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Multicentric epithelioid hemangioendothelioma of bone. Report of a case with radiologic-pathologic correlation

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Abstract Epithelioid hemangioendothelioma (EHE) is a rare vascular tumor of uncertain biologic behavior. Most cases come out as a single lesion of the soft tissue but also may appear in the lung, liver, and other locations. Epithelioid hemangioendothelioma of bone is an extremely rare tumor and more prevalent in the second and third decades of life; its behavior is uncertain, it most commonly is unifocal, and it affects preferentially lower extremities. In this work, we present the clinical, radiologic, and pathologic findings of a 19-year-old man with a multicentric EHE of bone that involved 3 vertebrae and developed lung metastasis.

Keywords: Epithelioid hemangioendothelioma; Vascular bone tumors; Multicentric bone tumors

1. Introduction

Epithelioid hemangioendothelioma (EHE) is a rare vascular tumor of uncertain biologic behavior; it generally arises in soft tissue as a single lesion, but it has been described in other sites such as lung and liver [1-3]. Epithelioid hemangioendothelioma primary in bone is an unusual neoplasm, it occurs in the second and third decades, and chiefly affects lower limbs, but it may involve any bone [4-6]. Multicentric EHE of bone is a rare occurrence and signifies a challenge for the radiologic diagnosis because it may be confused with other neoplastic and nonneoplastic conditions [7].

In this article, we present the clinical, radiologic, and pathologic findings of a 19-year-old man with a multicentric EHE that involved simultaneously 3 vertebrae and developed pulmonary metastases.

2. Clinical history

A 19-year-old man, after a falling traumatism, complained of pain in the thoracic spine with neurologic symptoms of paresthesia and loss of strength of the lower limbs. Computed axial tomography (CAT) scan and magnetic resonance imaging done in another hospital disclosed a pathologic fracture in 3 vertebral bodies (T1, T3, and T4) interpreted as "suggestive of an infectious process." Over the span of 3 months, the patient evolved with slow and progressive loss of muscular strength and, eventually, paraplegia. A biopsy from paravertebral tissue was obtained, bacteriologic cultures from this material resulted negative, and the pathologic diagnosis was "proliferation of epithelioid vessels most consistent with EHE." A chest x-ray and a CAT disclosed a left pleural effusion and a diffuse bilateral interstitial infiltrate. A thoracoscopy of lung and pleura revealed a serohematic pleural effusion of 500 mL, a 1-cm nodule in parietal pleura and multiple intrapulmonary nodules, and biopsy of lung and pleura; a biopsy was obtained, and the histopathologic diagnosis was metastatic EHE in lung and pleura. Palliative radiotherapy to the spine was administered. The patient's condition progressively worsened: in 3 months, he developed bilateral pulmonary nodules and right pleural effusion and orthopnea requiring mechanical assistance. He was discharged from the hospital and received palliative care until death 5 months after admission.

3. Image findings

Magnetic resonance image in T2 sequence shows pathologic fractures in dorsal vertebrae 1 to 4 (Fig. 1A);

