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# Breast Ewing Sarcoma/Primitive Neuroectodermal Tumor: A Case Report and a Review of the Literature

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# Breast Ewing Sarcoma/Primitive Neuroectodermal Tumor: A Case Report and a Review of the Literature

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## Abstract

Extraskeletal Ewing's Sarcoma is a rare soft tissue tumor that is morphologically indistinguishable from the more common Ewing's sarcoma of the bone. Primitive Ewing's sarcoma of the breast is exceptionally reported. We present the case of a 26-year-old woman with Ewing sarcoma/PNET diagnosed by molecular biology showing the specific transcript of Ewing/ peripheral primitive neuroectodermal tumor (pPNET). A definitive diagnosis of primary Ewing sarcoma/PNET of the breast was made following a negative staging evaluation.

## Introduction

Ewing sarcoma /primitive neuroectodermal tumor (PNET) is a morphological and immunohistochemical spectrum ranging from small monomorphic round cell tumor to "atypical" forms large cell or fusiform cells. It's characterized by a specific translocation and recurrent, often involving genes EWS and FLI-1. Although breasts may be affected by metastasis, achieving primitive breast is extremely rare. We report the case of a young woman with a primitive Ewing sarcoma/PNET of the breast

## Case Report(s)

We report the case of 26 years old women presented a mass of her right breast. She complained of rapidly growing in size. The Physical examination revealed an indurated mass of 12 in larger diameter. Sonography and mammography suggested a malignant mass of the right breast. Biopsy of the lesion was performed and suggested rich glycogen carcinoma of the breast. Right mastectomy was performed outside our institution. Macroscopically, the right breast measuring 19x14x10 cm. a large tumor was identified measuring 12x10x10 cm. This lesion was well circumscribed white with necrotic areas. Microscopic examination suggested always a rich glycogen carcinoma (PAS-positif on tumor cells) infiltrating all breast tissue without lymph node metastasis. Two months later, the patient presented local recurrence. Physical examination revealed a mass of 05 cm in larger diameter of the scar adherent to the wall with inflammatory signs. Ultrasonography revealed a heterogeneous hypoechoic mass measuring about 32x23 mm (figure 1). The patient underwent a local excision taking the pectoral muscle. Our laboratory has received a resection specimen measuring 20x16x8cm. The microscopic examination revealed sheets of monomorphic small to medium cells, with round nucleus. This proliferation was covered with bands of fibrovascular tissue (figure.2). On immunohistochemistry, the tumor cells were strongly positive for mic 2 (CD99), (figure 3), neuron specific enolase the (NSE). They were negative for cytokeratin AE1/AE3, EMA, S100 protein, synaptophysin, desmin and CD34. RT-PCR found a tumor specific transcript family PNET / Ewing transcript EWS / FLI1.Extraskeletal Ewing's sarcoma of the breast was confirmed. The patient refused further therapy was lost sight of.

## Discussion

Ewing sarcoma /primitive neuroectodermal tumor (PNET) is characterized by a specific trans-location and recurrent, often involving genes EWS and FLI-1 [1-4]. In 1975, Angervall and Enzinger reported the first case of extraosseous Ewing's sarcoma. The most common sites are the chest wall, paravertebral region, and retroperitoneal space. However, few cases have been reported in the kidney, breast, gastrointestinal tract, prostate, endometrium, the adrenal glands, brain and lungs. The diagnosis of primary breast Ewing sarcoma was very difficult here because of the unusual location and atypical morphology. The morphological appearance of the tumor is variable, ranging from small round cell tumor forms large cell or fusiform cells. The classic appearance shows a typical malignant prolifera-tion densely cellular, composed by undifferentiated small round cells, [5,6]. The nuclei are vesicular, with one or two indistinct nucleoli. Periodic shiffer acid (PAS) staining reveals, in most cases,

intracytoplasmic glycogen [5]. Immunohistochemical studies showed expression of vimentin, synaptophysin, NSE and PS100. The cytokeratin AE1/AE3 may be positive in 25% of cases. Marking with anti CD99 (Mic2) is non-specific, it can be seen in lymphoblastic lymphoma, the synovial sarcoma, osteosarcoma, small cell rhabdomyosarcoma and small cell carcinoma [7]. The differential diagnosis includes many undifferentiated tumors. In our case it was necessary to eliminate phyllode sarcoma, undifferentiated carcinoma, a synovial sarcoma and of course distant metastasis. Our patient had a tumor depends on the mammary gland and the clinical and radiological staging was not objectified another bone lesions or soft tissue or the chest wall. The cytogenetic study shows, regardless of the location, a translocation t (11, 22) (q24; q12) resulting in the formation of a chimeric fusion transcript between the EWS gene on chromo-some 22 and on FLI-1 chromosome 11 [8]. This translocation is found in more than 90% of cases. More rarely, other types of translocations can always see between chromosome 22 and other chromosomes 21, 7 and 17. [9] The cytogenetic study was done by the method of RT-PCR (reverse transcriptase polymerase chain reaction) on frozen tissue or by fluorescent in situ hybridization (FISH) can be performed on paraffin sections [9, 10]. The treatment is based on surgery coupled with anthracyclines chemotherapy with radiothera-py. Local recurrences are frequent and the prognosis is poor. [11] Our patient had a relapse after two months and was lost sight of before the start of adjuvant treatment.

