

PERIPHERAL SOFT TISSUE EWING'S SARCOMA : A RARE CASE REPORT

Farzana Shegufta, Mahfuz Ara Ferdousi, Md Abu Taher

Department of Radiology and Imaging, Bangladesh Institute of Research and Rehabilitation for Diabetes, Endocrine and Metabolic Disorders (BIRDEM), 122 Kazi Nazrul Islam Avenue, Shahbag, Dhaka-1000

Abstract

A 22 years male patient presented with gradual left forearm swelling for 6 months. X ray forearm revealed large soft tissue swelling with tiny calcification and mild scalloping at inner aspect of ulna and ultrasonogram (USG) revealed soft tissue mass having calcification and necrotic areas within and spectral Doppler showed arterial type of blood flow with no augmentation. Later computerized tomography (CT) scan showed soft tissue mass with necrotic area and calcification with no bony involvement. Magnetic resonance imaging (MRI) with contrast revealed a large heterogeneously enhancing lobulated mixed intensity lesion in antero-medial compartment of the left forearm involving flexor group of muscles causing displacement of fat plane. MRI and subsequent histopathology of the lesion revealed it as a rare soft tissue Ewing's sarcoma / primitive neuroectodermal tumor (PNET) in extremity.

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Introduction

Ewing's sarcoma or primitive neuroectodermal tumor (PNET) is a neural crest tumor. It is a rare tumor, usually occurring in children and young adults under 25 years of age. Five years survival rate is about 53%.¹ PNET belongs to the Ewing family of tumors. Among these, peripheral PNET (pPNET) usually present in second decade of life with slight male preponderance and accounts 4-17% of all pediatric soft tissue tumor. Most pPNET manifest in thoracopulmonary region (Askin tumor), rare in abdomen, pelvis, head, neck and extrimities.² We present here a rare case of pPNET in the extremities.

Case report

A young male of 22 years reported to radiology and imaging department of BIRDEM from Keranigonj for MRI examination of left forearm as per advice of a neurosurgeon. Chief complaints of the patient was gradual swelling over the left forearm for 6 months. General examination of the case revealed no

significant abnormality. Local examination of left forearm revealed a swelling of about 10cmx6cm mainly in the antero-medial aspect of left forearm which was firm in consistency, non tender and free from overlying skin. Biochemical parameters were normal. X ray forearm revealed large soft tissue swelling with tiny calcifications. Mild scalloping was seen at inner aspect of ulna (Fig-1). Ultrasonogram of swelling revealed a soft tissue mass measuring about 9cmx6cm with calcification and necrotic areas within at the medial aspect of left the forearm (Fig-2). Spectral Doppler revealed arterial type of blood flow within the lesion that represented no venous malformation. There was no augmentation of flow. Computerized tomography (CT) of swelling showed a soft tissue mass with necrotic areas and calcification in antero-medial aspect of the left forearm (Fig-3). Bone window showed no bony involvement. Magnetic resonance imaging (MRI) with contrast revealed a large (50mmx38mmx92mm) heterogeneously enhancing lobulated mixed intensity lesion in antero-

Address for Correspondence:

Dr. Farzana Shegufta, Department of Radiology and Imaging, Bangladesh Institute of Research and Rehabilitation for Diabetes, Endocrine and Metabolic Disorders (BIRDEM), 122 Kazi Nazrul Islam Avenue, Shahbag, Dhaka-1000



Fig-1. X-ray of left forearm antero-posterior and lateral view showing large soft tissue swelling (arrow) with tiny calcification and scalloping at inner aspect of ulna (arrow head).

medial compartment of mid part of the left forearm involving flexor group of muscles predominantly flexor digitorum profundus and pronator teres causing displacement of fat plane. Few signal void areas were seen within the lesion suggesting calcification. T1 hypo-intense and T2 hyper-intense areas were also evident within the lesion suggesting necrosis Fig-4 and 5). No obvious intralesional hemorrhage was seen.

