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Deep soft-tissue leiomyoma of the forearm mimicking a primary bone tumor of the ulna

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Leiomyomas of the soft tissues are rare in general, and extremely uncommon in the forearm. In general, leiomyomas are benign soft-tissue tumors that occur where smooth muscles are present. We present a case of soft-tissue leiomyoma of the forearm eroding the midshaft of the ulna, with emphasis on radiological diagnosis and histopathological correlation.

Introduction

Leiomyomas are tumors derived from smooth muscle. They represent 4.4% of all benign soft-tissue neoplasms (1). Smooth muscle is present in small quantities in the extremities. Leiomyomas are classified into three groups: cutaneous leiomyomas that arise from the erector pili muscle; vascular leiomyomas that arise from smooth muscle of the vein; and leiomyomas of deep soft tissues (2). Deep soft-tissue leiomyoma is extremely rare.

Case report

A 16-year-old boy complained of pain and swelling in the left forearm of two month's duration. The swelling was sudden in onset and progressive in nature. There was no history of trauma or fever. Routine blood investigations were within normal limits. Plain radiography of the left forearm in frontal and lateral projections (Figs. 1A and B) showed a soft-tissue shadow in the interosseous space causing scalloping and erosions of the midshaft of the ulna.

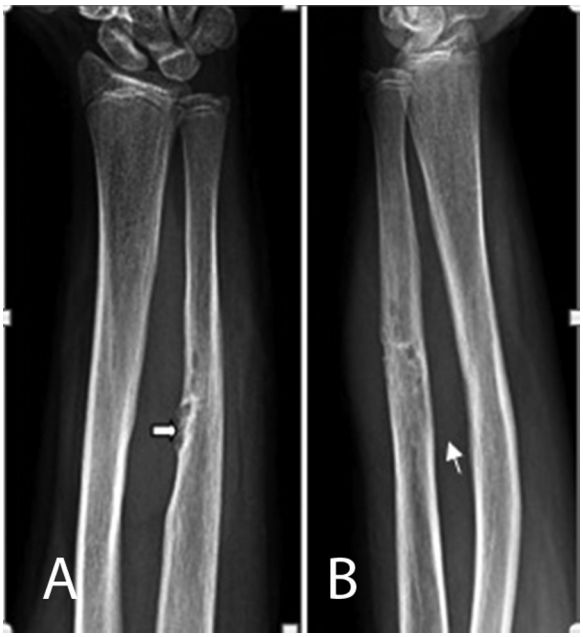

Figure 1. Plain radiographs in frontal (A) and lateral (B) projections show a soft-tissue shadow (upward arrow) in the interosseous space causing scalloping and erosion (right arrow) of the midshaft of the left ulna.

The patient was further evaluated with CT and MRI. Noncontrast CT of the left forearm in a reformatted coronal and sagittal view (Figs. 2A and B) demonstrated a relatively well-defined, soft-tissue-density lesion involving the intermuscular compartment of the left forearm in the dorsal aspect of the ulna. No calcification/hemorrhage was

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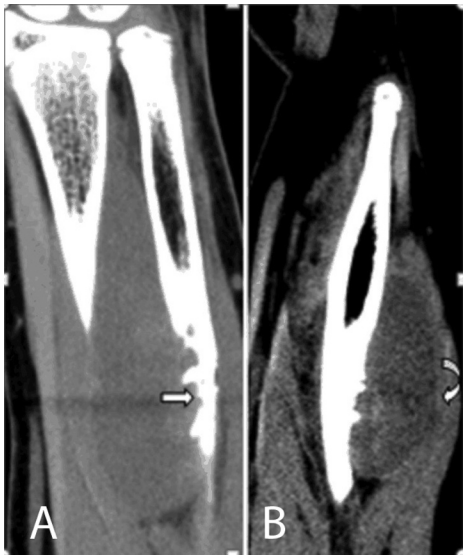

Figure 2. Nonontrast CT of the left forearm in reformatted coronal (A) and sagittal (B) views shows a well-defined, soft-tissue density (curved arrow) lesion involving the intermusclar compartment and causing scalloping and erosion of the adjacent ulnar cortex (right arrow).

noted within the lesion. The lesion was seen to cause erosion of the adjacent ulnar cortex. No periosteal reaction was noted.

