Global Surgery

Case Report ISSN: 2056-7863

Extraskeletal Ewing's Sarcoma of the Little Finger, a rare case

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Abstract

When Ewing's sarcoma arises in soft tissue rather than bone, it is referred to as extraskeletal Ewing's sarcoma (ESS). It is generally accepted to be between 15% and 20% of that of Ewing sarcoma of bone. Extraskeletal Ewing sarcoma usually manifests in young patients, with 85% of cases detected between 20 months and 30 years of age. The most commonly reported locations of extraskeletal Ewing sarcoma include the paravertebral region (32%), lower extremities (26%), chest wall (18%), retroperitoneum (11%), pelvis and hip (11%), and upper extremities (3%). Radiological features of Ewing's sarcoma are nonspecific. It is confirmed by features on histological analysis. In young people who present with soft tissue tumours, ESS should be considered. In the management of patients with tumours, imaging techniques are useful for biopsy guidance, evaluating the possibility of resection, and tumour response to treatment.

Introduction

Ewing's sarcoma (ES) was first described in 1921 [1]. It is the second most common primary bone tumor of childhood and adolescence [2]. Tumors morphologically indistinguishable from Ewing sarcoma of the skeletal system can present as soft tissue masses. In some cases, they simply represent soft tissue extensions of tumor originating in the underlying bone. In others, bone involvement is absent, and these are regarded as primary Ewing sarcomas of soft tissues [3-5]. The prevalence of extraskeletal Ewing sarcoma is generally accepted to be between 15% and 20% of that of Ewing sarcoma of bone [6,7]. Extraskeletal Ewing sarcoma usually manifests in young patients, with 85% of cases detected between 20 months and 30 years of age [7]. The most commonly reported locations of extraskeletal Ewing sarcoma include the paravertebral region (32%), lower extremities (26%), chest wall (18%), retroperitoneum (11%), pelvis and hip (11%), and upper extremities (3%) [5,8,9-11].

Case report

A 18 year old right hand dominant boy farmer by occupation presented with a 1 year history of swelling over left little finger with insidious onset and gradually progressive in size. It was associated initially with dull aching pain that later worsened with rapid increase in size since last 1 month. He had history of incision and drainage by a quack 2 months back presuming it to be abscess. There was no history of trauma, fever, cough, dyspnea, and body aches. There was history of reduced appetite and weight loss since 1 month. He had no other co-morbid medical illness.

On physical examination, the mass was 6×3.5×3 cm, immobile and fixed involving 5th finger extending upto distal palmar crease. Both 4th & 5th fingers were widely apart (Figures 1 and 2). There was restriction of little finger movements. There was no regional lymphadenopathy. All laboratory findings were within normal range.

Conventional radiographs revealed enlarged soft tissue opacity in left 4th web space, extending to proximal and middle phalanx of left 5th finger with evidence of well-defined osteolytic lesion associated with cortical break noted in lateral aspect of left 5th proximal metacarpal. Rest of metacarpals, phalanges and carpal bones appeared normal. Incision biopsy report was in favor of Ewings sarcoma.

MRI Lt. wrist & hand (Figure 3) showed evidence of well-defined soft tissue lesion along medial aspect of little finger of the left hand in between two tendons of extensor muscles of 4th and 5th finger. There was widening of the space between 4th and 5th finger. The lesion was seen abutting the medial aspect of proximal and middle phalanx of little finger with fine marginal erosion of the medial cortex of middle phalanx and altered signal intensity of the proximal phalanx could suggest possibility of secondary involvement. The lesion was showing

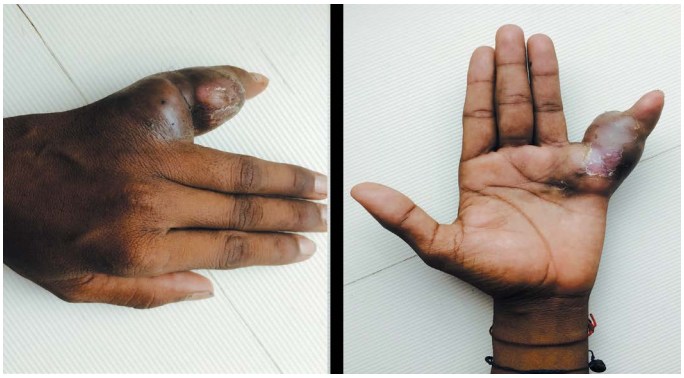

Figure 1. Swelling of Lt. Little finger.