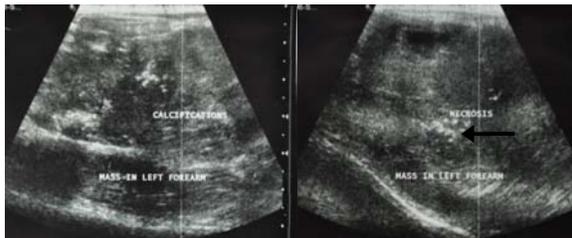


Fig-2. Ultrasonogram of left forearm swelling revealed a soft tissue mass with calcification (long arrow) and necrotic areas (small arrow).

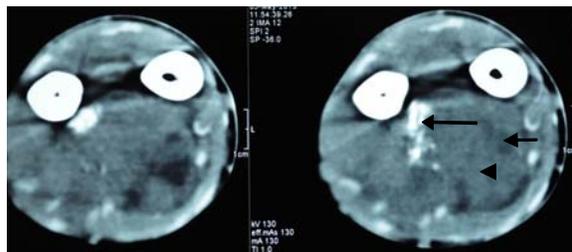
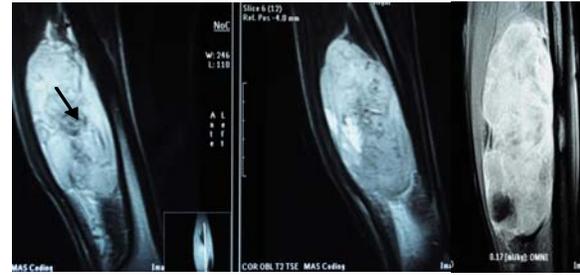


Fig-3. CT scan of swelling shows a soft tissue mass (arrow head) with necrotic areas (small arrow) and calcification (long arrow) in antero-medial aspect of left forearm.



T1W T2W Post contrast
Fig-4. Coronal T1, T2 and post contrast MR images of left forearm showing large heterogeneously enhancing lobulated mixed intensity lesion with calcification (arrow) and necrosis (small arrow) within.

All the features suggested a soft tissue sarcoma. As sarcomatous lesion metastasize early to lung, so X ray chest was performed and it revealed a pleural based lesion along right lateral chest wall without any rib destruction (Fig-6). CT scan of chest showed soft tissue density (50 HU), which was better appreciated in mediastinal window (Fig-7). No bony

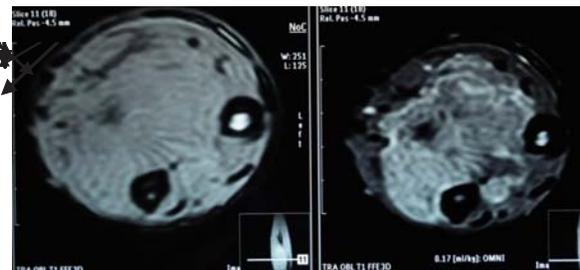


Fig-5. Axial T1W(6A) and post contrast MR (6B) images shows heterogeneously enhancing soft tissue mass (arrow).



Fig-6. Chest x-ray P/A view revealed a pleural based lesion (arrow) along right lateral chest wall without any rib destruction.

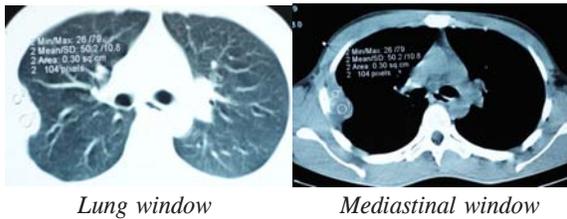


Fig-7. CT scan of chest shows soft tissue density (50 HU) mass (arrow) without bony destruction.

destruction was seen. Fine needle aspiration cytology (FNAC) from left forearm swelling reported the lesion as PNET. Subsequently, histopathology of the tissue reported a small round cell sarcoma (Fig-8). The tumor cells were present in groups, nests and pseudorosettes. Foci of tumor necrosis and fair number of mitoses were present. The tumour cells showed positive reaction with PAS, which was confirmatory for PNET as the cells of Ewing's sarcoma/PNET usually contain large amount of cytoplasmic glycogen as demonstrated by PAS stain or by electron microscopy. This represented an important feature for the differential diagnosis with other small round cell tumours.³ Based on the above, the lesion was diagnosed as a case of pPNET.

