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An Extraosseous Ewing Sarcoma of the Index Finger Masquerading as a Benign Tumor

Roos Wolthuizen, MD,* Judith Nieken,† Jelle Overbosch,‡ Shariselle Mirna Wietske Pool, MD, PhD*

Ewing sarcoma is a malignant, small, round, blue cell tumor that often affects the long bones and pelvis. It is rarely seen in the bones of the hand. A minority of Ewing sarcoma cases arise in soft tissue. This case report details the clinical and radiological presentation of an extraosseous Ewing sarcoma of the index finger in a 22-year-old woman. Treatment consisted of an index ray amputation followed by adjuvant chemotherapy. After 1 year, the left hand had excellent function with an acceptable cosmetic appearance and no evidence of metastasis or local recurrence. (*J Hand Surg Am.* 2020;45(4):366.e1-e4. Copyright © 2020 by the American Society for Surgery of the Hand. All rights reserved.)

Key words Case report, Ewing sarcoma, Ewing tumour, extraosseous, phalanx.

WE PRESENT THE CASE OF A 22-year-old woman who was referred to our clinic in July, 2017 with a 6-month history of local swelling affecting the volar surface of the left index finger. Until then, primary care consisted of expectant management. Over 4 weeks, the mass grew progressively although the patient reported no pain. She had no comorbidities, fever, systemic symptoms, or history of trauma to the hand. There was no family history of malignancies.

CASE REPORT

Clinical examination revealed a reddish blue transparent swelling $2 \times 1 \text{ cm}^2$ in size on the volar side of the proximal phalanx of the index finger. The lesion was firm, nonpulsatile, and nontender on palpation (Fig. 1). There was a flexion deficit of 45° degrees of the proximal interphalangeal joint of the index finger.

Plain radiographs showed an extraosseous mass around the proximal phalanx without indications of bone involvement such as periosteal reaction or disruption of the cortex (Fig. 2). Ultrasound examination revealed a soft tissue mass with varying echogenicity, both hypoechoic and hyperechoic. Extensive arterial and venous flow signals were also seen, without a clear supplying or draining vascular system. As a result of these findings, the differential diagnoses included a vascular malformation.

Magnetic resonance imaging (MRI) revealed a well-demarcated mass in the subcutaneous tissue on the volar side of the proximal phalanx, measuring $2.0 \times 2.7 \text{ cm}$ in the axial plane and 2.3 cm longitudinally (Fig. 3). There was clear contact with the flexor tendon without signs of involvement or disruption of the tendon, consistent with the prior ultrasound. There was a small volume of fluid in the tendon sheath, probably reactive to the mass. There was also clear contact with the skin and the radial neurovascular bundle, without signs of involvement. There were no signs of bone involvement. The mass showed a relatively homogeneous intermediate signal on the unenhanced T1-weighted images, heterogeneous high signal on the fluid-sensitive images, and relative homogeneous enhancement after intravenous administration of gadolinium-based contrast agent, with the exception of a few small cystic areas. There

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FIGURE 1: Left hand with **A** anterior and **B** lateral view of the swelling on the index finger.

FIGURE 2: Posteroanterior X-ray of the left hand without evidence of bone involvement.

FIGURE 3: Sagittal MRI of the left index finger showing a well-demarcated subcutaneous mass on the volar side of the proximal phalanx, without soft tissue involvement. Coloring in the pulp is caused by focal moderate fat suppression of (normal) subcutaneous adipose tissue and bone marrow.

were no calcifications and feeding or draining vessels were not evident. Because of the demarcation and lack of involvement of surrounding tissue, sarcoma seemed less likely; however, it was not excluded owing to progressive growth.

After a delay for logistical reasons related to the arrangement of imaging studies, we performed

FIGURE 4: Macroscopic picture of the excised tumor.

excisional biopsy of the tumor 2 months after the patient was referred to our clinic. No ingrowth was identified in surrounding healthy tissue. Histopathological examination showed monomorphic atypical cells with enlarged, round nuclei with a finely divided chromatin pattern (Figs. 4, 5). The cells exhibited immunoreactivity for CD99, vimentin, and pankeratin. The pathology diagnosis was an incompletely excised, extraosseous Ewing sarcoma (EES)/primitive neuroectodermal tumor. The multidisciplinary team confirmed the diagnosis as Ewing sarcoma (ES) with proven ES breakpoint region translocation.

Two weeks after the diagnosis was confirmed, the patient underwent an index ray amputation with free surgical margins (Fig. 6). Computed tomography of the neck, thorax, and abdomen obtained before chemotherapy showed no evidence of metastases. The cervical lymph nodes were enlarged with features suggestive of reactive lymphadenitis. Biopsies of the iliac crest bone did not show tumoral infiltration. She received postoperative chemotherapy consisting of 6 sessions of vincristine, ifosfamide, doxorubicin, and etoposide and 8 of vincristine, actinomycin-D, and ifosfamide. Seven months after ray amputation, she completed hand therapy with excellent active range of motion of the third metacarpophalangeal joint. More than 1 year after ray amputation, there was no evidence of metastasis or local recurrence on computed tomography and magnetic resonance imaging scan, respectively.

FIGURE 5: Microscopic histologic slide of the ES: **A** Overview and **B** close-up.

FIGURE 6: Left hand 10 days after surgery, after index ray amputation.

DISCUSSION

Ewing sarcoma most often affects the long bones and pelvis. Hand and wrist involvement represent less than 1% of all ES. The metacarpal and proximal phalanges are the most common locations in the hand, and the thumb (28%) and middle finger (28%) are the most affected.^{1,2} Ewing sarcoma often mimics osteomyelitis both clinically with signs of local inflammation and radiographically because of the cortical destruction and aggressive periosteal reaction.^{1–3} A minority of cases of ES arise in soft tissue. The lower extremities, head, neck, pelvis, and paravertebral spaces are commonly affected extraskeletal sites.^{3,4} Only a single case of an EES in the hand³ was previously described in the literature.

Conventional radiographs and MRI are normally used to assess the primary origin (osseous or

extraosseous) and the extent of the tumor.³ Ultrasonography of EES often reveals a hypoechoic, heterogeneous mass with intratumoral flow signals. Extraosseous Ewing sarcoma is often represented by hypointense to isointense T1-weighted MRI images and hyperintense T2-weighted images with heterogeneous contrast enhancement.⁴ The definitive diagnosis ultimately follows from the histopathological examination of the resected specimen. Several studies^{1,3,4} suggested radical resection of the tumor and systemic chemotherapy as the best combination for management of ES.

This case is interesting in that the clinical and radiological findings indicated that the soft tissue tumor was benign. In retrospect, biopsy was required for the correct diagnosis and treatment should have been started much earlier. This delay occurred because a benign lesion, such as a vascular malformation, remained at the top of the differential diagnosis. The difficulty is that EES has no specific clinical manifestation. The sarcoma's site of origin particularly determines the type of accompanying symptoms.^{3,4} A rapidly growing mass with local pain is the most frequently presenting symptom. Remarkably, the patient did not report pain. This case highlights an exceptional but important differential diagnosis in patients presenting with a rapid growing soft tissue tumor in the fingers, with or without progressive pain. Although EES is rarely seen in fingers, it should be considered a possible diagnosis owing to the aggressive nature and associated mortality.

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