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FOOT & ANKLE

LEIOMYOSARCOMA OF THE OS CALCIS

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ABSTRACT

A primary leiomyosarcoma of the os calcis, occurring in a 66-year-old woman and treated by below-the-knee amputation, is reported. The diagnosis was confirmed by electron microscopy, and supported by immunochemistry. These techniques can be performed in a primary spindle cell osseous neoplasm to distinguish leiomyosarcoma from fibrosarcoma and malignant fibrous histiocytoma. A total of 28 cases of primary leiomyosarcoma of extragnathic bones have been reported in the literature with one occurring in the foot. This is the first reported case involving the os calcis and the first reported leiomyosarcoma of the foot in the English literature.

INTRODUCTION

Primary leiomyosarcoma of the bone is uncommon. A search revealed only 28 cases in the literature.[1,3,4,6–14] Of these cases, only one occurred in the foot and was located in the talus.[12] This paper reports a case of primary leiomyosarcoma of the os calcis, discusses the criteria for diagnosis, and briefly reviews the literature.

CASE REPORT

A 68-year-oid white female presented with a 4-year history of pain and swelling of the right foot and ankle. During the preceding 6 months, the pain progressed, requiring narcotic medication and a walker for ambulation.

The patient had a previous history of "ankle sprain," 7 and 4 years prior to admission. Radiographs of the foot and ankle during that time were considered normal. The "sprains" resolved uneventfully with conservative treatment.

In 1983, 2 years prior to presentation, she was hospitalized elsewhere for the evaluation of increasing right ankle pain. She had no history of associated trauma. An extensive rheumatological evaluation was performed including an open biopsy of the right ankle synovium. All results were considered normal and no diagnosis was made.

The pain and swelling of the right foot and ankle continued and became more severe, prompting the patient to seek further evaluation.

Physical examination revealed marked medial and lateral swelling of the ankle. There was extreme tenderness over the peroneal and posterior tibial tendons with limitation in ankle and subtalar motion. Routine laboratory tests and chest x-rays were normal. Radiographs of the ankle and foot revealed marked osteopenia with no evidence of a lytic lesion or obvious neoplasm (Fig. 1). A computed tomography (CT) scan was not performed based on the assumption that the problem was soft tissue. Magnetic resonance imaging (MRI) was not available. The initial diagnosis was chronic peroneal and posterior tibial tenosynovitis.

In April 1985, the ankle was explored due to unremitting pain. At surgery, the posterior tibial tendon sheath was opened and mild tenosynovitis discovered. An underlying mass was noted and incision through the sheath exposed a neoplasm involving the entire os calcis and extending through the medial cortex and into the subtalar joint. The biopsy was interpreted as a spindle cell sarcoma. A work-up for primary or metastatic disease, including a CT scan of the abdomen and pelvis and a long bone survey, was negative. The patient subsequently underwent a below-the-knee amputation. She is alive and clinically disease free 38 months following amputation.

Pathology

The specimen consisted of multiple irregular portions of soft gray white tissue replacing the os calcis. The mass extended into the talocalcaneal joint, adjacent soft tissue and skeletal muscle. The remainder of the foot, including the amputation margin was free of tumor.

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Microscopic examination revealed interlacing fascicles of spindle cells with pale eosinophilic cytoplasm and elongated, variable hyperchromatic nuclei. There were frequent prominent nucleoli with blunted ends (Figs. 2 and 3). Mitosis ranged from two to five per high power field. Trichome stain revealed red staining of variable intensity of some tumor cells. Reticulin stain disclosed reticulin fibers individually wrapping many tumor cells.

