



## Ewing's Sarcoma of Hand - A Rare Case Report

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### Abstract

*Ewing's sarcoma of the hand is a rare entity which may be missed unless a high degree of suspicion is maintained. A case of a 19 year old female is presented who came with complaints of swelling involving her left hand. It was associated with ulceration and foul smelling discharge. Patient was from a rural set up hence initially her condition may have been neglected resulting in massive growth of the tumour causing physical & social morbidity. Biopsy of this swelling was suggestive of malignant small round cell tumour. Immunohistochemistry was CD 99 positive. After appropriate work up patient underwent left below elbow amputation. Histopathological examination confirmed Ewing's sarcoma arising from the metacarpals of the left hand. Post-operative course was uneventful. Patient refused to undergo adjuvant chemoradiation therapy. Follow up of 9 months has shown her to be disease & symptom free. A brief case report with review of literature is presented.*

**Keywords:** *Ewings sarcoma, hand, adolescent female.*

### Introduction

Ewing's Sarcoma is the second most common primary bone tumour in childhood and adolescence<sup>[1]</sup>. It usually affects the diaphysis of the long bones and flat bones of the pelvis. It is exceptionally rare in the small bones of the hands and feet where incidence is no more than 1%.<sup>[2]</sup>. Initially thought to be derived from primitive neuroectodermal cells, many now believe it to arise from mesenchymal stem cells<sup>[3]</sup>. We hereby present a rare case of Ewing's sarcoma of left hand in a 19 years old female. She was hailing from a rural place and had neglected her condition

initially resulting in a huge tumour of left hand with ulceration & foul smelling discharge. She was investigated and treated with surgical intervention. Present case highlights the ignorance and paucity of medical care in rural places resulting in delayed presentation of such patients to tertiary centres.

### Case Report

A 19 years old female came with complaints of pain and swelling involving her left hand since 8 months. Patient was apparently alright 8 months back when she noticed a swelling over the dorsum

of her left hand. Swelling was insidious in onset, initially painless and small in size, gradually progressed over the course of 8 months to involve the entire left hand. She complained of increasing blackish discolouration over the swelling over the past 2 months associated with yellowish foul smelling discharge from multiple points of ulcerations over the swelling. Patient also gave history of increasing pain associated with the swelling over the past two months affecting her range of movement at all the fingers of her left hand and left wrist joint, hindering her daily activities. She also complained of fever, intermittently over the last two months which got relieved by anti-pyretic medication. Patient gave h/o loosening of clothes over the past 8 months suggestive of weight loss. There was no history of similar swelling elsewhere in the body, chest pain, breathlessness or trauma. Patient used to cover her left hand in cloth and restrict herself within home to avoid social contact as her swelling was huge with foul smelling discharge. This social morbidity may have resulted in her delayed presentation to a tertiary centre.

On clinical examination, patient had pallor and was found to be severely anaemic on investigation (Hemoglobin-6g/dL). Physical examination revealed a 20x20cm mass over the whole dorsum of left hand, with redness, tenderness and patches of blackish discolouration alongwith multiple foul smelling pus discharging sinuses (Fig. 1). The range of movement of the fingers of the left hand and flexion, extension at the wrist joint were absent.

Plain radiograph of the left hand (AP and oblique view) revealed a large, lobulated soft tissue mass arising from the second metacarpal bone which appeared to be completely destroyed (Fig. 2). Few calcifications in a spiculated pattern were seen within the centre of the mass. The second carpo-metacarpal joint was not well visualised and was involved. Non visualisation of the distal carpal row was suggestive of their involvement. Involvement of the medial four metacarpal joints

as well as the intercarpal and radiocarpal joints was evident.

