was referred to the National Naval Medical Center.

Examination. General medical examination was unremarkable. Lumbar paraspinal muscle spasm was present bilaterally, with decreased range of motion of the lower spine in all planes. Moderate weakness and marked atrophy of the right quadriceps femoris was evident, with the left measurements across comparable points at mid-thigh 6 cm greater than the right. A mild subjective sensory loss to pinprick was demonstrable along the right medial thigh. Deep-tendon reflexes were normoactive and symmetrical, except for minimal reduction of the right patellar reflex. Babinski sign was absent.

Lumbar spine radiographs obtained on admission were interpreted as normal. An electromyogram performed on December 19 disclosed evidence of a right radiculopathy at L-3. Myelography was attempted on December 21, but the procedure was terminated when a bloody tap was obtained. On the morning of December 27, the patient noted the acute onset of severe left (contralateral) lower extremity burning paresthesias. Marked weakness of the right quadriceps muscle was now evident, and straight-leg raising elicited pain bilaterally at 30° to 40°.

A lumbar puncture at the L5–S1 interspace drained only 1 to 2 cc of clear yellow fluid, which promptly clotted. A spinal needle was then introduced laterally.
Primary epidural Ewing's sarcoma

FIG. 1. Anteroposterior (left) and lateral (right) views of a lumbar Pantopaque myelogram, with the contrast medium introduced from above. Note complete obstruction to caudal flow of contrast material opposite L-2, in a feathered-edge pattern characteristic of epidural mass.

into the subarachnoid space at C1–2, where the cerebrospinal fluid was found to be clear and colorless. Pantopaque, 6 cc, was instilled, disclosing a completely obstructing extradural lesion at L-2 (Fig. 1).

Operation. Surgical decompression was performed as an emergency procedure. The laminae of L1–3 were resected, and a fleshy tumor was found at L2–3, totally confined to the epidural space. There was no evidence of extension of the tumor either through the dura or into the contiguous bone or muscle elements. The tumor was excised posteriorly and laterally, but "tailed-off" anterior to the dura, where resection could not be considered complete.

Sections disclosed a neoplasm composed of sheets of fairly uniform round-to-oval cells, with few mitotic figures (Fig. 2 upper left). The cytoplasm was relatively clear and indistinct, but contained large quantities of PAS-positive, diastase-digestible material, indicating high concentrations of glycogen (Fig. 2 upper right). There was a tendency toward organization of tumor cells into nests, separated by thin trabeculations (Fig. 2 lower).

Postoperative Course. The patient had immediate resolution of paresthesias, but was unable to void. Catheterization was required for 5 days, with subsequent restoration of voluntary control. Quadriceps strength gradually improved, and had returned to almost normal by 6 weeks after surgery.

A thorough radiographic search for an alternate primary lesion or a secondary metastasis was unrevealing. A gallium scan and technetium radionuclide scans of bone, liver, spleen, and brain disclosed no abnormalities. Intravenous pyelogram revealed prompt renal function and excretion bilaterally, without evidence of parenchymal masses or calyceal abnormalities. Serum protein electrophoresis demonstrated a normal pattern.

Radiation therapy was instituted on January 10, 1978. A total of 4500 rads was delivered to the thoracolumbar spine in fractionated doses (25 treatments of 180 rads). Chemotherapy was subsequently instituted on March 14; intravenous adriamycin, 75 mg/sq m, was given every 3 weeks, to a total dose of 550 mg.

The patient was managed on an outpatient basis after March 23, and was discharged from care on May 27, 1978. At last report, he is asymptomatic and has continued to do well.

Discussion

In 19216 and again in 1924,7 Ewing collected and reported on a series of round-cell sarcomas of the bone and delineated these as a single entity. This was even-
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Fig. 2. Photomicrographs of the tumor tissue. Upper Left: Uniform sheets of round-to-oval cells can be seen with clear cytoplasm and indistinct cell borders. Harris hematoxylin, $\times 240$. Upper Right: Note the dense content of PAS-positive (dark) material (upper half), and large numbers of PAS-positive granules (lower half). PAS, $\times 240$. Lower: Thin trabeculations of reticulin-staining elements divide groups of tumor cells into nests or clusters. There is a general absence of reticulin-staining elements between individual cells. Reticulin, $\times 240$.

tually defined as a malignant osseous tumor of children and young adults, arising usually from long bones, and composed of sheets of relatively uniform, closely packed round cells with poorly defined cytoplasm.\textsuperscript{4,11} Original concepts regarding etiology and classification have subsequently been modified, but by the 1940's this neoplasm was generally accepted as a distinct entity, together with its eponymous designation as Ewing's sarcoma of bone. It is currently regarded as a primary neoplasm of bone, probably derived from immature reticulum cells.\textsuperscript{1}

In 1975, Angervall and Enzinger\textsuperscript{1} reported on a series of 39 tumors collected at the Armed Forces Institute of Pathology (AFIP) between 1957 and 1969, which were histologically identical to Ewing's sarcoma, but distinguished by the fact that all had arisen in soft tissues. Indeed, osseous abnormalities were radiographically evident in only 10 patients, and these were clearly of a reactive nature. Twelve of the 39 patients manifested tumors that arose in proximity to the spine, only three of which were primary tumors of the epidural space. Analysis of the data regarding these 12 cases (Cases 1, 5, 10, 11, 14, 17-19, 29, 34-36) reveals an equal male:female incidence (six males, six females) with an average age at presentation of just under 20 years ($\bar{x} = 19.89$; median = 18.5; range 20 months to 41 years). The average duration of symptoms was 4½ months. The three patients who presented with a primary epidural sarcoma were two males and one female, 17, 20, and 18 years old, respectively, who had an average duration of symptoms of 2 months. Survival data are incomplete, but only one patient in the group of 12 may have been alive at the time of the AFIP report, and the longest survivor died 6½ years after surgery, chemotherapy, and two courses of radiation therapy. The three patients with primary epidural sarcoma all died within 1 year after surgery.

The histological picture characteristic of this tumor requires differentiation from other small-cell sarcomas, including reticulum-cell sarcoma and
lymphosarcoma. Other neoplasms which may be considered on the basis of histological similarity include the primitive, poorly differentiated, neuroectodermal tumors (such as neuroblastoma or neuroepithelioma), alveolar rhabdomyosarcoma, and undifferentiated metastatic carcinomas (such as bronchogenic carcinoma).

This is a group of tumors significantly different from that usually considered in an analysis of epidural mass lesions. The vast majority of epidural tumors are metastatic malignancies which include the spectrum of lymphomas and lymphosarcomas and various metastatic carcinomas, especially those originating from prostate, breast, kidney, and lung. Primary epidural neoplasms, on the other hand, are quite rare and include dermoid and epidermoid tumors, angioma, fibroma, neurofibroma, meningioma, and lipoma, a variety of tumors arising from the contiguous spinal elements (such as myeloma or osteosarcoma) and, especially, primary lymphoma. This case suggests an additional consideration in the differential diagnosis, namely, primary Ewing's sarcoma.

References


The opinions or assertions contained herein do not necessarily reflect the views of the Bureau of Medicine and Surgery of the Navy Department or the Naval Service at large.

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