Case Report

Ewing's sarcoma of proximal phalanx of the hand with skip metastases to metacarpals

Anshu Shekhar, Suresh Korlhalli[1], Gururaj Murgod[1]

Abstract

Ewing's sarcoma is the second most common malignant primary bone tumor of childhood and adolescence affecting mainly the diaphysis of long bones and flat bones. This tumor is extraordinarily rare in small bones of the hand and presents as a swelling with atypical radiological features of cystic and lytic lesion with scant periosteal reaction. The common differential diagnosis include osteomyelitis, tuberculosis, enchondroma and benign tumors. Moreover, skip metastasis to adjacent bones is even rarer. The prognosis of this condition is greatly influenced by the presence of metastasis at presentation, further emphasizing the importance of early diagnosis. Multimodality treatment using surgery, radiotherapy and chemotherapy is currently recommended though no consensus exists. We report a case of Ewing's sarcoma of the little finger proximal phalanx which was initially missed and developed skip metastasis to several metacarpals within 4 months.

Key words: Ewing's sarcoma, hand, phalanx, radiographic features, skip metastases
MeSH terms: Hand, sarcoma, Ewings, metastases

Introduction

Ewing's sarcoma is a malignant nonosteogenic primary tumor of the bone, which is mainly seen in the diaphysis of the long bones and the flat bones of the pelvis in young patients. Originally, James Ewing described it as a tumor arising from undifferentiated osseous mesenchymal cells; however, recent studies suggest that Ewing's tumor may be neuroectodermally derived from the primitive neural tissue.[1] This tumor is stated to comprise approximately 6% of the total malignant bone tumors, but is exceptionally rare in the small bones of hands and feet, where the reported incidence is not more than 1%.[2]

Department of Orthopaedics, S. D. M. College of Medical Sciences and Hospital, Sattur, Dharwad, [1]Department of Orthopaedics, Karnataka Institute of Medical Sciences and Hospital, Hubli, Karnataka, India

Address for correspondence: Dr. Anshu Shekhar, Department of Orthopaedics, S. D. M. College of Medical Sciences and Hospital, Sattur, Dharwad - 580 009, Karnataka, India. E-mail: dr.anshushekhar@gmail.com

Access this article online	
Quick Response Code:	Website: www.ijoonline.com
	DOI: 10.4103/0019-5413.156229

Case Report

A previously healthy 10-year-old girl presented with a large swelling arising from the right little finger of 6 months duration. The swelling was initially small and painless to start with and was associated with fever and chills. She was seen at a local hospital, where a radiograph was taken which revealed a cystic and lytic lesion of the little finger proximal phalanx [Figure 1]. It was diagnosed as osteomyelitis, and she was treated with analgesics and antibiotics. As the swelling and pain progressed, she was referred to this institute. The swelling was 6 × 6 cm, tender, pulsatile and firm in consistency. The fresh radiograph showed a large soft tissue mass with some calcification within and complete destruction of the phalanx [Figure 2a and b]. Magnetic resonance imaging (MRI) scan of the hand revealed an ill defined expansible lesion in the proximal phalanx with marrow signal changes that were T1 hypointense and T2/Short tau inversion recovery hyperintense with juxtacortical soft tissue mass [Figure 3]. Fine needle aspiration cytology (FNAC) of the lesion was performed and reported as having features of "small blue round cell tumor." Computed tomography (CT) of the chest, abdomen, and pelvis was normal. Axillary lymph nodes were enlarged, and FNAC had features suggestive of reactive lymphadenitis. The patient underwent wide excision with fifth carpometacarpal joint disarticulation. Histopathology of excised tissue showed small round cells with high nuclear-cytoplasmic ratio and coarse chromatin with occasional pseudorosettes, suggestive of Ewing's sarcoma. The cells were periodic acid

Schiff positive and immunohistochemistry was positive for CD99 [Figure 4a and b].