Discussion

Ewing's sarcoma are round cell sarcoma showing varying degrees of neuroectodermal differentiation.⁴ The soft tissue Ewing's sarcoma are subdivided into three morphological types with very similar age incidence, of which 34% is Ewing's sarcoma, 29% peripheral neuroectodermal tumour and 37% primitive neuroectodermal tumour.⁵

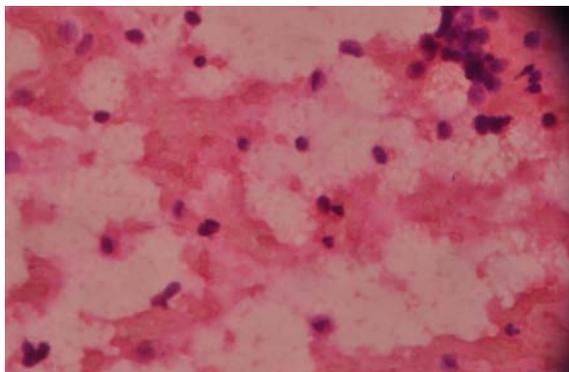


Fig-8. Cytopathology of tissue from the left forearm swelling showing small round cells (arrow).

Primitive neuroectodermal tumors (PNETs) are highly malignant tumors consisting of small round cells of neuroectodermal origin, mainly involving soft tissue and bone. Its nomenclature is based on the fact that most of the cells in the tumor are derived from neuroectoderm but have not developed and differentiated in normal way, so the cells appear primitive. Batsakis *et al*⁶ divided the PNET family of tumors into the following 3 groups based on the tissue of origin.

- CNS primitive neuroectodermal tumours (PNETs) - tumors derived from the central nervous system.
- Neuroblastoma - tumor derived from the autonomic nervous system.
- Peripheral primitive neuroectodermal tumour (pPNET) - tumours derived from tissues outside central and autonomic nervous system.

pPNETs are classified as part of the Ewing family of tumors. Both Ewing family of tumors (EFT) and pPNETs represent different presentation of the same tumor, having similar genetic alterations. Ewing sarcoma is more common in bone and pPNETs in soft tissue. Stout first described PNETs in 1918 and these tumors were thought to arise directly from nerves.⁷ EFTs and pPNET share the same reciprocal translocations in chromosome 11 and 22, based on molecular cytogenetic analysis. Tumors that demonstrate neural differentiation by light microscopy, immunohistochemistry or electron microscopy are called PNETs and those that are undifferentiated are known as ES.⁸ pPNETs are very rare, the annual incidence of tumor is 2.9 per million population from birth to 20 years of age.² These tumors are rare in African American and Asian children with most cases occurring in white and Spanish adolescents children. Sex does not play significant role in the incidence of Ewing's soft tissue sarcoma. In 25 years, the age incidence rates in males and females increased from 0.2 per million to 1.5 per million and from 0.2 per million to 1.3 per million respectively. These rates are similar to the rate of 1.0 per million reported by Toro *et al*.⁹ Ewing's sarcoma is most commonly increased in patients under the age 25 years.¹⁰ 45% of soft tissue sarcomas arise in the brain. Soft tissue sarcomas also arise in other sites including the connective tissue of the trunk (15%) and the limbs (12%). Clinical symptoms depend on the site of presentation but invariably include pain and swelling of the surrounding structures due to mass effect.

pPNETs are highly aggressive and usually present with metastases. The most common sites of metastases are lung, bone and bone marrow. Five year survival rate was found to be 56-68% in a study carried out in 25 years time period and was slightly higher for PNET in the limbs and lowest for those diagnosed in the brain.¹¹ Prognosis is worse if tumour is large with evidence of necrosis and if metastasis is present to lung and bone.¹² Cytogenetic and immunohistochemical studies for neural markers are essential in accurate diagnosis of pPNETs. Therefore, radiological studies together with histopathology may help in diagnosing such rare cases of pPNET.

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