



## Case Report

### EWING'S SARCOMA OF FIFTH METACARPAL IN WOMEN: AN UNUSUAL PRESENTATION

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#### Abstract

Ewing's sarcoma is the second most common highly malignant primary mesenchymal bone tumor of childhood and adolescence affecting mainly the diaphysis of long 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 scanty periosteal reaction. Prognosis 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 5th metacarpal bone in 40 years old lady which was initially missed and so there was delayed onset of treatment.

**Keywords:** Ewing's sarcoma (ES), Metacarpal, Tumour.

#### 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<sup>1</sup>. It is relatively uncommon in the small bones of hands and feet with 1% incidence<sup>2</sup>.

#### Case Report

A 40 years old lady presented with swelling in the dorsolateral aspect of right hand for one year. She complains of pain for the last one month. She was a diagnosed case of pulmonary tuberculosis & completed anti TB treatment. She had no history of blood transfusion & no history of cough, hemoptysis, jaundice, hematemesis, melaena & any other bone pain. No other of her family members suffered from TB, malignancy & any other chronic diseases

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On local examination, a globular swelling in the dorso-lateral aspect of the right hand measuring about 2x3cm, small scar mark present, no venous engorgement & ulceration. Temperature normal & tenderness present. Margin well defined, hard in consistency & fixed with the underlying structure but not overlying skin. There was no restricted movement of metacarpophalangeal and wrist joint. No other neural & lympho-vascular abnormality was present.

Routine hematological & biochemical examinations were within normal limits. Chest X-Ray P/A view, USG of whole abdomen was normal. The early X-rays of local part revealed a cystic and lytic lesion with large soft tissue mass with some sclerosis and negligible periosteal reaction in 5th metacarpal (right) bone. The patient was subjected to do FNAC of the swelling & diagnosis revealed the lump as tubercular osteitis first time and patient started anti tubercular drugs for bone TB for three months. Due to no response of anti TB drugs, repeated FNAC revealed Ewing's sarcoma second time. Then patient was sent for open biopsy & histopathology and it revealed poorly differentiated

fibrosarcoma. Patient was referred to oncologist and was advised for radiotherapy followed by chemotherapy. After completing concurrent chemoradiation, ray amputation was done. Histopathological report of the specimen shows features of Ewing's sarcoma. The surgical margins were negative. Patient was advised for adjuvant chemotherapy. Patient received chemotherapy but after 2 years there was distal metastasis in right humerus. Patient survived two & half years after diagnosis of Ewing's sarcoma & died due to metastatic complications.



## Discussion

Ewing's sarcoma is a mesenchymal cell tumor<sup>3</sup>. It is usually seen in the first or second decade of life, generally affecting long tubular bones and pelvis<sup>4</sup>. It is very rarely seen in the bones of the hand. Less than 1% of the cases diagnosed with ES are localized in the hand and the wrist. The most common locations in the hand are the metacarpal and proximal phalanges<sup>5</sup>. The thumb (28%) and the middle finger (28%) are most commonly affected<sup>3</sup>. Men (69%) are affected more than the women, with the average age of occurrence being 18.5 years (5 months–51 years). ES in the hand has a generally better prognosis compared to other sites<sup>3</sup>. Survival rate is highest in lesions involving distal bones of the extremities<sup>6</sup>. Pain and swelling are the most common complaints of the affected finger<sup>4</sup>. At the beginning of the clinical presentation, the patient's general health condition remains good with no fever or weight loss being present<sup>7</sup>. In the current case, the patient was in good health, with the only complaints being pain and swelling of the hand.

Investigative modalities should be 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<sup>8</sup>. 18F-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<sup>9</sup>. 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<sup>10</sup>. 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 atypical radiological appearance of Ewing's sarcoma of the hand is important in ensuring a prompt and accurate diagnosis<sup>11</sup>.

ES can be easily confused with acute infection of bone presenting with pain, swelling, localized tenderness with increased total leukocyte count and raised ESR<sup>8</sup>. ES can also present with atypical radiological appearance in hand involvement mimicking enchondroma, osteomyelitis, spinaventosa, bone infarction and sickle cell disease<sup>11</sup>.

Management, preferably at a specialist center by a multidisciplinary team, has included both local control, by either surgery, radiation or a combination of these, and systemic chemotherapy<sup>12</sup>. With the advent of modern chemotherapy, the long term, 5-year survival rate has improved to approximately 70%<sup>13</sup>. 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<sup>12</sup>. Because Ewing's sarcoma and related tumors occur so rarely in the hand, there are no standardized treatment recommendations. Currently, the most favorable treatment of ES of the small bones of extremities is radical excision followed by chemotherapy<sup>3</sup>. This treatment approach is successful in tumors of the fingers. Radiotherapy is useful when radical surgical excision is not performed and in partial response to chemotherapy. Neoadjuvant chemotherapy plays an important role to control local disease, skip metastasis and response of tumour to drugs.

### Conclusion

Lack of recognition of this condition might be secondary to the absence of features traditionally associated with malignant bone neoplasms. A broader differential diagnosis after intervention failures offers the opportunity for diagnosis and appropriate treatment. In spite of its multifaceted presentation, Ewing's tumor is an interesting variety of malignant tumor. Every case should be assessed and evaluated thoroughly. It is important to bear in mind that early recognition of an unusual appearance and location of Ewing's sarcoma is necessary for its adequate treatment.

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