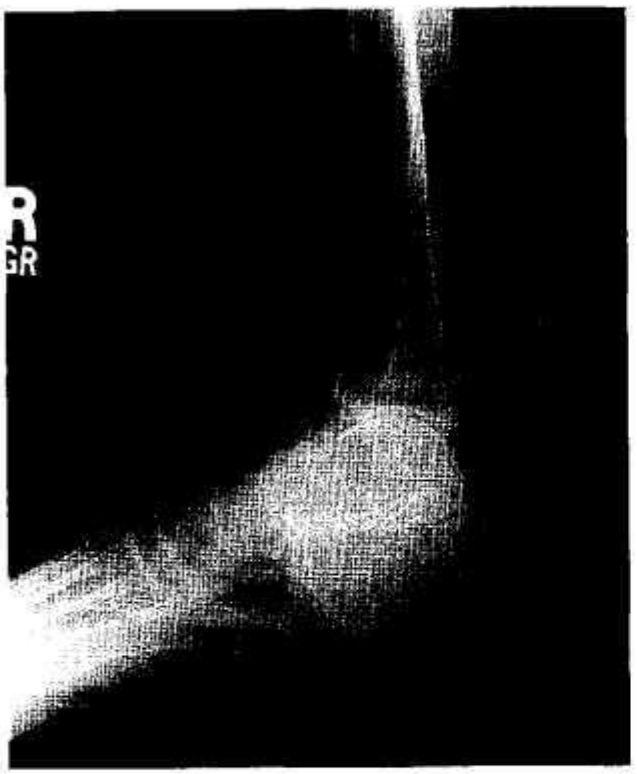

Fig. 1. Lateral radiograph of right ankle showing diffuse osteopenia and an indistinct posterior facet of the os calcis.

Electron microscopy revealed abundant intracytoplasmic bundles of fine filaments, mostly arranged parallel to the long axis of the tumor cell. Dense body formation was conspicuous in the bundles of fine filaments and occasionally in apposition to the incomplete external lamina. Pinocytic vesicles were prominent in places (Fig. 4). These features are typical of smooth muscle and confirm the diagnosis of leiomyosarcoma.[5,6,8,10]

Peroxidase-antiperoxidase immunochemistry demonstrated the presence of smooth muscle actin and the absence of desmin and cross striations in the tumor cells. This supported the diagnosis of leiomyosarcoma.[1,5,6]

DISCUSSION

Primary leiomyosarcoma of bone may be confused with the more commonly occurring spindle cell sarcoma of bone.[2,3,6,10,11] A well or moderately differentiated leiomyosarcoma can be recognized by light microscopy, confirmed by electron microscopy, and supported by immunochemistry. On light microscopy, malignant cells of smooth muscle origin frequently have cytoplasmic glycogen, and longitudinally arranged, parallel rows of linear striations.[10,13] On electron microscopy, myofilaments, dense bodies, basement membranes, and pinocytotic vesicles are characteristic of smooth muscle.[6,10] These features, although diagnostic, may be less prominent and sometimes absent, in poorly differentiated leiomyosarcomas.[5]

