Atypical fibrous histiocytoma of the thoracic spine

Case report

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A 14-year-old boy presented with mid-thoracic pain, leg weakness, and bladder dysfunction. Plain x-ray films and bone scan were normal whereas myelography demonstrated an extradural mass extending from T-2 to T-5. Exploration revealed a large tumor adherent to dura and bone. Pathological verification of an atypical fibrous histiocytoma prompted the report of this rare tumor of the vertebral column.

Key Words • histiocytoma • paraplegia • myelography • thoracic spine

A typical fibrous histiocytoma is a common tumor of soft tissue. Although this tumor may be found in bone, involvement of the thoracic vertebral column has not been previously reported. A case of a teen-age boy with a rapid, progressive T-5 myelopathy is presented. Diagnosis was made more difficult due to the history of trauma.

Case Report

This 14-year-old boy presented in August, 1978, complaining of pain in the thoracic spine. Five months before admission, the patient was kicked forcefully over the thoracic spine while playing basketball. The pain persisted and for 1 month before admission he noticed increasing leg weakness and difficulty in voiding.

Examination. The patient had restricted lumbar flexion, marked tenderness over the T-5 spinous process, and paravertebral muscle spasm. There was severe bilateral leg weakness with greater involvement of the distal rather than proximal muscles. Bilateral ankle clonus, hyperreflexia, and extensor toe responses were noted. A T-5 sensory level was present.

Radiological studies included normal plain films, bone scan, and anteroposterior tomograms of the thoracic spine. Myelography revealed a total extradural block at T-5. Lateral C1-2 puncture was required for visualization of the T-2 block. Cerebrospinal fluid studies revealed crystal-clear fluid with an elevated protein content of 89 mg%, and gamma-globulin of 24.8 mg%. Alkaline phosphatase was increased to 972 U/hr. Complete blood count, sedimentation rate, and sickle-cell preparations were normal.

Operation. At surgery a three-level (T2-5) decompressive laminectomy was performed, allowing exposure and gross radical removal of the tumor. The extradural location of the tumor was not associated with soft tissue or paraspinous muscle extension. The nature of the tumor required a piecemeal removal, the largest fragment measuring 2.5 x 1.5 x 1 cm.

Histological Examination. The tissue revealed a cellular, neoplastic lesion with hyperchromatic and enlarged cells. The nuclei were oval and there was a moderate amount of cytoplasm. There were scattered multinucleated cells with areas rich in both chondroid and osteoid matrices. Mitotic activity was extremely infrequent (Fig. 1).

Postoperative Course. Two weeks after surgery, the patient regained considerable strength in his legs, could walk without aid, and was engaged in an active rehabilitation program. Postoperative myelography showed a normal flow of Pantopaque through the area of previous obstruction. Radiation and chemotherapy were withheld pending further clinical and myelographic follow-up studies. Re-examination at 6 months showed continued clinical improvement.
FIG. 1. Photomicrograph of the tumor. Both mono- and multinucleated, slightly atypical histiocytic cells are found within a loose, amorphous matrix. Although nucleoli are prominent, the chromatin pattern is delicate. H & E, × 450.

Discussion

Malignant fibrous histiocytoma (of soft tissue) was first described by O'Brien and Stout in 1964.9 The histiocyte is believed to be the cell of origin, giving rise to a heterogeneous group of tumors.1,6,9,14,16 Typically, the characteristics include a storiform pattern, histiocyte spindle-like cells (facultative fibroblasts), indented nuclei, abundant foamy cytoplasm, large nucleoli, and multinucleated giant cells.2,4,7,8,13,14 Of this fibrohistiocytic group, the atypical fibrous histiocytoma can be differentiated from the other three histological types: malignant fibrous histiocytoma, malignant histiocytoma, and epithelioid sarcoma. This lesion also may lack the classic storiform pattern.15 Differential diagnosis includes osteogenic sarcoma, fibrosarcoma, and chondrosarcoma. Spanier, et al.,13,15 suggest that any malignant tumor in which osteoid changes can unequivocally be demonstrated as produced by tumor cells should be classified as osteogenic sarcoma. In Weiss and Enzinger's analysis of 200 cases of malignant fibrous histiocytoma, metaplastic osteoid or chondroid tissue was present in some tumors.10 The diagnosis of atypical fibrous histiocytoma is appropriate when multiple samples of the entire specimen fit the above histological pattern.2,7 Five- and 10-year survival rates are reported as 90% and 80%, respectively, for soft-tissue tumors.19 Although cases of pelvic and sacral involvement have been identified, there are no reported instances of this tumor arising from the thoracic spine with cord compression.2,4,7 Despite radical removal of the tumor, confirmed by normal postoperative myelography, the locally aggressive nature of this lesion makes long-term prognosis guarded.

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References


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