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Key words: *Extra Skeletal Ewing's Sarcoma, Little Finger, MRI, histopathology*

Received: August 12, 2016; **Accepted:** August 29, 2016; **Published:** August 31, 2016

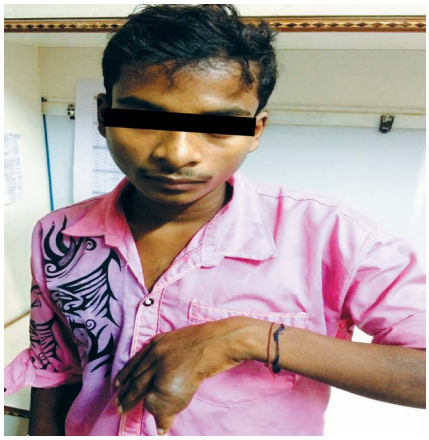

Figure 2. Swelling of Little finger.

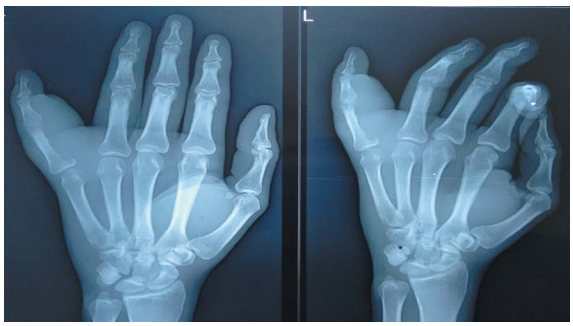

Figure 3. MRI of Lt wrist & hand.

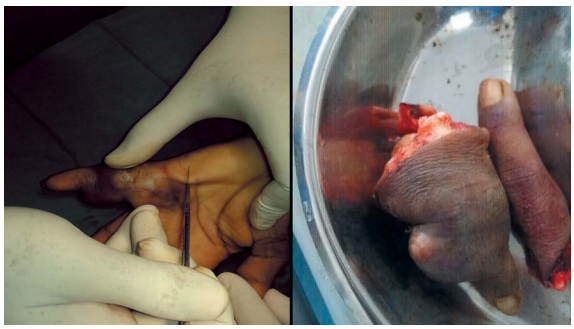

Figure 4. Excised fingers.

area of central necrosis. Size of the lesion was 6 x 3.5 cm. with area of necrosis around 2 cm. On post contrast images it showed good contrast enhancement with circumferential pattern. Rest of the visualised bones, muscles appeared normal. Soft tissue mixed intensity lesion with areas of circular or whorled contrast enhancement along medial aspect of little finger suggested possibility of neoplastic aetiology than infective cause. Possible differential diagnoses were – Soft tissue Ewings sarcoma, other soft tissue sarcoma, Lymphangiosarcoma (less likely). Marginal destruction of cortex of middle phalanx of little finger and altered signal intensity of the proximal phalanx could suggest secondary bone involvement (Haematogenous spread) (Figure 4).

Amputation of left 4th and 5th fingers including head & partly body of corresponding metacarpals were done to get R**0** resection. Gross examination (Figure 4) - showed, amputated 4th and 5th fingers. Little finger specimen measured 8.5x5x4 cm with firm to hard soft tissue mass encircling the bone. The 4th finger measured 8x2x2 cm in size. Microscopic examination (Figure 5 & 6) Sections from decalcified bony tissue from 4th& 5th finger as well as separated bony part (head of 5th metacarpal bone) showed no bone marrow involvement by malignancy. Section from soft tissue mass showed uniform round tumor cells having vescicular nucleus with scanty cytoplasm and few showing prominent nucleoli (white arrow). Figure 7 showed conventional radiograph of hand in post-operative period [8-11].