Computed Tomography (CT) angiography of the left upper limb was done suggestive of a large exophytic, ill defined heterogeneously enhancing mass measuring 11.2x13x20cm arising from the diaphysis of the metacarpal of the left index finger (Fig. 3). It showed multiple non enhancing necrotic areas within. There was destruction of the diaphysis with a central stellate area of calcification. Multiple vessels were seen coursing through the mass. Medially it caused thinning of the cortex of the third metacarpal. The bulk of the mass was seen to be growing dorsally with no extension into the adjacent joints. Branches from the left radial artery and digital arteries of the left index finger were seen to be supplying the mass. Multiple superficial varicosities were noted throughout the left upper limb with prominence of basilic and cephalic veins. Few enlarged reactive lymph nodes were seen in the left axillary region largest measuring 3.4x4.1cm.

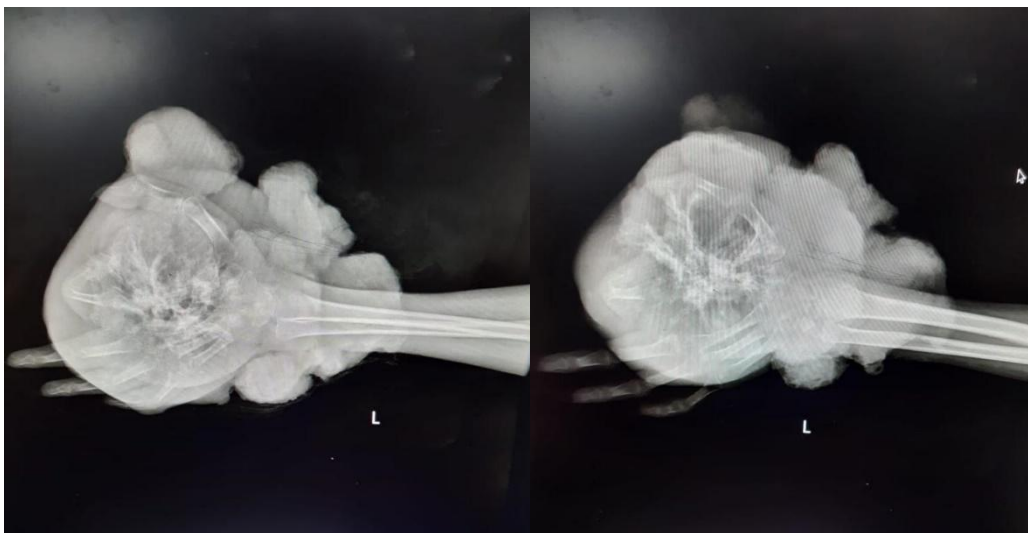
Biopsy of the swelling revealed malignant round cell tumour with immunohistochemistry suggestive of CD99 positive. The pathological diagnosis was Ewing's sarcoma. Left below elbow amputation was planned for this patient. She was given blood transfusion in view of anaemia prior to the surgery and haemoglobin was built upto 10g/dL. After optimising the patient pre-operatively, she then underwent left below elbow amputation (Fig. 4). Histopathological examination revealed a tumour arranged in groups, nests, sheets separated by fibrovascular septa (Fig 5). Tumour was seen to be invading the underlying bone. Individual tumour cells were monotonous with scant to moderate, eosinophilic to clear cytoplasm, round to oval nuclei and prominent nucleoli. Mitoses were noted 5-6/hpf. Areas of haemorrhage and necrosis with vascular emboli were seen. Resection margins of skin, bone and soft tissue were free of tumour. This confirmed our diagnosis of malignant small round cell tumour suggestive of Ewing's sarcoma. Post-operative course was uneventful. Patient was

advised PET scan and post-operative chemotherapy, however, patient refused further treatment and went to her native place. Patient did not follow up so telephonic consultation was done

with patient on which she sent her photograph (Fig. 6). She did not come personally for follow up & further evaluation.

