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Spondylectomy of Epithelioid Hemangio-endothelioma of Thoracic Spine and Technical Resection Issues

(Case Report)

Abstract

Background Primary Epithelioid Hemangioendothelioma (EHE) of spine is an extremely rare malignant vascular neoplasm with unpredictable outcome. As a malignant tumor, total resection offers the best possible disease - free survival, while the location poses difficult technical problems .

Method: A 38-year-old man with a large tumor of T10 vertebra was investigated, had biopsy-confirmed diagnosis of EHE, and underwent simultaneous anterior and posterior resection and reconstruction with anterior strut grafting and posteriorly instrumented fusion.

Results: The patient has had good a post-operative course with no neurological damage and no recurrence found in the one-year follow-up .

Conclusion: EHE of thoracic spine is a rare finding and a well-planned total surgical excision can produce a good outcome.

Keywords: Hemangioendothelioma, Thoracic Vertebrae, Vascular Neoplasms, Spine, Spinal fusion

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Introduction

Primary Epithelioid Hemangioendothelioma (EHE) of spine is an angiocentric vascular tumor, representing only 1% of all malignant tumors[1]. These tumors can be asymptomatic. Local pain is the most common presentation of spinal EHE. PET/CT imaging is essential for diagnosis and follow-up of the tumor[1,2]. Angiosarcoma is defined as a high-grade malignant tumor Enneking stage IIB, extra-compartmental. The term EHE, low grade angiosarcoma, is identified as Enneking stage III, benign aggressive extra-compartmental tumor, has metastatic potential[3,4,5,6]. We are reporting a case of EHE involving T10 vertebra, for whom T10 spondylectomy was performed

Case presentation

A 38-year-old man, with a 4-months history of back pain and bilateral leg pain (visual analogue scale 6) was referred to our Spine unit at Shiraz University, Iran. During this period, he has been receiving symptomatic treatment. On physical examination, no spinal mass or skin change was detected in the thoracic region. The patient was ambulatory without support and spine movements were normal. He endorsed mild pain on forward flexion. Neurologic evaluation showed intact strength and sensory exams.

A spine x-ray radiograph showed well preserved vertebral height, normal thoracic kyphosis, with an ill-defined lytic lesion. Spine MRI showed an enhancing mass replacing most of T10 vertebra (Figure 1). The patient underwent a CT-guided biopsy that showed typical vascular channels with fibrous stroma and epithelioid cells with eosinophilic cytoplasm. The tumor location, in accordance with the Weinstein Boriani Biagini (WBB) location classification ,was unilateral zone 3-7 anterior/ posterior (Figure 2).

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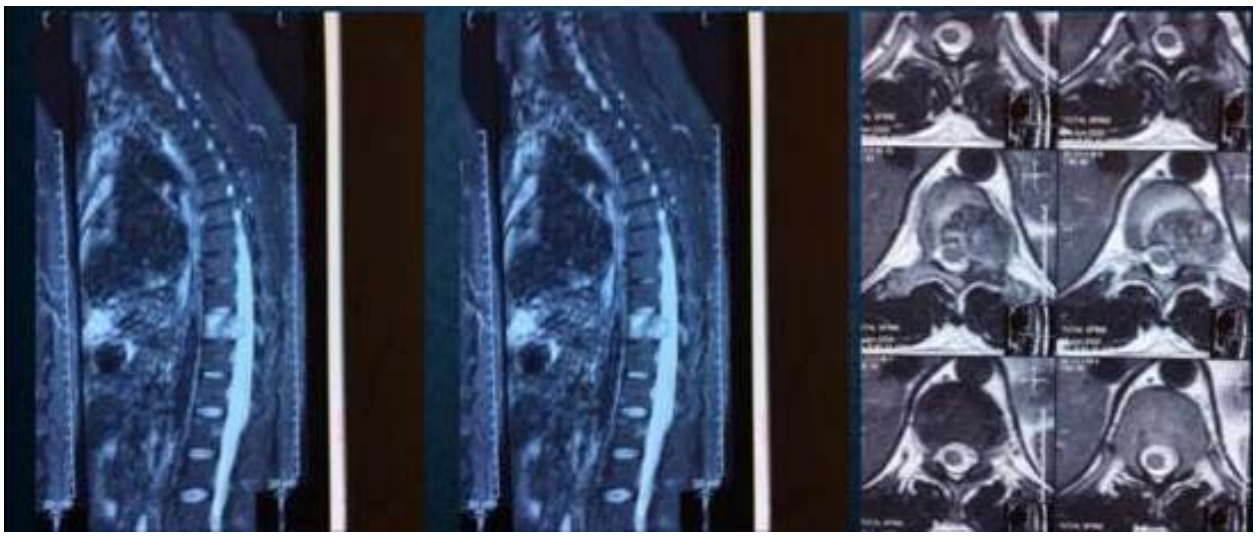

Figure 1: MRI shows heterogenous tumor mass involving T10 vertebrae with extension of tumor into the spi nal canal.

