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Case Report

Primary pleomorphic liposarcoma of the thoracic epidural space: case report

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Abstract

BACKGROUND CONTEXT: Pleomorphic liposarcoma (PLS) is a rare malignant soft tissue tumor comprising 5%–15% of liposarcomas and characterized by high malignant potential. To our knowledge only three cases of this entity have been reported in the spine.
PURPOSE: We describe the only reported case of a purely epidural PLS with no macroscopic bone involvement at diagnosis.
STUDY DESIGN/SETTING: A case presenting clinical evidence that PLS may arise from the epidural fat is reported.
METHODS: The clinical presentation, management, and outcome in a case of primary PLS of the thoracic spine, and a review of the literature, are presented.
RESULTS: A 70-year-male presented with sudden onset lower extremity weakness, constipation, and back pain. Magnetic resonance imaging revealed an epidural lesion at T5 with noted mass effect compressing the spinal cord and extension to the T5–T6 foramen. Urgent decompressive laminectomy with gross total resection was performed. Histopathology revealed high-grade PLS. Adjunct radiotherapy was prescribed. The tumor recurred 3 months later. In spite of repeat surgery, additional radiation, and chemotherapy, the patient developed widespread metastases and succumbed to his disease 1 year after treatment began.
CONCLUSIONS: Spinal PLS is a rare entity, but nonetheless may arise from epidural fat and should be considered in the differential diagnosis of primary spinal cord lesions. © 2015 Elsevier Inc. All rights reserved.

Keywords: Bone invasion; Epidural fat; Pleomorphic liposarcoma; Primary spinal tumor; Spinal cord compression; Spinal liposarcoma

Introduction

Liposarcoma is the most common soft tissue sarcoma in adults, accounting for 20% of all mesenchymal malignancies. The World Health Organization classifies malignant liposarcomas into four histologic subtypes: myxoid, mixed, pleomorphic, and dedifferentiated [1]. Pleomorphic liposarcoma (PLS) is a high-grade sarcoma that most commonly presents in the thigh and pelvis of older adults; 25% of cases present with metastatic disease, most commonly to the lungs, liver, and skeleton [2]. It is the rarest of the liposarcomas, with only 5%–15% meeting diagnostic criteria for this subgroup [3–5]. In patients with localized disease at diagnosis, there is a 5-year survival rate of approximately 60% [3,4,6–8]. Recurrent disease, no surgical resection, and positive resection margins are independent poor prognostic indicators [2].

Primary PLS of the spine is a particularly rare entity, with only three cases reported [7–9]. In all of the cases, the tumor presented with a significant involvement of the vertebral body

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at diagnosis. We present a case of a primary PLS in the thoracic spine that was purely epidural, extending for the full height of the T5 vertebral body at the right foramen. Bony involvement was not apparent on preoperative imaging or during the initial surgery. When the tumor recurred, involvement of the vertebral body was evident on magnetic resonance imaging (MRI), and microscopic bone invasion was found on histopathologic examination.

Case report

A 70-year-old man presented to the Emergency Department complaining of constipation. The patient was sent home after symptomatic treatment, only to return after 3 days with complaints of continued constipation, back pain, and sudden onset of progressive bilateral lower extremity weakness. Neurologic examination on admission was significant for +4/5 motor strength in the bilateral lower extremities, bilateral Babinski, and sensory level cut at T10 (American Spinal Injury Association [ASIA] Impairment Scale [AIS] D [10]). The patient underwent emergency MRI of the thoracic and lumbar spine, which revealed a hypointense solid, cystic, epidural nonenhancing mass. The mass was located in the right lateral aspect of the spinal canal at T5, spanning the discs at T4–T5 and T5–T6 with noted extension into the right foramen at T5–T6. No bone involvement was evident. The tumor measured approximately 2.5 cm×1 cm×1 cm, creating a prominent mass effect with marked cord compression (Fig. 1).

First surgery

The patient underwent emergency T4–T5 laminectomy. At surgery, the lesion was a white caseating mass in the spinal canal, emanating from the epidural space and expanding into the right T5–T6 foramen. No macroscopic bone involvement was observed. Gross total resection was achieved via piecemeal excision followed by T5–T6 foraminotomy. The patient tolerated the procedure well with no perioperative complications.

