Case Report

Ewing’s Sarcoma /Primitive Neuroectodermal tumor of plantar aspect of foot (soft tissue) in adult female: A case report

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Abstract
Ewing’s sarcoma/primitive neuroectodermal (PNET) is uncommon round cell tumor that occurs in bone and soft tissue. Histopathology, Immunohistochemistry (IHC) and cytogentic needs for the correct diagnosis. We reported a rare case of extraskeletal Ewing’s sarcoma (EES)/primitive neuroectodermal tumor (PNET) of plantar aspect of right foot in adult female. Diagnosis is confirmed by IHC with positive markers CD 99, Vimentin, S 100 protein.

Keywords: Ewing’s sarcoma, Primitive neuroectodermal tumor, Extrasosseous Ewing’s sarcoma

1. Introduction
Ewing’s sarcoma/PNET is defined as round cell sarcoma that shows varying degree of neuroectodermal differentiation. Nearly 80% patient are younger and ages of 10 to 30 years, extremely uncommon older than 30 years with predilection for males with ratio of 1.4 to 1.1. Most cases arise in deep soft tissue particularly on the trunk, the lower limb and Other common sites are the head, neck region, the paravertebral region and the pelvis is reported2,3. Extraskeletal Ewing’s sarcoma (EES)/primitive neuroectodermal tumor (PNET) is a rare, malignant small round cell neoplasm of undifferentiated mesenchyme origin / soft tissue sarcoma resembling Ewing’s sarcoma histologically yet not appearing to arise from bone were reported by Teff et al in 1969.3,4

2. Case Details
A 22 years adult female came to hospital with history of swelling over the plantar aspect right foot, round to oval in shape and of size approximately 8x8 cm. Firm to hard in consistency, fix to skin and underlying structure with moderate tenderness. Past history of excision of swelling done at similar site one and half year back, but again swelling appear at same site of present size. Records of excision are not available.

MRI examination shows approximately 7.8X 6.4 X 6.3 cm sized well defined, heterogeneously enhancing exophytic mass lesion over plantar aspect of right foot involving plantar fascia and adductor muscle, intraosseous muscle. No bony involvement. Diagnosis: Soft tissue neoplastic lesion.

Fine Needle Aspiration Cytology (FNAC) was advised. It showed dissociated and clusters of large, round and monomorphic cell having a large nucleus with clumped chromatin and 1-2 prominent nuclei. The cytoplasm is scanty and basophilic. Rosette like structure is seen. Cytological diagnosis of round cell tumor was made and advised biopsy for confirmation. We received amputed leg below knee joint. On plantar aspect of right foot tumour was measuring 10x7x3.5 cm. External surface was irregular with areas of ulceration. Cut surface was solid, firm in consistency with areas of necrosis and base was formed by muscle. Histopathology of tumor showed solidly packed lobular round cell with round to ovoid nucleus with distinct nuclear membrane and scanty blue cytoplasm (fig 4). Immunohistochemical staining was performed (by polyme detection kit). Tumour cells were positive for CD 99(fig 1), vimentin (fig 2), S100 protein (fig 3) and negative for cytokeratin, CD45, myoglobin and desmin. A final diagnosis of Extraskeletal Ewing’s sarcoma /Primitive ectodermal tumour was reported.

Figure 1: Strong membranous positivity for CD99 x100
3. Discussion

The Peripheral primitive neuroectodermal tumour, firstly recognized by Arthur Purdy Stout in 1918, reported a case with ulnar nerve tumor composed of undifferentiated round cell that formed rosettes, is a member of family “small round cell tumors.”

In 1975 Angervall and Enzinger described the first Ewing’s sarcoma arising in soft tissue extraskeletal (ES) and subsequent report confirmed by clinical and pathological feature of extraskeletal ES.

Primitive neuroectodermal tumor is malignant neoplasm that originates from cells of primitive neural crest, the PNET concept has been expanded to include histologically similar, peripherally located tumors referred to as PNET. The tumors usually involve bone and soft tissue within limbs.

The principle sites of extra skeletal ES are paravertebral region and chest wall, generally in close association with vertebra and ribs. These tumors may arise in soft tissue of lower extremities rarely in pelvic and hip region, the retroperitonium and upper extremities. Unusually described in lungs, uterus, ovary, urinary bladder, myocardium, parotid gland and kidney. There have been a number of case report for peripheral PNET including in eye, maxillofacial, peripheral limbs, gynaecological organ and intraabdominal. The tumours of ES/PNET family reported in unusual sites including kidney, urinary bladder, testes, parotid gland, oesophagus, pancreas, small intestine, hepatic duct and gall bladder.

In general, the tumor present as a rapidly growing often painless mass axial or deeply located measuring 5-10 cm in greatest diameter. Superficially located do occur but are rare. Tumor in bone and soft tissue is tan grey and often necrotic and haemorrhagic.
Diagnosis requires histopathological examination, immunohistochemistry and cytogentic. Microscopically EWS/PNET is composed of uniform round cell with round nuclei containing fine chromatin, scanty clear or eosinophilic cytoplasm and indistinct cytoplasmic membrane. Mitotic figure are common and Homer Wrights rosettes are seen. Immunostain CD99 is highly sensitive for Ewing`s sarcoma/Primitive neuroectodermal tumor, this marker should be used as part of panel in immunostain. CD99 activity is diffuse and membranous. As many as 97% of EWS/PNET have been found to express the product of MIC2 gene. Vimentin stains most neural cells and neural marker such as neuron specific enolase frequently expressed. In present case IHC supported the diagnosis of PNET, our patient showed positivity for CD99, vimentin and S100 protein and negative for desmin, CD45 and cytokeratin. These tumour are rare, aggressive and often recur and metastasize especially to lungs, liver, bone, bone marrow and lymph nodes. The diagnosis in Ewing`s sarcoma/primitive neuroectodermal tumor has improved in modern era of treatment, important prognostic factors include stage anatomic location and size of the tumor. Most patient with localised tumor are now cured by surgery, radiation therapy/ multiagent chemotherapy or various combination. In one study of extra osseous ES, 64% disease free survival in patient receiving radiation therapy and chemotherapy. Current survival rate is estimated to be 41%. In EES, five year survival rate of 38% to 67% have been reported. The PNET have a very high rate of local recurrence and propensity to metastasize. The most common sites of dissemination have been reported in bone (35-50%), bone marrow (29-33%), lung (18-50%), liver (20%), lymph node (10-20%) and spleen (10%) and metastasis in multiple sites (35%) have been noted. Multimodality treatment including radical surgical resection, non adjuvant and adjuvant chemotherapy and irradiation, is the current treatment of choice. Overall survival rate is 19% in patient who presented with metastatic disease. PNET is a highly malignant tumor with very poor prognosis. After curative resection, post-operative chemotherapy regimens are mandatory because of the particularly high risk of both local recurrences and distant metastasis.

We reported a rare case of EES/PNET of planar aspect of right foot in adult female. Diagnosis is confirmed by IHC with positive markers CD 99, Vimentin, S 100 protein. From various literatures we concluded that EES/PNET is highly malignant tumour, so early confirmed diagnosis and treatment is required for this tumour.

References