CASE REPOSITORY

Extraskeletal Osteosarcoma of the Hand

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Extraskeletal osteosarcoma is a rare disease that uncommonly affects the upper extremity. A 46-year-old African American man presented for evaluation of a right middle finger mass. Excisional biopsy demonstrated extraskeletal osteosarcoma of the soft tissues. We performed a transmetacarpal ray resection. *(J Hand Surg Am. 2017;■(■):1.e1-e4. Copyright © 2017 by the American Society for Surgery of the Hand. All rights reserved.)*

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Online Exclusive

EXTRASKELETAL OSTEOSARCOMA (ESOS) is a rare mesenchymal malignancy that usually occurs in the fifth or sixth decade of life. Extraskeletal osteosarcoma is reported to account for approximately 1% of soft tissue sarcomas and 4% of all osteosarcomas.[1–3] It is typically a high-grade tumor; fewer than 10 cases of low-grade ESOS have been described.[4]

The location of the tumor varies but it most commonly affects the lower extremity and arises deep to the fascia.[4] Other reported locations include the retroperitoneum and, less commonly, the upper extremity. It typically does not present in the hand.[1] In the extremities, ESOS typically presents as an enlarging soft tissue mass with associated pain.[5,6]

Extraskeletal osteosarcoma is an aggressive tumor. It has a local recurrence rate of 45% to 70% and rates of metastasis up to 80%.[5] As with other variants of osteosarcoma, the lungs are the most frequent location of metastatic spread. Extraskeletal osteosarcoma has a reported 5-year survival rate ranging from 24% to 46%; the median survival for metastatic ESOS is 8 months.[1] In localized disease, good prognostic factors include negative margins at resection and a superficial location of the tumor.[4] Management of ESOS involves wide excision of the tumor with consideration given to radiotherapy and chemotherapy, although to date, studies have failed to prove long-term survival benefits of these adjuvants.[4,5]

As is requisite for the diagnosis of any osteosarcoma, ESOS produces osteoid; however, it does so with no attachment to bone or periosteum.[1,4] It has also been known to produce cartilaginous matrix. Like other soft tissue sarcomas, prior radiation therapy is a risk factor for the development of ESOS, and up to 10% of patients with ESOS have a reported history of radiation.[3,4]

This case describes a 46-year-old African American man who presented for evaluation of a right middle finger mass. Excisional biopsy proved it to be extraskeletal osteosarcoma; a partial ray resection was performed. This tumor is exceedingly rare and only 2 other cases of ESOS of the hand have been documented.

CASE REPORT

A 46-year-old African American man with no history of malignancy was referred to the hand clinic of our institution in September, 2014 with a 2-year history of a right middle finger mass. He reported that it had been unsuccessfully lanced by 2 outside providers. The mass progressively enlarged, ultimately interfering with his occupational function. Associated symptoms at the time of presentation included a reported sensory deficit to the radial aspect of the middle finger and spasms in the right upper extremity.

On examination, the right middle finger revealed a mass located over the proximal and middle phalanges.