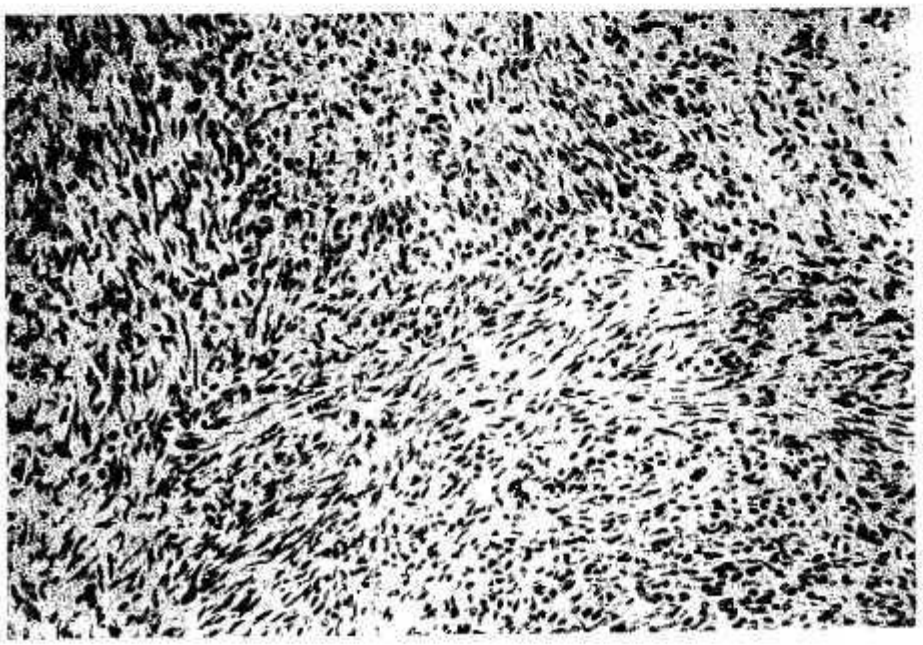

Fig. 2. Neoplasm of os calcis consisting of interlacing fascicles of fusiform malignant smooth muscle cells. Hematoxylin and eosin *x* 40.

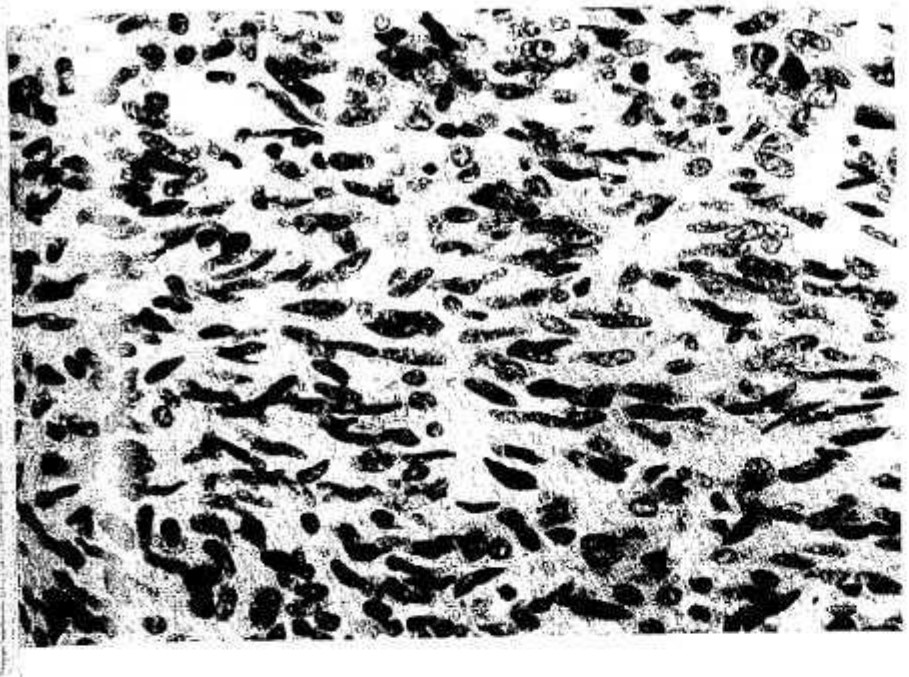

Fig. 3. Higher magnification of Figure 2 showing many pleomorphic tumor cells with blunt end nuclei and a large nucleolus. Hematoxylin and eosin x 100.