MRI of the left forearm revealed a well-defined mass lesion in the dorsal aspect of the midshaft of the ulna. The lesion appeared mildly hyperintense on T1W images (Fig. 3A) and was not suppressed on T1W fat-suppressed images

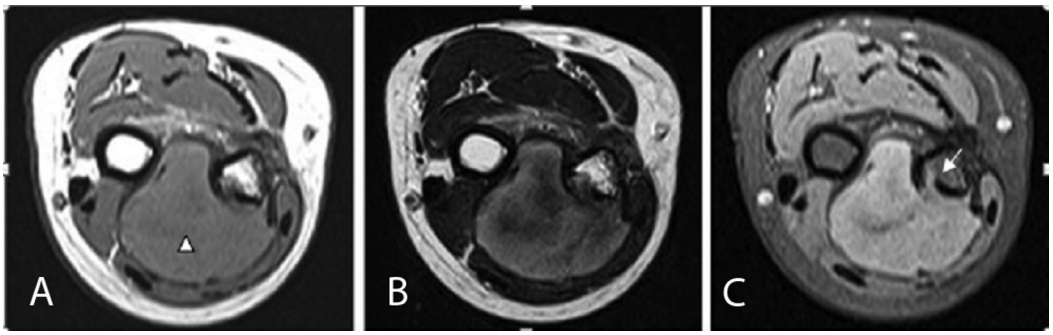

Figure 3. F18-FDG PET/CT study 4 months before the MPS demonstrating the left upper lung (A), left 5th rib (B), and right T5 vertebral lesions (C) with subtle increase in F18-FDG uptake.

(Fig. 3B). The lesion was hyperintense to the adjacent muscle on T2-weighted images (Fig. 3C). The lesion was seen to anteriorly displace and impinge on the anterior intraosseous nerve, and posteriorly displace the extensor muscles of the forearm.

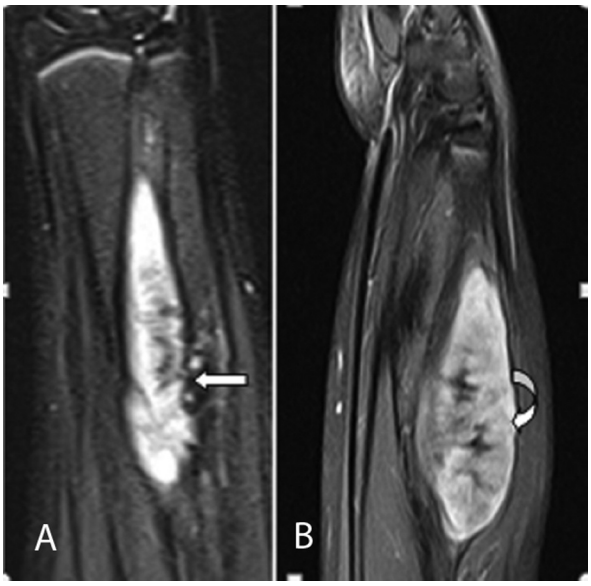

Figure 4. MRI of left forearm in coronal (A) and sagittal (B) T2W STIR shows well-defined, hyperintense, intermuscular lesion involving the dorsal aspect of ulna (curved arrow) causing scalloping and cortical irregularity with a mild intramedullary extension (left arrow).

The lesion was also seen to cause scalloping and cortical irregularity of the ulna, with a mild intramedullary extension (Fig. 4).

Considering the patient's age and the nature of the lesion on imaging, the possibility of Ewing's sarcoma was raised. Core biopsy of the lesion showed a benign, smooth-muscle neoplasm at microscopy, favoring a diagnosis of fibromatosis. The tumor was excised completely (Fig. 5) and sent for histopathological examination. Initial microscopic examination (Fig. 6) revealed a well-circumscribed lesion composed of spindle cells arranged in a whorled pattern. The tumor cells contained elongated nuclei with a moderate amount of eosinophilic cytoplasm. There was no evidence of necrosis, mitoses, or nuclear atypia. Further immunohistochemical (IHC) staining showed strong reactivity for vimentin, smooth-muscle actin, and desmin, and a negative reaction against S-100 that favored a diagnosis of leiomyoma (Fig. 7). Nonvascularized fibula bone grafting of the left ulna with extensor tendon repair was performed a month later.

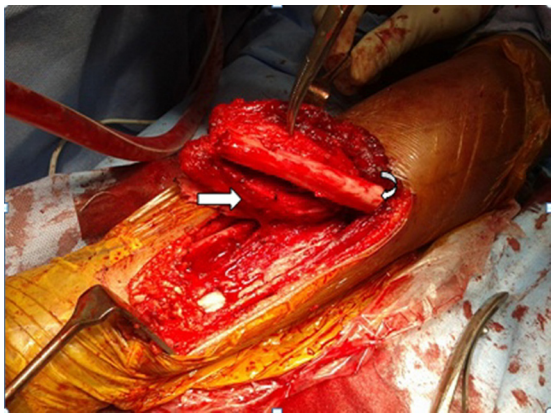

Figure 5. Intraoperative image shows the soft-tissue tumor (white arrow) resected with the ulnar bone (curved arrow).