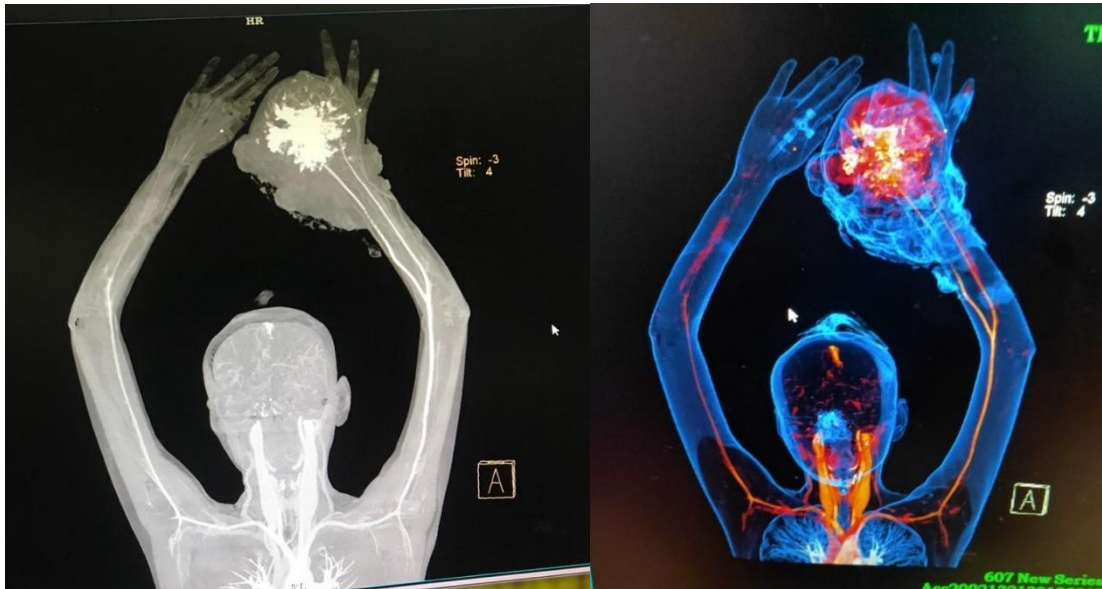


**Figure 1-** Ewing's tumour of left hand with ulcerations and blackish pigmentation



**Figure 2-** AP & Oblique X-Ray of left hand showing destruction of second metacarpal and spiculated calcification with destruction of surrounding tissues.

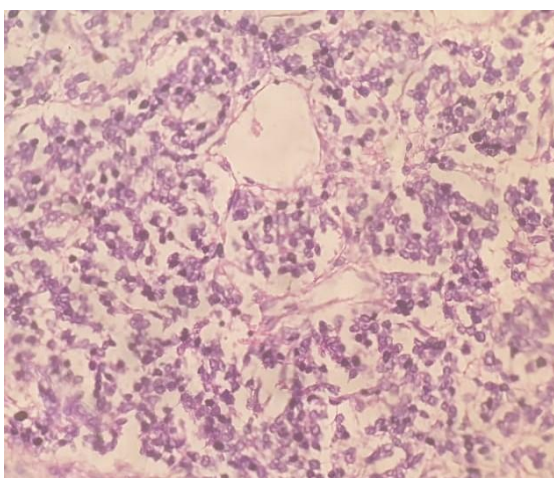




**Figure 3** CT Angiography of left upper limb showing tumour with increased vascularity.



**Figure 4** Amputated specimen of left hand containing tumour



**Figure 5** Histopathology slide showing sheets of small round malignant cells (hematoxylin and eosin stain 40 X)



**Figure 6** Amputation stump 9 months after surgery.

## Discussion

Ewing's sarcoma first described in 1921 and was initially thought to be derived from primitive neuroectodermal cells. Many now believe Ewing's sarcoma to arise from mesenchymal stem cells<sup>[3]</sup>. Genetic factors play a role in its pathogenesis. There is a form of anomaly with chromosomal translocation of two chromosomes 22 and 15<sup>[4]</sup>. It is the second most common primary bone tumour in childhood and adolescence<sup>[1]</sup>, after osteosarcoma. It usually affects the diaphysis of the long bones and flat bones of the pelvis and is exceptionally rare in the small bones of the hands and feet where incidence is no more than 1%<sup>[2]</sup>. Metacarpals and proximal phalanges are the most common locations in the hand<sup>[5]</sup>. It is usually seen in the first or second decade of life. Men (69%) are affected more than the women, with the average age of occurrence being 18.5 years (5 months–51 years)<sup>[3]</sup>. Present case was a 19 years old female with involvement of left hand which is rare.

