# Ewing's sarcoma humerus presenting along with metastatic breast nodule: An unusual case report

Sunita Singh, Shilpa Garg, Monika Sangwan, Jyoti Sharma, Rajeev Goyal<sup>1</sup>

Departments of Pathology, 'Orthopedics, Post Graduate Institute of Medical Sciences, Rohtak, Haryana, India

### **ABSTRACT**

Ewing's sarcoma is a highly malignant primary bone tumor and most frequently observed in children and adolescents but rare in adults. Usual sites of metastasis are lung, pleura, other bones and occasionally to lymph node, central nervous system, and liver. Metastasis into breast is extremely rare and only one case has been reported. Most common tumors metastasizing into breast are lymphoma, malignant melanoma, and rhabdomyosarcoma. A 25-year-old female presented simultaneously with both left arm swelling and breast lump. Fine needle aspiration done from both swellings revealed similar morphology and comprised monomorphic population of round cells dispersed singly with scant cytoplasm and reticulogranular chromatin. IHC done on cell block material was positive for CD99. Hence diagnosis of Ewing's sarcoma metastasizing to breast was made. Hereby, we present a rare case of Ewing's sarcoma humerus with simultaneous metastatic deposit in breast as the initial presentation.

Key words: CD99, cell block, Ewing's sarcoma, metastasis breast

# INTRODUCTION

Ewing's sarcoma is a rare malignant round cell tumor of the bone. It is the second-most common primary malignant tumor of bone found in children. It commonly affects long bones, pelvis, and ribs.[1] It can also arise in soft tissue (extraskeletal).[2] The common sites to which Ewing's sarcoma metastasize are the lung, pleura, and other bones. The metastasis of Ewing's sarcoma to the breast is very rare and only a few cases have been reported in the literature. [3] Metastasis to the breast from other organs are uncommon, the prevalence of such lesions ranges from 1.7 to 6.6%.[4] Lymphoma, malignant melanoma, rhabdomyosarcoma, and lung carcinoma are the common tumors that metastasize into breast. Sarcomas are extremely rare tumor sources of breast metastases and metastasis of Ewing's sarcoma is further rarest. [3] Here we report a case of Ewing's sarcoma humerus presented as breast metastasis which was clinically and radiologically

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misdiagnosed as carcinoma breast metastasizing to humerus.

## **CASE REPORT**

A 25-year-old female presented with a diffuse swelling in left arm since 3 months and she noticed lump left breast since 2 months. On local examination swelling arm was ill-defined diffuse involving the medial aspect of arm and was firm, warm, and tender to touch. Breast lump was 3 × 3 cm irregular, firm and mobile on palpation involving the upper outer quadrant of left breast with axillary lymph node enlargement. X-ray and sonographic examination of arm revealed soft tissue swelling. Sonography and mammography of breast suggested a malignant mass. Hence clinically as well as radiographically diagnosis of carcinoma breast metastasizing to arm was made and MRI and fine needle aspiration (FNA) were advised. MRI revealed a lobulated enhancing soft tissue mass with irregular margins involving upper and outer quadrant of left breast [Figure 1]. Diaphysis of left humerus revealed altered signal intensity with soft tissue mass which was infiltrating into surrounding muscles. Multiple vertebral bodies also revealed altered bone marrow signal and collapse. MRI findings confirmed the possibility of carcinoma breast with metastasis in left arm. FNA was performed from both the sites. FNAC smears from both the swellings revealed similar morphology and showed presence of monomorphic population of round cells with

Address for correspondence: Dr. jyoti sharma, Department of pathology, DH-98, Phase 1, Pallavpuram, Meerut (U.P), India. E-mail: drjyoti1212@yahoo.in

scant cytoplasm and reticulogranular nuclear chromatin and inconspicuous nucleoli [Figures 2-4]. Cytologically diagnosis of malignant small round cell tumor was made and two possibilities of Ewing's sarcoma or Non-Hodgkin's lymphoma of bone metastasizing to breast were kept. On cytochemistry the tumor cells were diastase-sensitive PAS positive. Subsequently, immunohistochemistry was performed on the cell block prepared and tumor cells were positive for CD99 and negative for LCA, CK, EMA, S100, Synaptophysin, Chromogranin, and Desmin. Final diagnosis of Ewing's sarcoma of left humerus metastasizing to breast was made.