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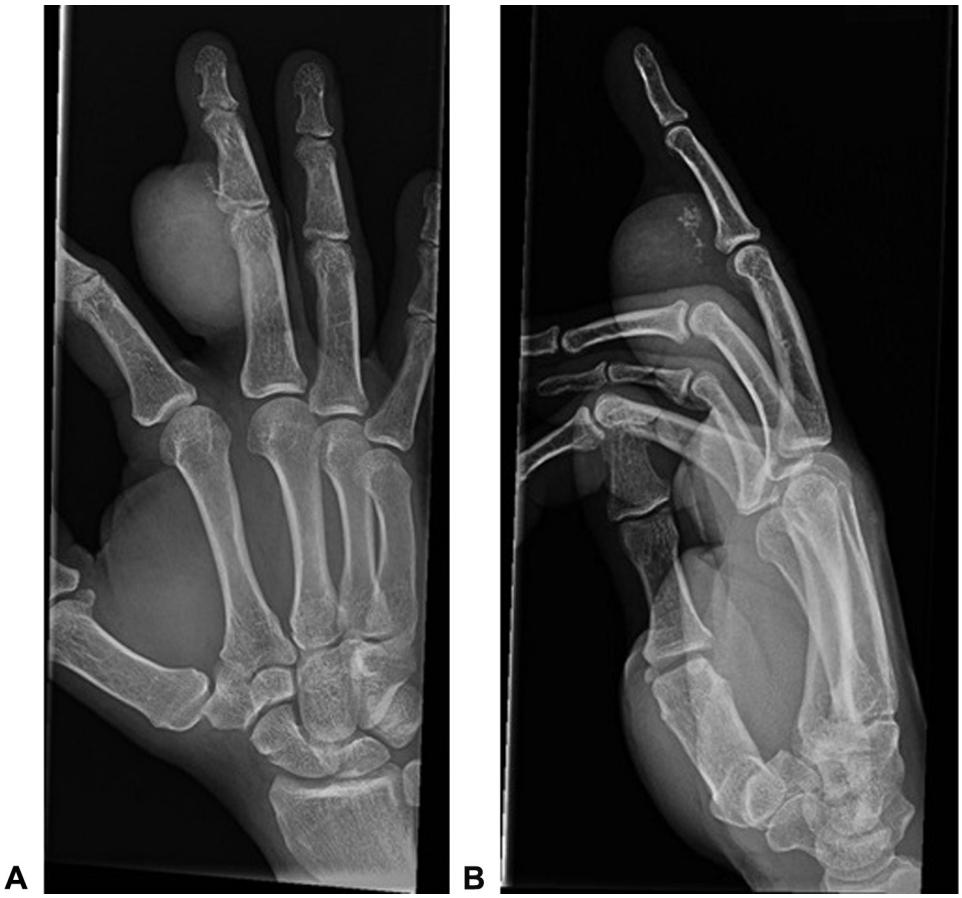

FIGURE 1: A Oblique and **B** lateral radiographs of right hand demonstrating a large soft tissue mass of the middle finger without osseous involvement. Stippled calcifications within the soft tissue mass are also noted indicating a possible cartilaginous matrix.

It was centered about the volar and radial aspects of the digit at the level of the proximal interphalangeal joint. The mass was firm and nonfluctuant, with no associated erythema or tenderness to palpation. Sensation distal to the mass was intact. Motion was restricted at the proximal interphalangeal joint owing to a mass effect, but the patient was able to flex and extend the distal interphalangeal joint normally.

Radiographs of the digit (Fig. 1) revealed a large soft tissue mass with no destruction of the adjacent phalangeal cortex, periosteal reaction, or other osseous involvement. There was an area of stippled calcification within the soft tissue mass, suggesting a cartilaginous matrix. Magnetic resonance imaging of the tumor (Fig. 2) revealed a 3.7 × 2.9-cm heterogeneous soft tissue lesion. It was largely hypointense on both T1 and short TI inversion recovery sequences and did not enhance with contrast. The differential diagnosis for the mass at that time consisted of both benign and malignant processes. Consideration was given to a giant cell tumor of tendon sheath, extraskeletal chondroid neoplasms, and synovial sarcoma, among others. Subsequently, the risks, benefits, and alternatives of excisional biopsy were discussed with the patient in detail. The patient consented to the procedure and excisional biopsy was performed.

Intraoperatively, the digital anatomy was greatly distorted because of the tumor's size. The radial neurovascular bundle was sacrificed owing to its complete encasement in the mass. The mass was removed and sent to pathology for tissue examination.

The pathology team examined the mass and further consultation was obtained from 2 tertiary musculoskeletal pathology centers. There was consensus that the tumor was a high-grade, giant cell–rich, extraskeletal osteosarcoma with telangiectatic features. Under low magnification, the tissue demonstrated dense sheets of osteoid alternating with cellular foci rich in multinucleated giant cells (Fig. 3). Under high magnification, the tissue demonstrated severe cytologic atypia and mitotic activity (Fig. 4).

After the diagnosis was established, a staging workup was initiated. Chest computed tomography scan revealed a solitary, enlarged mediastinal lymph node, and a whole-body bone scan revealed no evidence of osseous metastatic disease.