Clinically, Ewing's sarcoma presents with fever, elevated erythrocyte sedimentation rate and signs of local inflammation. This may cause it to be confused with tubercular dactylitis<sup>[5]</sup>. Ewing's sarcoma often mimics osteomyelitis both clinically with signs of local inflammation and radiographically because of the cortical destruction and aggressive periosteal reaction<sup>[3,6]</sup>. Present case presented with swelling, pain, ulceration and foul smelling discharge because of long standing due to initial neglect as she was from a rural place. Patient besides physical morbidity had social morbidity as she remained at home due to unsightly tumour with foul smell.

X-ray and MRI of the primary site, chest CT and bone scintigraphy are crucial not only to differentiate the aforementioned lesions, but also to detect the site and size of the primary tumour and stage the tumour so as to plan the next line of management. An X-ray of Ewing's sarcoma will show typical destructive lesions with indistinct margins, onion-skin-type periosteal reactions and soft tissue masses in the diaphysis or, more rarely,

in the metaphyseal/diaphyseal region<sup>[7]</sup>. This can be differentiated from tubercular dactylitis which shows characteristic expansile lesion of the hand and absence of periosteal reaction on X-ray. Osteosarcoma found more commonly in the metaphysis, presents with an increase in alkaline phosphatase, not seen in Ewing's sarcoma. Present case showed typical destructive lesions.

MRI is an ideal method for evaluating soft tissues, the intramedullary involvement of the disease, and the involvement of the primary tumour in the surrounding soft tissues. Bone scintigraphy assists in detecting the bone involvement and, to an extent, the amount of intramedullary involvement. Conventional Tc99 bone scintigraphy is ideal for metastatic disease scanning<sup>[8]</sup>. Present case involved left hand and CT scan with angiography along with biopsy was diagnostic.

The treatment of Ewing's sarcoma is achieved through a Common European protocol called Euro Ewing 99<sup>[9]</sup>. The aim of surgical management is to achieve acceptable cosmesis and preserve as much functionality of the hand as possible. After the initial surgical management, the prognosis depends on the tumour volume and the histological response to VIDE induction chemotherapy (vincristine, ifosfamide, doxorubicin, etoposide). The tumours will be irradiated if the histological study reveals more than 10% of tumour cells after surgery and chemotherapy<sup>[10]</sup>. Localising the tumour on the hand is an important prognostic factor as the survival rate is much higher when the distal part of the extremities are affected. The best treatment combination is radical excision and postoperative chemotherapy.

Our patient underwent left below elbow amputation after discussion with patient. She was advised post-operative chemotherapy and was asked to follow up with PET-CT scan. She was also referred to the prosthetic department for prosthesis and rehabilitation. However, she refused further treatment and went to her native place. She was contacted telephonically. She refused to follow up personally, however, she sent

her amputation stump picture 9 months after surgery on phone.

### Conclusion

Ewing's sarcoma of the hand is a rare entity which may be missed unless a high degree of suspicion is maintained. Though no consensus regarding optimal management exists, a multidisciplinary approach using surgery, radiotherapy and chemotherapy is currently recommended. In rural India, owing to the lack of awareness, paucity of resources and social taboo, these patients tend to present late in the course of their illness leaving amputation as the last and only resort. Education at rural level and primary health care centres may help in early diagnosis and management of such cases.

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