The patient was discharged and advised to review after 3 weeks for chemotherapy. However, the patient was non-compliant and returned about 4 months after the surgery with pain and swelling of the dorsum of the hand with dilated veins [Figure 5a]. There was a history of fever. Blood investigations revealed hemoglobin of 9 g% and erythrocyte sedimentation rate (ESR) 42 mm at 1 h. Radiograph revealed cystic lesion in the third metacarpal diaphysis with sclerosis and minimal periosteal reaction [Figure 5b]. MRI scan showed marrow signal changes involving second, third and fourth metacarpal bones with cortical erosion and breakdown of third metacarpal bone [Figure 6]. CT scan of the chest revealed bilateral multiple metastatic deposits. Tc 99 bone scan did not reveal any other site of metastasis. The patient underwent a transradial amputation about 5 cm. proximal to the wrist joint. Histopathology and immunohistochemistry of the excised specimen confirmed it as Ewing's sarcoma. Postoperatively, chemotherapy was started using cyclophosphamide, vincristine, actinomycin, and doxorubicin. The patient is surviving at 24 months of follow-up. The chest metastasis has partially resolved, and there is no local recurrence or evidence of metastasis to other sites.

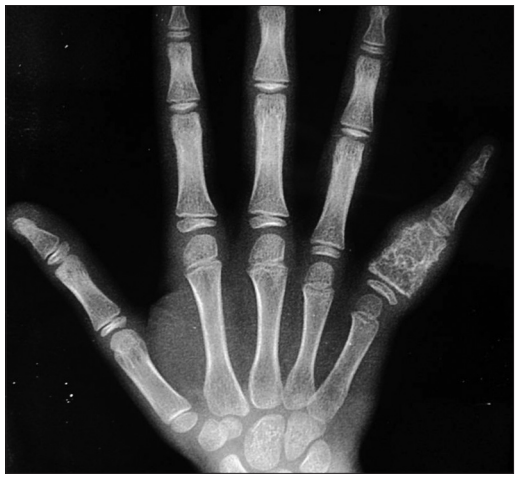

Figure 1: Initial radiograph of hand anteroposterior view showing a cystic, lytic lesion with minimal periosteal reaction in the proximal phalanx

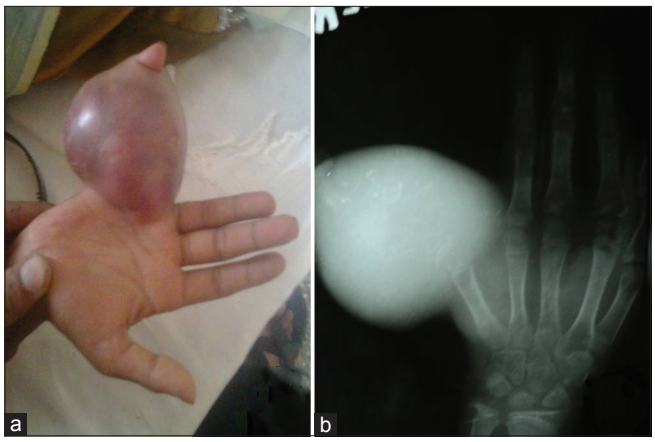

Figure 2: (a) Clinical photograph of hand showing swelling at the time of presentation to us about the size of a golf ball (b). Radiograph of hand anteroposterior view showing destruction of the entire phalanx with large soft tissue component

Discussion

Ewing's sarcoma is the second most common malignant primary bone tumor of childhood and adolescence.[3] Bernstein *et al*., reporting 1426 patients from the European Intergroup Cooperative Ewing Sarcoma Study, recorded only 1% of primary Ewing's sarcomas in the hand.[4] Case reports indicate that within the hand, the metacarpal is the most common locale.[5,6] Ewing's sarcoma/primitive neuroectodermal tumor classically presents mimicking an acute infection with pain, swelling, and erythema in the affected area with associated leukocytosis and raised ESR.[7] Siddiqui *et al*. reported a case of calcaneal Ewing's sarcoma with skip metastasis to adjacent tarsal bones.[1] To the best of our knowledge, this is the first reported case of

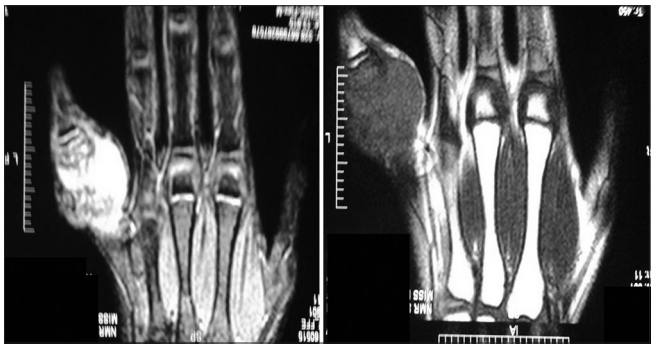

Figure 3: Magnetic resonance imaging in T1 and T2/soft tissue inflammatory response sequences showing the extent of the lesion in the little finger