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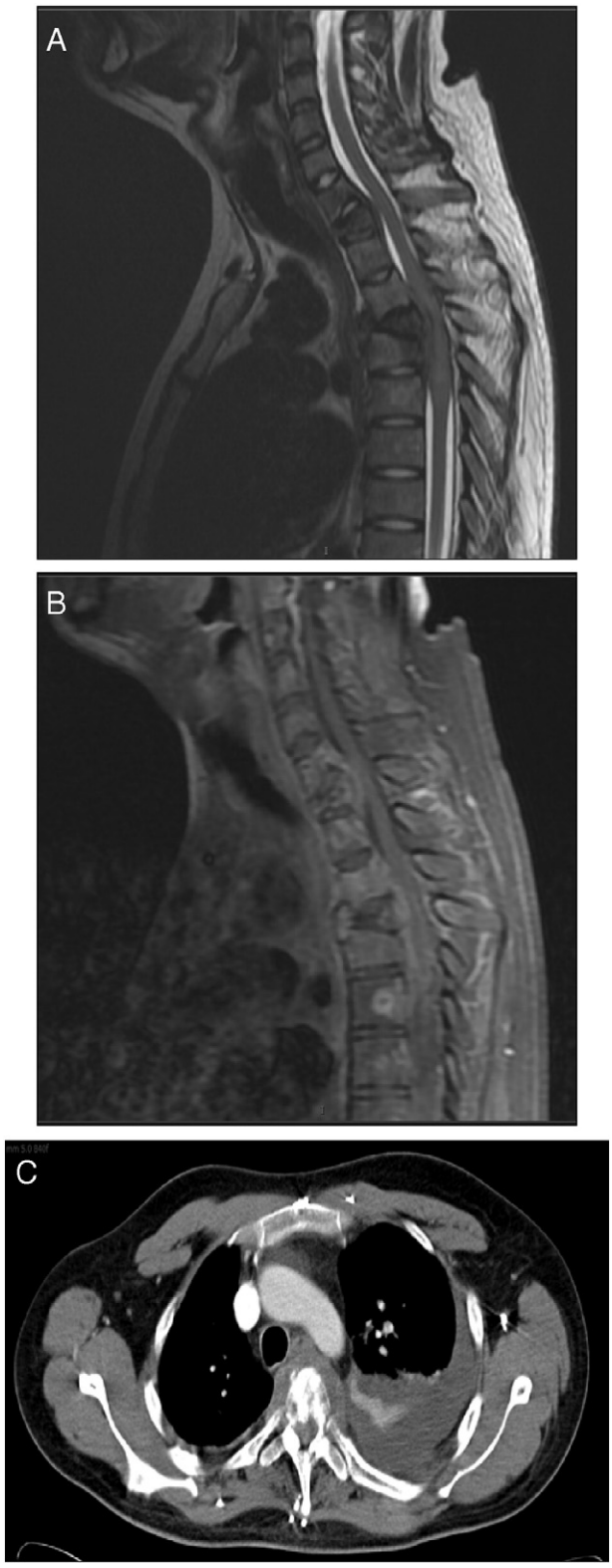

Fig. 1. Magnetic resonance (T2) image shows pathologic fractures in vertebrae D1, D3, and D4 (A); the image is enhanced with gadolinium in T1 (B). Computed tomographic scan showing a pathologic fracture in D4 vertebra (C).

there is alteration in the contour of the vertebral bodies with decrease in height and displacement of the posterior wall of D4, which compresses and narrows the spinal cord. After intravenous contrast (Fig. 1B), there was an enhanced signal in the vertebral bodies described above and also in C2, C5, C6, D6, D7, and D8.

A computed tomographic scan (Fig. 1C) demonstrated a pathologic fracture in T4 vertebra with collapse of the vertebral body. The radiologic findings were suggestive of a vascular tumor, but the possibilities of metastases and infectious process could not be ruled out.

4. Pathologic findings

Histologic study of the vertebra showed neoplastic tissue formed by epithelioid cells arranged in cords and nests surrounded by abundant myxoid stroma with hyaline areas (Fig. 2A). The tumor cells were large, polygonal, with well-defined borders and ample eosinophylic cytoplasm, and with intracellular lumina containing erythrocytes in some cells. The nuclei were central, ovoid, and with little variation in size and shape (Fig. 2B). The pulmonary and pleural nodules there were also formed by neoplastic tissue displaying similar histologic characteristics but with more abundant myxoid stroma (Fig. 2C).

4.1. Immunohistochemistry

The neoplastic cells of the vertebral were positive for CD34 (Fig. 2D), CD31 (Fig. 2E), and epithelial membrane antigen (EMA) (Fig. 2F). In the lung and pleural tumors, the neoplastic cells were positive only for CD34 and negative for cytokeratin cocktail (CK) and EMA (not shown).

5. Discussion

Epithelioid hemangioendothelioma is a rare tumor of vascular origin and uncertain malignant potential. Epithelioid hemangioendothelioma of the soft tissue was described in 1982 by Weiss and Enzinger [8], and it has been described with lesser frequency in visceral organs and other sites [1-3]. Epithelioid hemangioendothelioma of bone is rare [4-6,9,10], it represents 0.5% to 1.0% of the primary malignant tumors [11], its age range is from 20 to 30 years, and both sexes are equally affected.

Osseous EHE is sometimes seen as a multifocal or multicentric disease [7,12]; it is located mainly in the long tubular bones of lower extremities and in the pelvis, affects upper extremities and other flat bones less frequently, and rarely arises in vertebrae as in the present case.

All the histologic characteristics described in EHE were present in our case: the cells were polygonal, arranged in trabeculae and cords surrounded by myxoid stroma, and displayed ample cytoplasm with intracellular lumina containing erythrocytes.