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Medical imaging quiz – Case 79

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ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2025, 42(1):135–136

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A 35-year-old male patient presented with waist worsening pain. He referred having recurrent episodes of waist pain the last two years. He did not have any underlying disease or history of cancer. Laboratory evaluation revealed none pathological finding; thus, a magnetic resonance (MR) imaging was performed, which revealed the underlying pathology. Diagnosis was confirmed after a computed tomography (CT)-guided percutaneous fine needle biopsy (fig. 1).

Comments

Epithelioid hemangioendothelioma (EHE) is a rare relatively low grade vascular tumor first described in 1982 by Weiss and Enzinger. It occurs around medium to large venous structures. Liver and lung are the most frequently involved sites, but epithelioid hemangioma (EH) can be found in soft tissues and bones. The peak incidence occurs in the second and the third decades, but EH can occur at almost any age. Osseous EH has a predilection for occurrence in males (male:female ratio is 2:1). Osseous EH involves skull, axial skeleton, and the lower extremities in most cases. The most frequently involved long bones are the tibia (23%), femur (18%), and humerus (13%). In over 50% of cases, lesions are multifocal, especially with involvement of the lower extremity. Multifocal lesions have a tendency to involve bones of the same region. Synchronous involvement of contiguous bones is common, such as the tibia and fibula, but in some cases, separated tumor foci are present in distant bones.

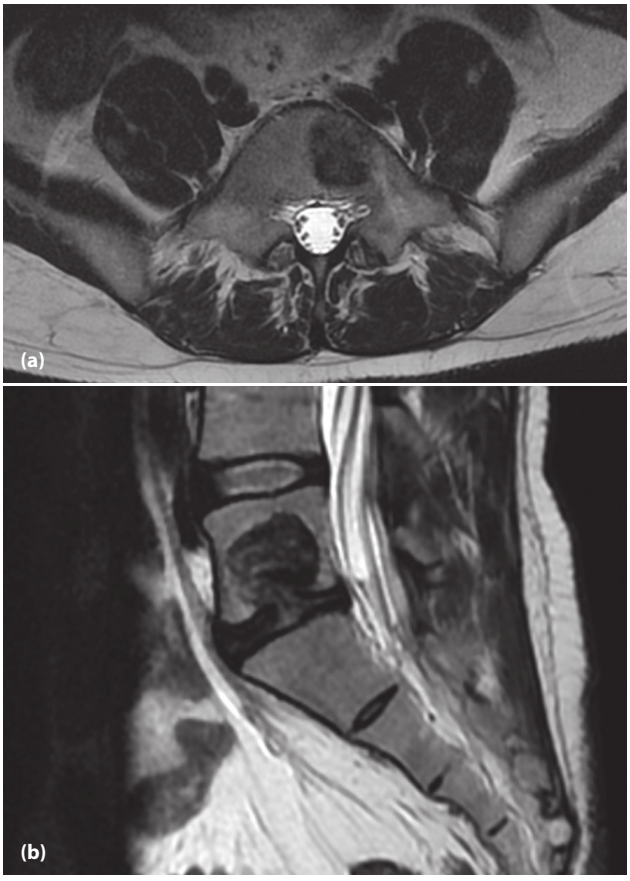

Figure 1. Magnetic resonance (MR) images of lumbar spine. (a) On a transverse T2-weighted image, atypical lesion with intermediate signal intensity. (b) On a sagittal image, the lesion appears in the body of L5 vertebra, with focal intermediate signal intensity foci.

Clinically, local pain and swelling that last weeks to years are the main symptoms. A pathologic fracture can be associated in 10% of patients. Constitutional symptoms, such as hemolytic anemia and consumption coagulopathy are rarely reported. Radiologically, EH presents as osteolysis without mineralization. Calcifications and periosteal reaction are rare. Lesions may be well defined or poorly demarcated. Expansile remodeling, cortical disruption and soft tissue extension can be seen. In CT scans, a soft tissue mass shows isoattenuation to muscle with homogeneous enhancement on a contrast enhanced CT scan. The signal intensity on MR imaging is not specific. EH has been reported to show low to intermediate signal intensity on T1-weighted image and high signal intensity on T2-weighted images. After injecting gadolinium contrast agent, the mass has been reported to show homogeneous enhancement. The presence of flow voids, suggestive of vascular channels, may represent a neoplasm of vascular origin, but this finding does not indicate hemangioendothelioma, but rather should suggest other diagnosis such as hemangiopericytoma. The radiologic differential diagnoses include a simple bone cyst, fibrous dysplasia with cystic

degeneration, or cystic angiomatosis.

The gross features of EH is a bright red hemorrhagic tumor with irregular scalloped borders. Microscopically, the EH usually consists of an anastomosing cord of epithelioid cells that occasionally form poorly defined vascular channels. Cells are plump with abundant, granular, eosinophilic cytoplasm. The vacuolization of cytoplasm is characteristic, presenting attempts to form a primitive vascular lumen. The nuclei are round with prominent nucleoli. The individual epitheliod cell cytoplasmic lumen may contain red blood cells. Mitotic activity is usually low with 1 to 2 mitoses per 10 high power fields. Epithelioid cells express endothelial markers such as factor VIII-related antigen, CD31 and CD34. The clinical course and prognosis of EH is somewhere between that of hemangioma and angiosarcoma. Most cases show locally destructive indolent behavior, but the prognosis of EH of bone is hardly predictable because prognosis cannot be made on the basis of histologic grade alone. Visceral involvement seems to be the most important indicating poor prognosis. There is no established standard treatment of EH of bone. The number, size, location, and presence of metastasis determine the treatment. For localized disease, wide resection followed by radiation therapy is recommended. Radiation therapy or chemotherapy has been used for multicentric disease.

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