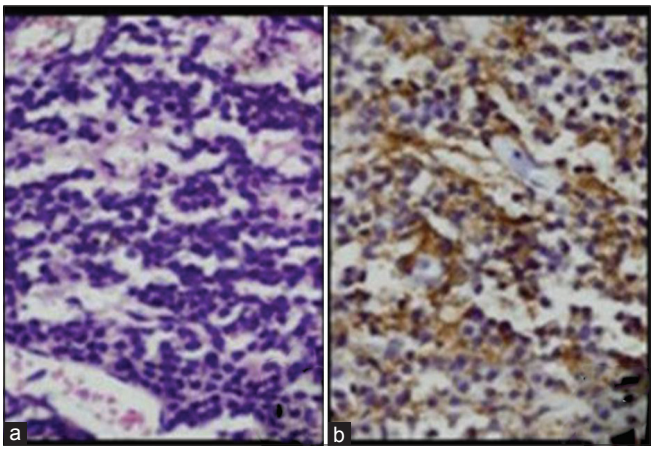

Figure 4: Periodic acid Schiff positivity of the cells seen (a) and Immunohistochemistry showing positivity with CD99 (b)

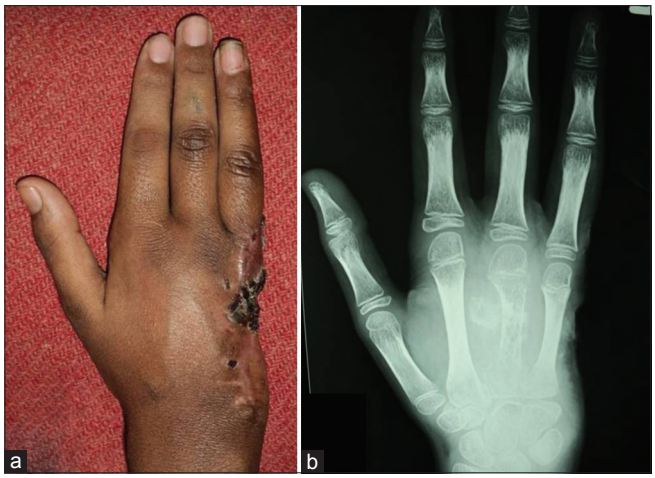

Figure 5: (a) Clinical photograph at followup after 3 months shows discernible fullness over dorsum of the hand. (b) Radiograph of hand anteroposterior view showing destruction of the third metacarpal with little periosteal reaction

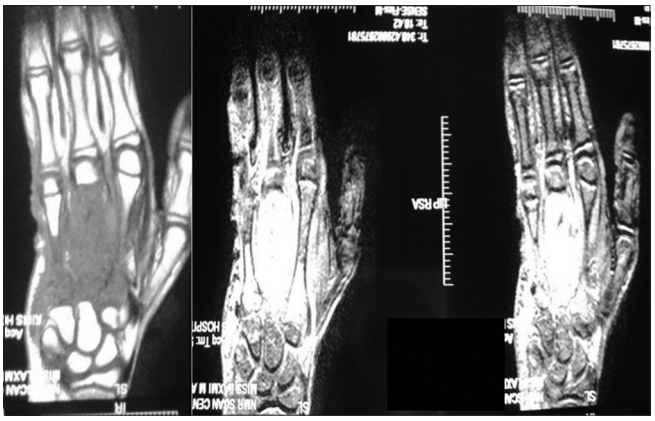

Figure 6: Magnetic resonance imaging revealing marrow signal changes involving the second, third and fourth metacarpals with cortical breakdown of third metacarpal in T1 and T2 sequences

Ewing's sarcoma arising from phalanx of the hand with skip metastases to multiple metacarpals.

Investigative modalities done as a routine for "tumor work up" includes a radiograph of the part, MRI scan, complete blood work up, chest X-ray, CT/ultrasonography of abdomen and Tc 99 bone scan.[7] ^{18}F-Fluorodeoxyglucose positron emission tomography-CT scan is a newer and excellent modality to detect unsuspected or unusual metastatic sites, monitor response to chemo or radiotherapy and to detect recurrence in primary skeletal Ewing's sarcoma.[8,9] Reinus *et al*. when studying the radiographic appearance of Ewing's sarcoma of hands and feet, found that the most common feature was that of a permeative bone lesion with poorly defined margins and an associated soft tissue mass.[10] Even in our patient, the early X-rays revealed a cystic and lytic lesion with large soft tissue mass with some sclerosis and negligible periosteal reaction. Knowledge of the often atypical radiological appearance of Ewing's sarcoma of the hand is important in ensuring a prompt and accurate diagnosis.[3] The differential diagnosis of this lesion includes osteomyelitis, spina ventosa, enchondroma, coccidioidomycosis, bone infarction and hand foot syndrome of sickle cell disease.[3] The prognosis does not appear to be related to the radiographic appearance or dimension of the primary lesion but to the presence of known metastasis at the time of presentation.[10]

