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 immunocytochemistry. 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. Of these cases, only one occurred in the foot and was located in the talus. 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-old 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," and 4 years prior to admission. Radiographs of the foot and ankle were considered normal. The "sprains" resolved uneventfully with conservative treatment.

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.
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. Trichrome stain revealed red staining of variable intensity of some tumor cells. Reticulin stain disclosed reticulin fibers individually wrapping many tumor cells.

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.1,5,6

Peroxidase-antiperoxidase immunocytochemistry 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

DISCUSSION

Primary leiomyosarcoma of bone may be confused with the more commonly occurring spindle cell sarcoma of bone.1,2,3,6,10,11 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.1,10,13 On electron microscopy, myofilaments, dense bodies, basement membranes, and pinocytic vesicles are characteristic of smooth muscle.6,10 These features, although diagnostic, may be less prominent and sometimes absent, in poorly differentiated leiomyosarcomas.5
Immunohistochemistry is of value in the diagnosis of myogenic neoplasms. Desmin has been useful as a marker to confirm the rhabdomyosarcomatous nature of a malignant tumor. Its value in leiomyosarcoma is variable. Smooth muscle actin enhances the recognition of this neoplasm. Its absence excludes rhabdomyosarcoma, fibrosarcoma, and malignant fibrous histiocytoma.

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

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, sacroiliac region, acetabulum, and talus have been reported with one
eas 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. 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. Ten patients died less than 5 years after diagnosis, all with metastasis. 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.

REFERENCES