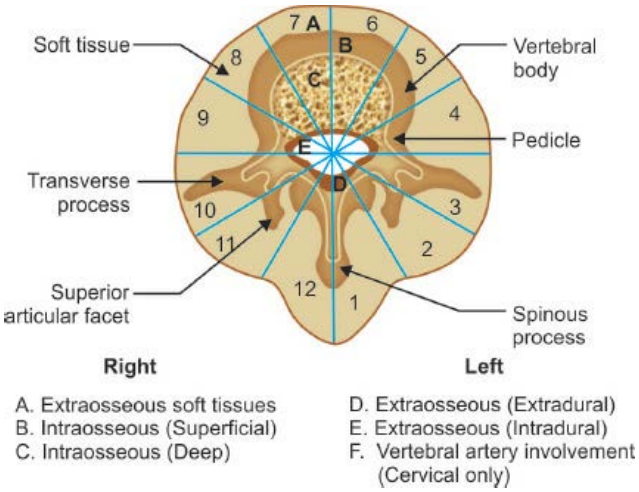

Figure 2: Weinstein Boriani Biagini staging system showing zones of tumor involvement [10].

Operative Technique

Based on the diagnosis of EHE and considering the WBB classification, surgical intervention was planned. T10 spondylectomy was completed via posterior approach on prone position. The T7 to L1 vertebrae were exposed posterolaterally. Pedicular screw fixation from T7 to L1 was performed. At the pathologic level, dissection was completed by exposing the lamina, facet joints, and ribs bilaterally. T10 laminectomy and facetectomy was completed. At the T10 level, spinal dura was released, and nerve roots were bilaterally ligated. After rib resection (4cm) on each side, the chest wall was opened. Discs at T9-T10 and T10-T11 were defined anteriorly after releasing the vascular tissue (aorta and vena cava).

After placing a temporary rod from T9 to T11, T10 vertebra was resected anteriorly through the disc space using Gigli saw .Next, the pathologic vertebra was mobilized and removed posteriorly. Anterior reconstruction with cage and bilateral fibular grafting was performed. Then, the temporary rod was removed and replaced by permanent rods. Following completion of the anterior and posterior reconstruction, intraoperative wake up test was successfully performed and intact nerve function was confirmed. Bilateral chest tubes were inserted posteriorly. The estimated blood loss was 2500 mL with 4 hours of operating time.

The patient started mobilizing after 48 hours with the physiotherapist. There was no neurologic deficit and chest tubes were removed 72 hours post-op without any pulmonary complications. The hospital course was uneventful, and patient was discharged 7 days post-op. In the one-year follow-up ,the patient is asymptomatic clinically and radiographically.

Discussion

EHE is a rare benign aggressive vascular tumor with backpain as the main presenting symptom. Presence of any neurologic symptoms depend on the location of tumor[2]. Histologic features of EHE range between hemangioma and high-grade sarcoma.

Microscopically, the tumor is composed of anastomosing cords, solid nests, and round eosinophilic epithelial cells. The lesion does not display well-formed vascular structures such as those of hemangioma. It also lacks the cytologic atypia seen in angiosarcoma[4,7].

X-ray findings of EHE are not specific and it can appear as an osteolytic lesion. "Soap bubble" appearance with bone expansion has been described in some of the cases[7,8]. CT imaging can be used to evaluate the degree of bone destruction. MRI findings are also non-specific with decreased signal intensity on T1 and slight increase in signal intensity on T2-weighted images[7] EHE may occur at any age with predilection between 20-30 years and male to female ratio of 2:1[3,4]. The overall survival for the patient with a. unicentric tumor has been reported as 89%

Figure 3: Post-operative resection and reconstruction with bilateral strut fibular grafting.

compared with 50% in patient with multifocal disease. There are no standardized treatment guidelines because of rarity of the spinal EHE and its unpredictable natural course[3,4].

Treatment approaches for spinal EHE include surgical treatment, radiotherapy and chemotherapy[3,4]. A patient with a wide or marginal resection has better prognosis[4]. Partial corpectomy and vertebral reconstruction with bone grafting has been suggested as an alternative approach[9]. Radiotherapy is recommended to reduce risk of local recurrence after surgery. Our patient after one-year follow- up has been in stable condition, pain free with no signs of recurrence on reconstructed CT scan.

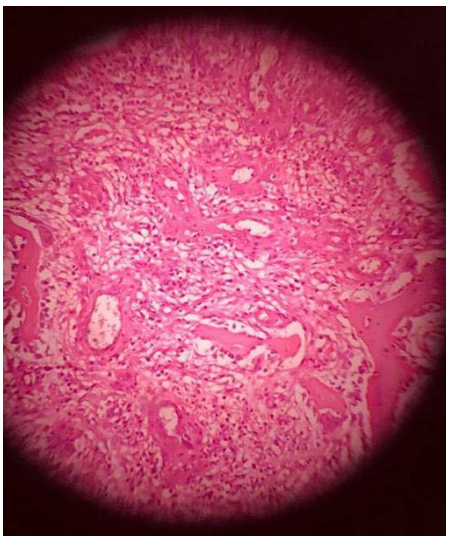

Figure 4: EHE composed of strands, cords of epithelioid cells with eosinophilic cytoplasm and vacuoles embedded in a myxohyaline stroma.

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