# **DISCUSSION**

Ewing's sarcoma is an uncommon primary malignant round cell tumor of bone named after Ewing who first described it in 1921.<sup>[1]</sup> Now ES, PNET, and Askin tumor are altogether referred as Ewing's tumor. These tumor shares a common cytogenetic and 85% cases are positive for balanced translocation t (11:22) (q24:12).<sup>[5]</sup>

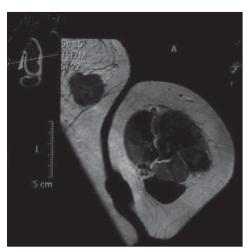


Figure 1: MRI with axial section sowing lesion in the arm as well as breast

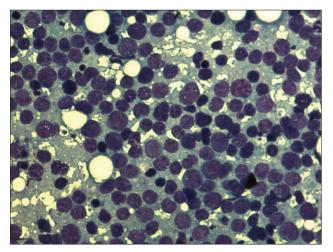


Figure 3: 400× showing monomorphic cells in aspirate from arm swelling on Giemsa

Despite being the second-most common primary malignant bone tumor found in children between 10 and 15 years of age it is still a rare tumor with an annual incidence of 0.8/million of population.<sup>[1]</sup> Approximately 80% of patients are younger than 20 years of age. Boys are affected slightly more frequently than girls.<sup>[5]</sup> Ewing's sarcoma can originate from any bone in the body, the most common sites are pelvis, thigh lower leg, upper arm, and ribs.<sup>[3]</sup> Most of the patients of Ewing's sarcoma present with pain and swelling at local site. Other symptoms include fever, weight loss, cough, anemia, and leukocytosis.<sup>[5]</sup> So clinically, they are often misdiagnosed and treated as osteomyelitis.<sup>[1]</sup> Along with above symptoms our patient presented with metastasis to breast and vertebrae at first visit which is very rare.

Ewing's sarcoma usually metastasizes to lung, pleura, and other bones. Lymph nodes, CNS, and liver may be occasionally involved. The metastasis of Ewing's sarcoma into breast tissue is extremely rare with only one case report being reported in literature. Metastatic cancer in the breast is often discovered as a superficial solitary mass (85%) in

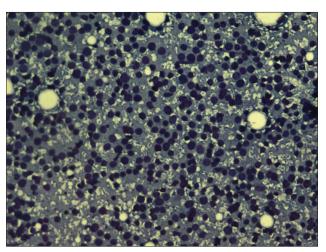


Figure 2: 200× showing monomorphic cells in aspirate from breast lump on Giemsa

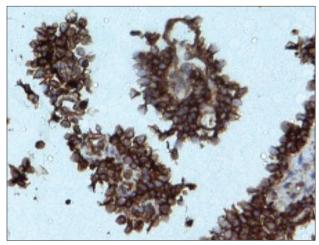


Figure 4: CD99-positive tumor cells on cell block material

the upper outer quadrant (66%). These metastases manifest clinically as well-defined, mobile, and rapidly enlarging masses. [6-7] Lymphoma/leukemia, malignant melanoma, lung carcinoma, carcinoma originating from the stomach, prostate, ovaries, kidneys, cervix, mouth, thyroid, are the common tumors that metastasize into breast tissue. Sarcomas rarely metastasize to breast. However, a few case reports are available revealing metastasis from fibrosarcoma and myofibrosarcoma to breast. [1]

Plain radiograph usually revealed a destructive lytic tumor that has permeative margins and extend into surrounding soft tissue. MRI is the imaging modality of choice because of its high-contrast resolution and ability to define the margins of soft tissue component especially on T2-weighted images. [5] Cytologically features are similar to the diagnosis of malignant small round cell tumor.[1] Diagnosis is usually supported by immunohistochemistry either on cell block material or on biopsy. Tumor cells are positive for CD99/MIC-2.[1] In our case, the MRI findings showed similar findings and cytological findings revealed a cellular lesion of monomorphic round cells with scant cytoplasm and reticulogranular chromatin. On Immunohistochemistry tumor cells were positive for CD99, and negative for LCA, CK, EMA, S100, Synaptophysin, Chromogranin, and Desmin to rule out possibility of other malignant small round cell tumor.

The immunohistochemical staining for CD99/MIC-2 suggested the diagnosis but is not pathognomic.

Ewing's sarcoma is a highly malignant bone tumor predominantly arising in young adults and an extremely rare cause of breast metastasis. Metastatic disease should be taken into consideration in the differential diagnosis of tumors of the breast, especially when there is history of extramammary neoplasm. MRI is the imaging modality of choice for local tumor evaluation. FNA along with immunohistochemical analysis is a useful approach for diagnosis. However, highlighting the specific transcript t (11:22) is necessary to confirm the diagnosis. The rarity of this condition prompted us to report this case. So such a possibility should always be kept in mind if a young patient presents with a soft tissue swelling and a breast lump simultaneously.

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