Management, preferably at a specialist center by a multi disciplinary team, has included both local control, by either surgery, radiation or a combination of these, and systemic chemotherapy.[5] With the advent of modern chemotherapy, the long term, 5-year survival rate has improved to approximately 70%.[11,12] With small soft tissue and bone sarcomas in the extremities, local control by surgery has been better than with primary radiotherapy. Patients with hand lesions are recorded to have survived for more than 41 months and European Intergroup Study data showed a 68% overall 3 year survival rates in patients with distal extremity lesions.[5] Because Ewing's sarcoma and related tumors occur so rarely in the hand, there are no standardized treatment recommendations. Based on current evidence, however, chemotherapy and wide local tumor resection should be considered the mainstays of treatment.[6]

The differential diagnosis of unusual finger swellings in children and the atypical radiographic picture this tumor should have high index of suspicion of Ewing's sarcoma. It is important to bear in mind that the presence of metastases is the most important prognostic factor and thus the need for accurate early diagnosis.

References

1. Siddiqui YS, Zahid M, Sabir AB, Asif N, Kumar G, Akhtar M. Calcaneal Ewing's sarcoma with skip metastases to the adjacent tarsal bones. J Clin Diagn Res 2011;5:117-9.
2. Mohan V, Gupta RP. Ewing's sarcoma of the small bones of hands and feet. J Radiol Imaging 2002;12:403-5.
3. Escobedo EM, Bjokengren AG, Moore SG. Ewing's sarcoma of the hand. Am J Radiol 1993;159:101-2.
4. Bernstein M, Kovar H, Paulussen M, Randall RL, Schuck A, Teot LA, *et al.* Ewing's sarcoma family of tumors: Current management. Oncologist 2006;11:503-19.
5. Jose Jerome JT, Varghese M, Thirumagal SB. Ewing's sarcoma of the distal phalanx of little finger. Internet J Oncol 2007;4
6. Sforzo CR, Scarborough MT, Wright TW. Bone-forming tumors of the upper extremity and Ewing's sarcoma. Hand Clin 2004;20:303-15, vi.
7. Thomas BP, Sasi K, Pallapati SC, Mathew A, Sreekanth R, Thomas M. Malignant tumours of the hand and wrist. Indian J Plast Surg 2011;44:337-47.

8. Sharma P, Khangembam BC, Suman KC, Singh H, Rastogi S, Khan SA, *et al.* Diagnostic accuracy of 18F-FDG PET/CT for detecting recurrence in patients with primary skeletal Ewing sarcoma. Eur J Nucl Med Mol Imaging 2013;40:1036-43.
9. McCarville MB, Christie R, Daw NC, Spunt SL, Kaste SC. PET/CT in the evaluation of childhood sarcomas. AJR Am J Roentgenol 2005;184:1293-304.
10. Reinus WR, Gilula LA, Shirley SK, Askin FB, Siegal GP. Radiographic appearance of Ewing sarcoma of the hands and feet: Report from the intergroup Ewing sarcoma study. AJR Am J Roentgenol 1985;144:331-6.
11. Wunder JS, Paulian G, Huvos AG, Heller G, Meyers PA, Healey JH. The histological response to chemotherapy as a predictor of the oncological outcome of operative treatment of Ewing sarcoma. J Bone Joint Surg Am 1998;80:1020-33.
12. Burgert EO Jr, Nesbit ME, Garnsey LA, Gehan EA, Herrmann J, Vietti TJ, *et al.* Multimodal therapy for the management of nonpelvic, localized Ewing's sarcoma of bone: Intergroup study IESS-II. J Clin Oncol 1990;8:1514-24.

How to cite this article: Shekhar A, Korlhalli S, Murgod G. Ewing's sarcoma of proximal phalanx of the hand with skip metastases to metacarpals. Indian J Orthop 2015;49:365-8.

Source of Support: Nil, **Conflict of Interest:** None.