Discussion

Leiomyomas occur at almost any age, with reports ranging from 3 to 62 years (mean age, 25 years) and a male:female ratio of 2:1 (3). Leiomyomas of the limbs are divided into superficial and deep soft-tissue tumors. Superficial tumors include both cutaneous and subcutaneous

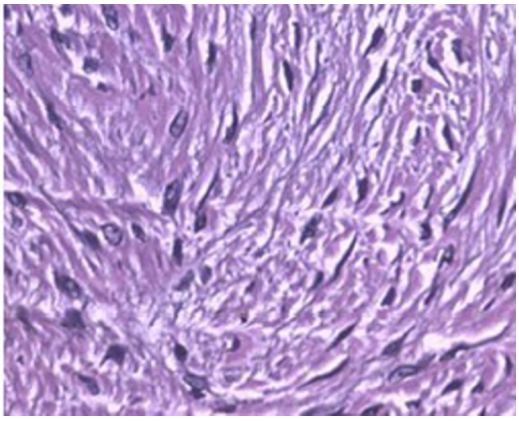

Figure 6. Histopathological examination shows a well-circumscribed lesion composed of spindle cells arranged in a whorled pattern. The cells show an elongated nucleus with a moderate amount of eosinophilic cytoplasm (H & E x 200).

lesions (4). The deep soft-tissue leiomyomas are further classified into vascular (originating from vessel wall smooth muscle) and nonvascular tumors (5). Only sporadic cases of deep soft-tissue leiomyoma with involvement of bone have been reported so far in the English literature.

Two theories have been postulated for the pathogenesis of this rare tumor. Goodman et al believe that deep leiomyomas arise from undifferentiated mesenchymal cells or smooth-muscle rests (6). Stout et al suggest that these tumors may instead arise from the smooth muscle in the walls of blood vessels (7).

These tumors are usually not recognized until clinical symptoms appear. Such symptoms include pain and swelling. In our case, we found that the pain was due to impingement on the anterior intraosseous nerve by the expanding mass lesion. Stout et al suggest that contraction of the smooth muscles in these tumors can be associated with spontaneous paroxysmal pain (7). Changes in the size of these tumors have been observed in the vascular type of leiomyoma, particularly during pregnancy (8).

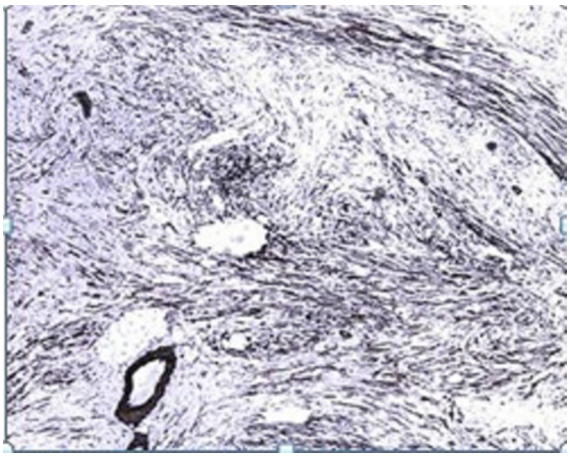

Figure 7. Immunohistochemical staining shows strong reactivity to smooth muscle actin (H & E x100).

Scattered calcifications have been reported in isolated deep soft-tissue leiomyomas, which may lead to their being mistaken for myositis ossificans (6).

Primary leiomyomas of bone are extremely rare, with fewer than 20 cases reported so far. The majority of the reported cases of leiomyoma involving the bone include the mandible, the maxillary tooth socket, and the temporal bone. Very few cases of leiomyoma involving the appendicular skeleton (which include the tibia, ulna, and femoral neck) have been reported (9).

The imaging features of deep soft-tissue leiomyomas are nonspecific and similar to those of many other soft-tissue neoplasms. The radiological differential diagnosis includes lipoma, leiomyosarcoma, schwannoma or neurofibroma, hemangioma, and soft-tissue giant-cell tumor of the tendon sheath.

Considering the patient's age and the imaging characteristics, we believed that the most probable diagnosis was Ewing's sarcoma. Ewing's sarcoma typically occurs in children and adolescents between 10 and 20 years of age and has a slight male predilection (10). These sarcomas tend to be poorly marginated, with periosteal reaction, and over 80% demonstrate extension into adjacent soft tissues. In long bones, the tumor is almost metaphyseal or diaphyseal in location (11). Retrospectively, the lesion in our case had a well-defined margin and did not show any typical periosteal

reaction. Moreover, the center of the lesion was predominantly in the intermuscular compartment--in contrast to Ewing's sarcoma, which is usually intraosseous.

The treatment of choice is complete surgical excision of the tumor along with surrounding muscle. The rate of recurrence is very rare, and this tumor rarely undergoes malignant transformation (12). However, there are no available data in the literature concerning the long-term prognosis for this rare tumor.

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