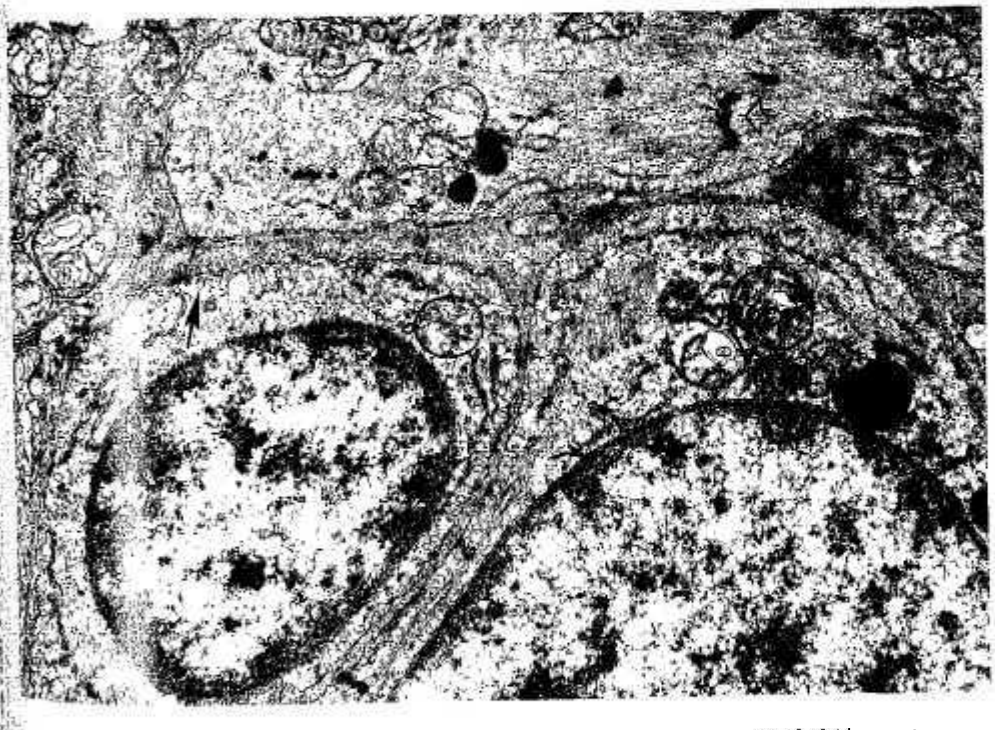

Fig. 4. Electron micrograph showing abundant intracytoplasmic fine filaments with dense body formation (open arrow) and presence of pinocytic vesicles (thin dark arrow), x 23.500.

Immunochemistry is of value in the diagnosis of myogenic neoplasms.[5,6] Desmin has been useful as a marker to confirm the rhabdomyosarcomatous nature of a malignant tumor.[1] Its value in leiomyosarcoma is variable.[1,5] Smooth muscle actomysin enhances the recognition of this neoplasm. Its absence excludes rhabdomyosarcoma fibrosarcoma, and malignant fibrohistiocytoma.[6]

The histogenesis of leiomyosarcoma of bone remains unknown.[8,13] Origin from the media of intraosseous blood vessels has been suggested and remains plausible.[7,8,10,13,14] Another source could be a primitive mesenchymal cell such as a myofibroblast that has the potential for smooth muscle differentiation.[10,13,14]

The small number of primary leiomyosarcomas of extragnathic bones makes it difficult to adequately characterize the lesion. Previously reported cases occurred in the femur (ten cases: two proximal, eight distal), tibia (seven cases: six proximal, one distal), humerus (four cases: all proximal), and the fibula (two cases: both distal). The clavicle, rib, sarcroiliac region, acetabulum, and talus have been reported with one

case each. The lesion is generally osteolytic, larger than 5 cm, and tends to involve the metaphysis, often with extension into the epiphysis or diaphysis. The peak incidence is in the seventh and eighth decades with a female to male ratio of 1 to 1.5. Fifty percent of the cases metastasize, most often to the lungs. Other sites include bone, liver, lymph nodes, and skin. [1,3,4,6–14]

Of the eight patients with follow-up greater than 5 years, five remain disease free, one is alive with metastatic disease and two died with metastatic disease.[1] Ten patients died less than 5 years after diagnosis, all with metastasis.[1,6,8,13] Our patient remains disease free 38 months after amputation.

Primary leiomyosarcoma should be considered in the differential diagnosis of primary malignant osteolytic neoplasms especially in the older age group. Its smooth muscle characteristics must be distinguished from other primary spindle cell sarcomas of bone. The accurate diagnosis requires light and electron microscopy, and perhaps immunochemistry.

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