# Conclusion

the primary breast Ewing tumors are very rare. The rarity of this condition prompted us to report this case. The immunohistochemical staining suggests the diagnosis but is not pathognomonic. Highlighting the specific transcript is necessary to confirm the diagnosis.

# Abbreviations(s)

PNET : peripheral primitive neuroectodermal tumor RT-PCR : reverse transcriptase polymerase chain reaction FISH: fluorescent in situ hybridization PAS: priodic shiffer acid

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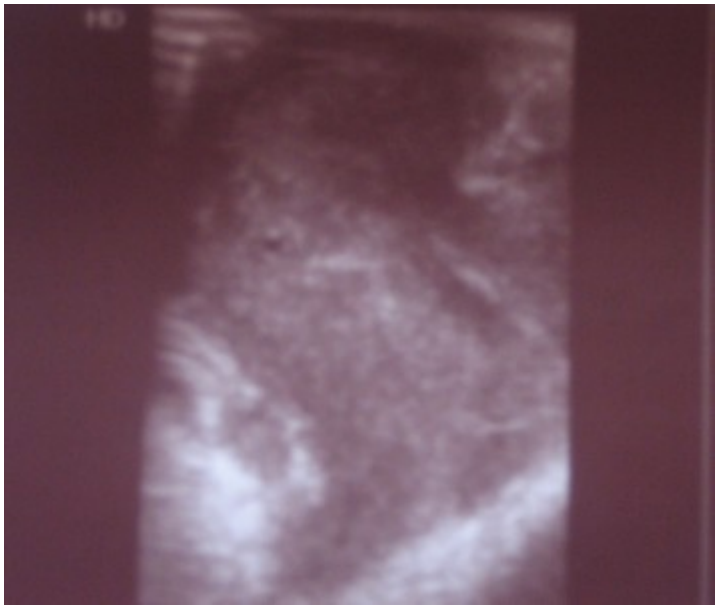
# Illustrations

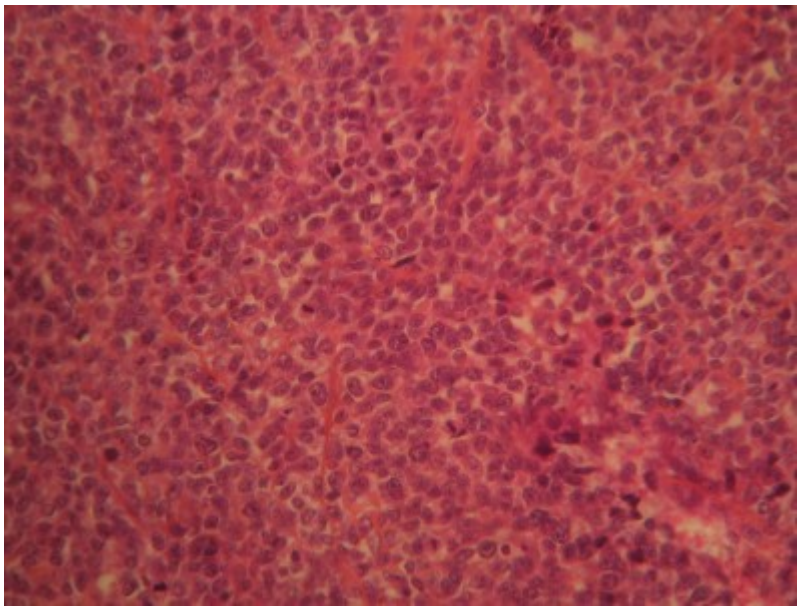
## Illustration 1

Illustration 1: Ultrasound image showing a large heterogeneous hypoechoic mass, measuring about 32x23 mm



## Illustration 2

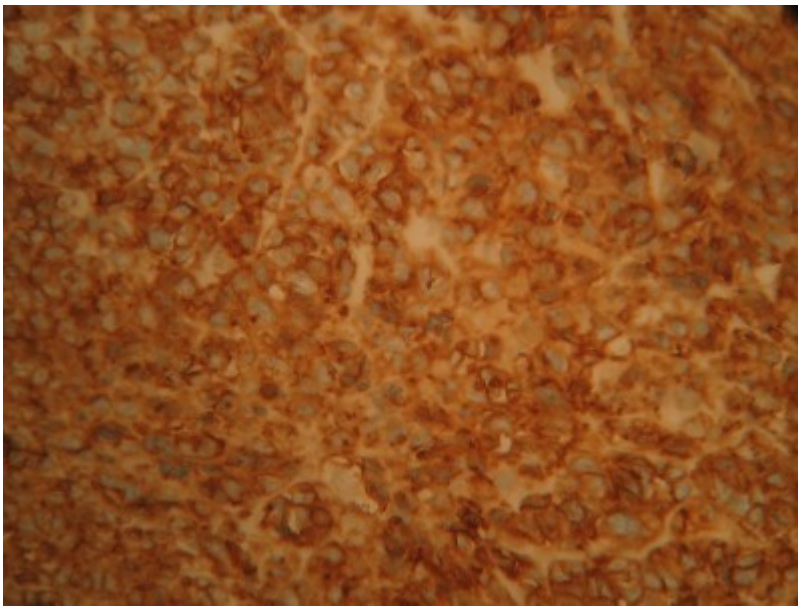
microphotography showing round cell tumor with atypical mitosis (hematoxylin-eosin at magnificationx20).



# Illustration 3

microphotograph showing positive staining of tumor cells for CD99 (Avidin Biotin at magnification x 40).



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