Discussion

Historically, Ewing sarcoma of soft tissue has included extraskeletal Ewing sarcoma and soft tissue primitive neuroectodermal tumour (PNET) [12-15]. In addition, extraskeletal Ewing sarcoma of the thoracopulmonary region is often referred to as Askin tumor [12-16]. Tefft and coworkers [17] described extraskeletal Ewing sarcoma in 1969 and reported four patients with paravertebral softtissue tumors that histologically resembled Ewing sarcoma. Because of chromosomal and histologic similarities with lack of differentiation, these lesions (extraskeletal Ewing sarcoma, soft tissue PNET, and Askin tumor) are considered to be in the Ewing sarcoma family of tumors [5,8,12-16]. These lesions are sarcomas likely of neuroectodermal origin that share the same cytogenetic marker, with translocation of chromosomes t(11;22)(q24;q12) [6,7,16,18,19] The pathologic appearance is identical to that described earlier for Ewing sarcoma of bone.

As with osseous lesions, extraskeletal Ewing sarcoma is rare in the black population [8,20]. Clinically, patients often have a large, rapidly growing, solitary, superficial or deep soft tissue mass measuring 5–10 cm at initial presentation [8,13]. Pain or tenderness has been reported in 49% of patients [8]. Extraskeletal Ewing sarcoma has been reported to show some differences in comparison with Ewing sarcoma of bone, including the following: *(a)* it does not show as distinct a predilection for male patients, but is more equally distributed between the sexes (although several studies have shown that male patients may be slightly more commonly affected); *(b)* patients are often slightly older (averaging around 20 years of age) by approximately 5–10 years; and

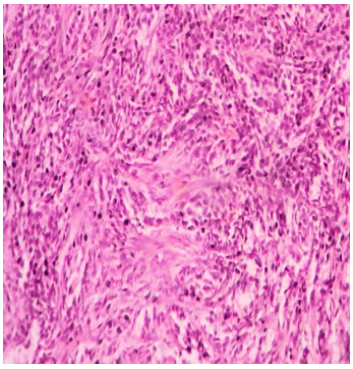

Figure 5. (10x) HP soft tissue.

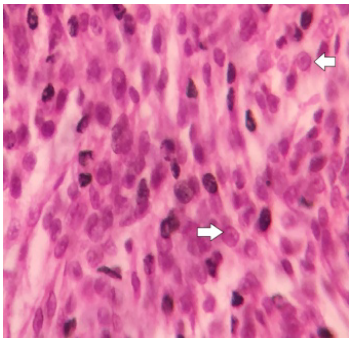

Figure 6. (40x) Prominent nucleoli.

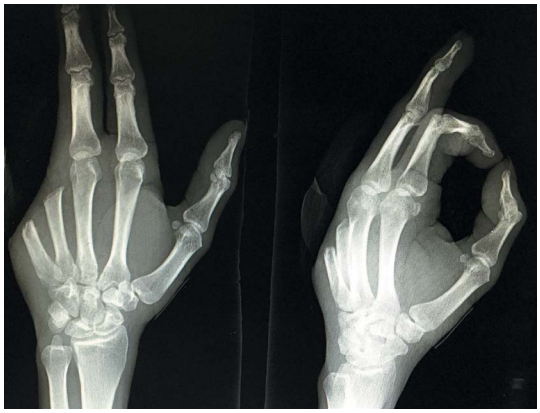

Figure 7. Post-op Xray Lt hand.

(c) it more commonly affects the trunk rather than the lower limbs [8,13,20,21].

At radiography, extraskeletal Ewing sarcoma may manifest as a large softtissue mass (50% of cases) or demonstrate a normal appearance. Adjacent bone erosion, cortical thickening, osseous invasion, or aggressive periosteal reaction may also be present (25%–42% of cases) [8,14,22]. Similarly, lesion calcification may be identified in up to 25% of cases [8,14,22]. MR imaging features are also nonspecific in evaluation of extraskeletal Ewing sarcoma. MR imaging demonstrates a soft tissue mass with heterogeneous signal intensity (91%) similar to that of skeletal muscle on T1weighted images and intermediate to high signal intensity on T2 weighted images in 100% of cases [22]. High signal intensity on long TR images predominates in 64% of cases [22]. Intermediate signal intensity areas seen on long TR images are likely due to a high degree of cellularity, as in osseous lesions. Areas of hemorrhage appear as high signal intensity on all pulse sequences and are not uncommon; fluid levels may also be evident. Focal areas of necrosis with low signal intensity on T1weighted images and high signal intensity on T2weighted images are also frequent [22]. As in other soft tissue masses, MR imaging is also useful for tumor staging and to evaluate the extent of involvement of surrounding structures [13].

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