

An unusual breast tumor – A case report

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Abstract: Extraskeletal Ewing's Sarcoma/Primitive Neuroectodermal Tumor (EWS/PNET) presenting as a breast mass is uncommon. EWS/PNET of the breast may impose a diagnostic challenge, mammography and ultrasonography features are nonspecific. The histopathological pattern is variable depending on the degree of neuroectodermal differentiation. Immuno-phenotyping is necessary and genetic study is the only confirmatory tool. We report a case of a 29 year old lady who developed a painless breast swelling during pregnancy and diagnosed to have ewing's sarcoma of the breast.

Keywords: Ewing sarcoma, Extrasosseous, Breast.

INTRODUCTION

Ewing sarcoma (ES) is a highly malignant tumor of long bones occurring in children and young adults and was first described by James Ewing in 1921. Extrasosseous Ewing sarcoma (EES) was introduced by Tefft in 1969 [1]. EES is a rare, aggressive, malignant soft tissue tumor with high recurrence rate and mainly occurring in adolescents and young adults between 10 and 30 years of age. ES/primitive neuroectodermal tumor (PNET) presenting as a breast mass is uncommon, with only a few cases been reported in the literature [2]. We report the case of a young lady who presented with breast lump and was diagnosed as EES.

CASE REPORT

A 29 year old lady presented with history of painless swelling right breast since 3 months. The swelling first appeared during the 3rd trimester of pregnancy and gradually increased in size during the lactation. There was no history of fever, loss of appetite, loss of weight, bone pain or respiratory symptoms. She was shown in a local hospital and fine needle aspiration cytology of the breast swelling was suggestive of carcinoma breast. She presented to us following modified radical mastectomy (MRM).

On examination her general condition was good. She had post mastectomy status with no clinical evidence of metastatic disease. She had no organomegaly or lymphadenopathy and other systems were normal. Gross examination of the MRM specimen showed circumscribed partly encapsulated grey white lobulated mass lesion measuring 5 x 4 x 4 cm. Histopathological examination of MRM specimen showed tumor cells with scanty to moderate amount of

cytoplasm, round to oval nuclei with fine chromatin and infiltrating the adjacent fat (Figure 1). On immunohistochemistry (IHC) the tumor cells were positive for MIC-2 (CD99) and negative for Synaptophysin, CD68, LCA, Myogenin, Tdt, P63, desmin (Figure 2). The picture was diagnostic of ES of the breast. Her hemoglobin was 13.6 g/dl, total WBC count was 10800/mm³, platelet count was 317000 /mm³, and ESR was 23mm/ 1st hour. Her renal function, liver function and serum electrolytes were normal. Serum LDH was 522 IU/L. Bone marrow study and bone scan were normal. Computed tomography (CT) scan showed no evidence of any lung metastasis. Our patient had non metastatic disease and presented to us following MRM. She is currently undergoing combination chemotherapy with vincristine, Adriamycin, cyclophosphamide alternating with ifosfamide and etoposide (VAC-IE).

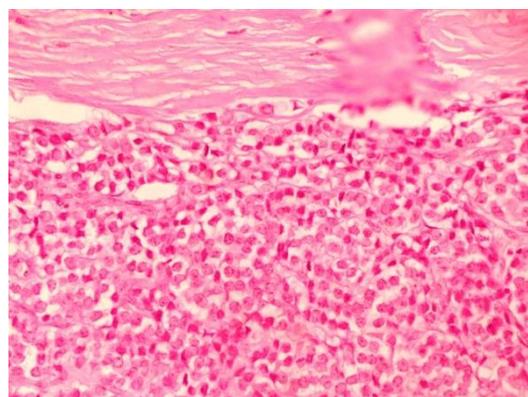


Fig-1: Neoplastic cells with scanty to moderate amount of pale, clear cytoplasm and round to oval nuclei with fine chromatin (H&E)

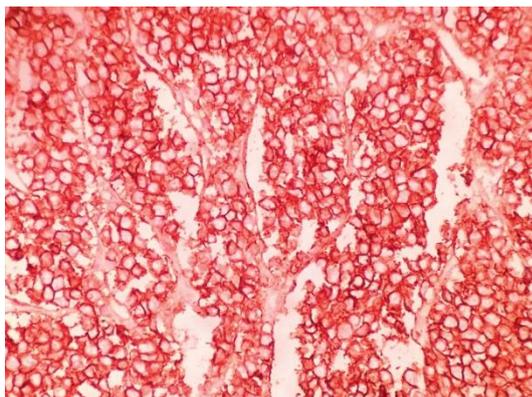


Fig-2: Neoplastic cells showing diffuse strong membrane positivity for CD 99

DISCUSSION

Carcinomas are the majority of malignancies involving the breast and sarcomas represent less than 1% of breast malignancies [3]. ES and PNET form a single group of bone and soft-tissue tumors with typical undifferentiated ES at one end of the spectrum and PNET with clear evidence of neural differentiation at the other. The most common sites of ES are chest wall, paravertebral region, retroperitoneal space, lower extremities, and gluteal region. However, few cases have been reported in the kidney, breast, gastrointestinal tract, prostate, endometrium, the adrenal glands, brain, and lung [4]. The breast is uncommonly involved [5].

Several studies of adult EWS/PNET from the Royal Marsden, the Memorial Sloan-Kettering and the Dana-Faber Cancer Centre have reported a median age of EES as 24-27 years [6]. Imaging modalities such as mammogram, ultrasound, MRI and PET-CT help in diagnosis, however, the imaging findings are nonspecific. Maxwell described sonographic findings of primary EWS/PNET of the breast as a superficial, circumscribed, hypo echoic mass with posterior acoustic enhancement and an apparent hypo echoic tract extending to the skin [7]. The diagnosis is usually confirmed by histopathological and IHC examination. Immuno-phenotyping is necessary to confirm the diagnosis of EWS/PNET, showing positivity of for vimentin, Fli-1 and CD99 [8]. This group of tumors is characterized by the presence of the typical translocation $t(11; 22)(q24; q12)$, the EWS-FLI1 chimere transcript at the molecular biology and the expression of CD99 antigen (MIC-2) at immunohistochemistry.

EWS/PNET is an aggressive tumor with a high incidence of local recurrence and distant metastasis. A combination of multiple treatment modalities, including surgery, chemotherapy and radiation therapy, is indicated for these patients. Systemic chemotherapy improves the 5-year survival rate in localized forms of PNET from 10% up to 65% which is primarily due to the elimination of micrometastases [9]. Although the

optimum combination chemotherapy has not yet been established, a regimen containing vincristine, adriamycin, cyclophosphamide and actinomycin D, was the standard first-line treatment for patients with localized disease. In patients with unresectable or metastatic disease, palliative chemotherapy is considered. Radiation therapy can be combined with surgery, in order to control local disease [10].

CONCLUSION

ES/PNET can occasionally involve the breast as an isolated mass. Careful histological examination and appropriate immunohistochemical studies are often necessary to confirm the diagnosis. ES/PNET should be kept in the differentials for a recurrent, rapidly growing breast or chest wall soft tissue lesion.

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