Fibromatosis Involving the Epidural Space


Summary: We present a case of fibromatosis involving the epidural space that resulted in symptoms of spinal cord compression.

Index terms: Spine, neoplasms; Spinal cord, compression

Fibromatoses are benign connective tissue tumors that have been reported to occur in the extremities, chest wall, abdomen, and pelvis. This report describes a patient with epidural fibromatosis who presented with spinal cord compression.

Case Report

A 16-year-old boy was in his usual state of good health until 2 weeks before admission when a tingling sensation over both knees developed and several episodes of falling occurred because of leg weakness. He had not noted symptoms in his upper extremities but did have difficulty with urination and erections. Physical examination was significant for hyperactive reflexes in both the upper and lower extremities with bilateral plantar reflexes, a positive Romberg sign, and a sensory level at C4–5.

Findings on magnetic resonance (MR) imaging of the brain were within normal limits. MR of the cervical and thoracic spine, performed both without and after the intravenous administration of gadopentetate dimeglumine, showed a moderately inhomogeneous circumferential epidural mass extending from the level of C-2 to T-5, bilateral apical/paravertebral masses at the level of T1–2 that appeared continuous with the epidural disease, and a poorly circumscribed mass in the left side of the neck (Fig 1). A faintly enhancing, poorly marginated mass was noted in the superficial soft tissues overlaying the spinous processes from C-2 to C-5 that was not clearly continuous with the epidural disease. The subarachnoid space was preserved at all levels except at T2–3, where the mass just impinged on the spinal cord (not shown). Computed tomography (CT) showed multiple round calcifications within the epidural space and within the paravertebral masses but did not show any abnormality corresponding to the linear and curvilinear signal voids noted on MR (Fig 2).

An open biopsy of paraspinal tissue from T-2 revealed hypocellular bundles of collagen displacing and invading connective tissue and skeletal muscle (Fig 3). The final pathologic diagnosis was fibromatosis.

The patient was placed on steroids and a chemotherapeutic regimen consisting of vincristine, dactinomycin, cyclophosphamide, and tamoxifen every 3 weeks for 7 months. Follow-up scans showed modest regression of the epidural mass (Fig 4). This mild improvement in cord compression at T2–3 produced dramatic resolution of long-tract signs and return of normal motor strength and genitourinary function.

Discussion

The fibromatoses are a diverse group of tumors characterized by poorly marginated masses of highly differentiated fibroblasts intermingled with a variable amount of collagen. Their biological behavior ranges between that of fibroma and fibrosarcoma (1). They have a tendency to invade locally and may infiltrate along fascial planes, but they do not metastasize. These nonencapsulated infiltrating masses are difficult to resect, and consequently there is high rate of local recurrence, up to 50% in some series (2).

The fibromatoses arise from the connective tissue of muscle, the fasciae or aponeuroses, and have been reported in the extremities, chest wall, mediastinum, shoulders, abdomen, pelvis, buttocks, and superficial soft tissues (3). Lesions are usually solitary, but synchronous multicentric lesions can occur (4).

The appearance of fibromatosis in the epidural space on imaging studies is consistent with the few descriptions of these lesions at other anatomic sites. On CT, such lesions are most typically of the same or lower attenuation than muscle on noncontrast CT and of a higher attenuation than muscle on postcontrast CT (5,

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However, lesions that are hyperdense relative to muscle on noncontrast CT or hypodense relative to muscle on postcontrast CT have also been reported (6). These lesions may contain calcifications identifiable on CT scans or plain films (2). Most authors report that contrast enhancement improves lesion conspicuity (6); however, occasionally contrast material renders the lesions isodense with the surrounding tissue. Poor definition of the margin of these infiltrating tumors in conjunction with the minimally enhancing or isodense components has resulted in both overestimation and underestimation of tumor size (5).

Although no relationship between the various CT appearances of these lesions and their histologic features has been demonstrated (5, 6), several authors have found that the histologic features of these lesions might account for their differing appearances on MR (1, 7, 8). The lesions of fibromatoses have been reported to be isointense or hyperintense relative to skeletal muscle on T1-weighted images and to be either isointense or hypointense relative to subcutaneous fat and less commonly to be hyperintense relative to fat on T2-weighted images (1). On both T1- and T2-weighted images, almost all lesions are noted to be inhomogeneous, with linear and curvilinear areas of signal void interspersed throughout the tumor (1). Areas of higher signal on T2-weighted images corre-
spond to hypercellular areas of spindle cells, whereas markedly hypointense areas correspond to portions of the tumor that are relatively hypocellular and contain mostly collagen. In particular, linear and curvilinear areas of signal void identified on both T1- and T2-weighted images, and identified in our patient on T1-weighted images, probably reflect dense collagen and are considered a hallmark of this entity (1). These linear and curvilinear areas of signal void can be indistinguishable from vascular flow voids, but gradient-echo imaging or other flow-sensitive techniques can be used to differentiate these dense collagen bands from vessels.

Many authors describe the minimal tissue reaction evoked by fibromatosis (4) and the paucity of surrounding edema (1) as characteristic features of the fibromatoses. This lack of tissue reaction might account for the relatively moderate degree of mass effect associated with the extensive epidural infiltration in this case. The differential diagnosis of this unusual presentation includes fibrosarcoma, neurofibroma, lymphoma, and granulomatous inflammation. All of these lesions would be likely to show more mass effect. In particular, fibrosarcoma, an entity that might share some histologic and imaging features with the fibromatoses, would typically elicit more tissue reaction (5). Neurofibromatosis would also show more mass effect, and would certainly involve rather than surround the foraminal nerve roots (Fig 2A), as well as possibly widening the neural foramina. Epidural lymphoma tends to create mass effect, and untreated lymphoma does not contain calcification.

MR and CT can provide complementary information, as occurred in our case. The multiplanar capability of MR provides better delineation of the multiple tumors and their relationship to the spinal cord. Most significantly, MR shows a pattern of signal intensity that might enable a preoperative diagnosis. CT may better show lesions surrounded by fat, such as the superficial lesion that was the ultimate site of biopsy. On contrast-enhanced MR, enhancing tumor can be difficult to differentiate from the hyperintense signal of surrounding fat unless fat suppression is available.

Fig 3. Histologic findings of fibromatosis (trichrome stain). Abnormally thick and hypocellular collagenous bundles replace paraspinal connective tissue. At least 95% of this tumor comprised fibrillar and amorphous extracellular matrix material. Although slightly more cellular at their presumed leading edges, the largest bundles were hyalinized and nearly acellular and often exceeded 1 mm in width. The ratio of cellular to extracellular material was small, even in relatively cellular bundles. Scale bar indicates 250 μm.

Fig 4. Sagittal T1-weighted postcontrast MR image (500/30/2) after 5 months of chemotherapy shows a very modest reduction in the size of the epidural mass at the level of C-5.
References