Pathologic diagnosis and clinical course

Histopathology revealed high-grade PLS (Fig. 2).

The patient's postoperative course was uneventful, and he made a complete neurologic recovery (ASIA E). MRI performed 1 month later showed no evidence of residual tumor. Positron emission tomography-computed tomography performed in the same period showed an area of increased uptake in the T4–T6 right paravertebral area with no evidence of other primary or systemic disease (Fig. 3).

The patient was referred for adjuvant external beam radiation therapy (EBRT); however, he was reluctant to begin treatment. He returned to the Emergency Department 3 months after his first surgery, after only one treatment, complaining of progressive lower limb weakness of 3 days' duration, difficulty initiating urination, numbness below the nipple line, and abdominal distention. On examination, the patient had

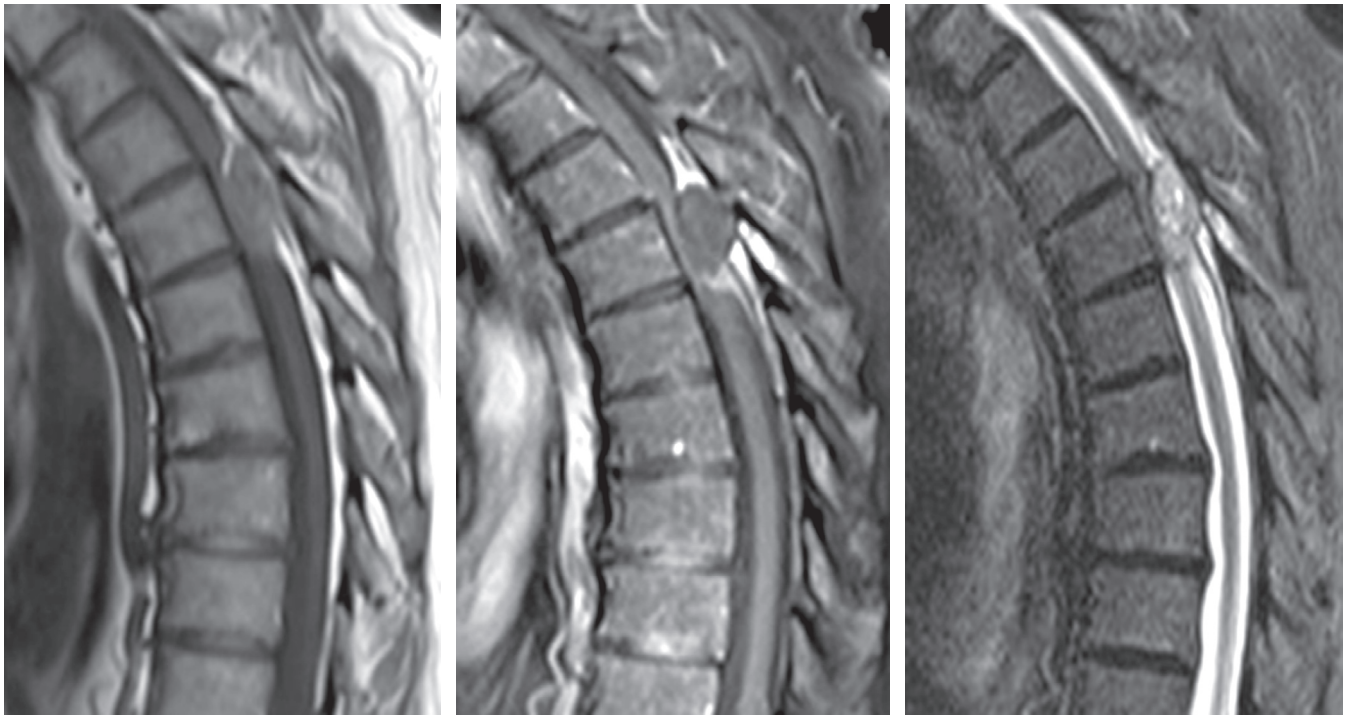

Fig. 1. Sagittal MRI in a 70-year-old man showing a posterior non-enhancing solid, cystic epidural tumor with cord compression spanning the discs at T4–T5 and T5–T6. (Left) T1-weighted; (Middle) Fat-suppressed, gadolinium-enhanced; and (Right) T2-weighted studies.

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