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CASE REPORT

A Rare Soft Tissue Tumor Confused Radiologically with Chondroma; Extraskeletal Osteosarcoma

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Abstract

Extraskeletal osteosarcoma is a malignant chondro-osseous neoplasia that is not associated with the skeletal system. It accounts for less than 1% of all soft tissue sarcomas. We report a case of a 60-year-old male with an isolated primary subcutaneous tumor in the dorsal aspect of his left foot. The lesion was initially considered an extraskeletal chondroma radiologically. After imaging; surgical excision and pathological analysis the diagnosis of extraskeletal osteosarcoma was made. This report includes the clinical and pathological findings of extraskeletal osteosarcoma in this case.

Keywords: Extraskeletal osteosarcoma- subcutaneous adipose tissue- chondroma- soft-tissue tumor- chondro-osseous tumor

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Introduction

Extraskeletal osteosarcoma (ESOS) is a malignant chondro-osseous neoplasia that is not associated with the skeletal system. It is characterized by bone matrix or osteoid formation. It may contain osteoblastic component as well as chondroblastic and fibroblastic components. Although extraskeletal osteosarcoma is histologically similar to primary osteosarcoma of bone; there are significant differences between them in terms of demographic characteristics, imaging characteristics, and prognosis [1].

It was first described in 1941 [2]. Although most of these tumors are located in the deep soft tissue, they can sometimes be localized in the dermis or subcutaneous tissue. When the literature is examined, it has been seen that there are ESOS cases reported in many different localizations such as vesicle seminalis, lymph node, mediastinum, and orbit [3-9].

Case Report

A 60-year-old male patient presented to our hospital due to swelling in the dorsal aspect of his left foot. The patient had a history of diabetes and hypertension. Contrast-enhanced magnetic resonance imaging revealed a 36x21 mm sized, largely calcified, well-circumscribed lesion with peripheral enhancement in post-contrast series in the subcutaneous fat tissue in the dorsal part of the foot at the level of the 2nd metatarsal bone. The described lesion did not appear to invade the extensor tendon or bone structure. The lesion was initially considered an extraskeletal chondroma radiologically (Figure 1A). A piece of nodular tissue, 3.8x3.5x1.8 cm, gray-white in color, and bone-hard, was sent to our department. Sampling was done after decalcification. Spindle and polygonal shaped bizarre tumor cells forming widespread neoplastic bone matrix were observed in tissue samples. In addition to osteoblastic differentiation, areas of chondroblastic and fibroblastic differentiation were also present in the tumor (Figure 1B-1D). As a result of the large immunohistochemical panel applied, tumor cells were positive with CD99, bcl-2; focal positive with SMA (smooth muscle actin), desmin, S100, p63; EMA (epithelial membrane antigen) showed negative immune reaction with panCK, CD34. Aberrant staining was observed with p53. 71 mitoses were counted in 10 high-magnification fields. Ki-67 proliferation index was 80%. Malignant neoplasms such as extraskeletal osteosarcoma, mesenchymal chondrosarcoma, synovial sarcoma, and undifferentiated pleomorphic sarcoma

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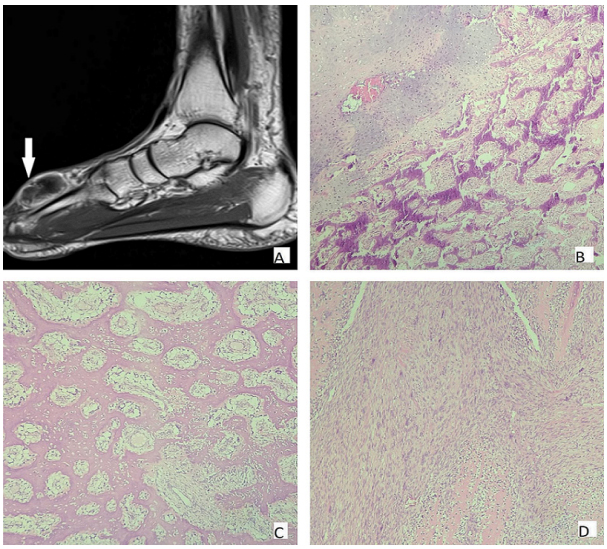

Figure 1. Extraskeletal Osteosarcoma. Figure 1A, MRI showing a well-circumscribed lesion in the dorsal part of the foot. Figure 1B, 1C, Low-magnification (×100) photomicrograph with hematoxylin-eosin stain demonstrating chondroid matrix and dense sheets of osteoid. Figure 1D, Low-magnification (×100) photomicrograph with hematoxylin-eosin stain demonstrating fibroblastic differentiation with severe cytological atypia.

were included in the differential diagnosis. As the mass was extraskeletal and showed extensive osteoblastic differentiation, the case was found to be compatible with ESOS. Additional resection was performed because of the positive surgical margins. Adjuvant radiochemotherapy was given after surgery.

Discussion

ESOS accounts for less than 1% of all soft tissue sarcomas and occurs mainly in the middle-aged. Most cases develop de novo, in some cases, radiation exposure is a predisposition factor [10]. Due to their rarity, the diagnosis and treatment of ESOS remain controversial.

On radiographs, extraskeletal osteosarcoma is often seen as a soft tissue mass with variable amounts of mineralization. Osteoid matrix formation or calcification is seen in approximately half of the lesions. An increase can be observed over time. Matrix mineralization is best evaluated on computed tomography (CT) rather than radiography or magnetic resonance imaging (MRI). On MRI, the tumor is usually a well-circumscribed heterogeneous mass and isointense to skeletal muscle on T1- and T2-weighted images [1].

ESOS is the soft tissue equivalent of osteosarcoma in bone. Morphologically, it can appear in many different forms. Tumors usually consist of epitheloid, spindle, or polygonal cells with pleo-morphism and cytological atypia. Osteoblastic, chondroblastic, and fibroblastic differentiation areas can be seen at different rates [10]. The neoplastic bone may grow in lace-like, trabecular, or sheet-like patterns. Tumor necrosis is common. The Special AT-rich sequence-binding protein 2 (SATB2) immunohistochemical stain can be used to detect osteoblastic differentiation [11]. But not specific for osteosarcoma. Osteoid production can also be seen in some non-ESOS high-grade soft tissue sarcomas like dedifferentiated liposarcoma, and undifferentiated pleomorphic sarcoma [12,13]. Bone-forming reactive lesions such as myositis ossificans should also be considered in the differential diagnosis. This may cause difficulties in reaching the correct diagnosis. Focal staining in immunohistochemical studies may also cause confusion. Staining with varying intensity and nonspecificity can be seen with immunohistochemical studies such as SMA, CD99, EMA, and desmin. Although histopathological evaluation is the gold standard for the diagnosis of ESOS, correlation with radiological findings is essential for a definitive diagnosis.

Although there is no standard treatment protocol for ESOS, the cornerstone of treatment is radi-cal surgery and polychemotherapy. Due to the rare nature of this disease, data on the type of chemotherapeutic combinations are not available. There are studies with positive results when treated like traditional osteosarcomas [14]. Adjuvant radiotherapy may be considered in the case of the unresectable or incompletely resected primary tumor [15].

In conclusions, ESOS is a rare malignant mesenchymal neoplasia with few reports in the literature. Radiological findings may be nonspecific. It should be considered in the differential diagnosis when non-skeletal tumors with osteoblastic differentiation are encountered. Since they differ from osteosarcomas of the bone in terms of treatment and prognosis, it is important for disease management to reach the diagnosis by correlation of histological and radiological findings.

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