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CASE REPORT

PRIMITIVE NEUROECTODERMAL TUMOUR IN INGUINAL LYMPHNODE WITH UNKNOWN PRIMARY, A CLINICAL SURPRISE

Dr. Sathish Christopher and *Dr. Vijay Ganesh, M.

Assistant Professor, Department of General Surgery, Velammal Medical College and Research Institute

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ABSTRACT

Primitive neuroectodermal tumour (PNET) is a tumour belonging to a group which presents classically with small blue round cells.42 year old female patient presented to us with complaints of painless swelling in the left groin for the past 9 months .CT abdomen and pelvis revealed left inguinal lymphadenopathy without identifying any primary. Patient underwent excision biopsy and further evaluation with translocation studies revealed it to be PNET, primary could not be identified. It is very rare to see such a case with PNET in lymphnodes with primary being unknown.

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INTRODUCTION

A 42 year old female patient presented to us with complaints of painless swelling in the left groin for the past 9 months with sudden increase in size for the past 3 months associated with swelling of the left leg. No other remarkable past history. On examination there was a swelling of size 10x8 cm over the left inguinal region, which was hard, non tender and mobile, with lobulated surface. Routine biochemical and hematological investigations were with in normal limits. Upper GI and Lower GI endoscopy were normal. CT abdomen and pelvis revealed left inguinal lymphadenopathy without identifying any primary. Core needle biopsy was inconclusive and hence proceeded with excision biopsy which on histopathological examination revealed destroyed lymphnode architecture with tumour composed of nests of small round cells with increased nulear-cytoplasmic ratio with scanty cytoplasm, with this finding PNET was a strong suspicion and patient was investigated for EWS-FLI-1 Translocation study by RT-PCR, which was found to be positive. A search for primary was done with whole body CT, MRI Brain, bone marrow study and bone scan, all of them failed to find a primary.

Corresponding author: Dr. Vijay Ganesh, M.,
Assistant Professor, Department of General Surgery, Velammal Medical College and Research Institute.

With this a diagnosis of PNET of Left inguinal lymphnode with Unknown primary was made and patient underwent VAC chemotherapy 6 cycles. Three months after completion of chemotherapy, patient was evaluated with PET scan which revealed no residual tumour. Patient is on regular follow up for the past 6 months without any complaints.

DISCUSSION

Primitive neuroectodermal tumour(PNET) is a tumour belonging to a group which presents classically with small blue round cells. PNET is usually classified into CNS PNET and peripheral PNET depending upon the location. The peripheral PNET was first recognized by Arthur Purdy Stout in the year 1918. PNET belongs to Ewing family of tumours, with classical EWS-FLI-1 translocation in about 85% of the tumours (Delattre *et al*., 1992). This translocation t(11;22) was identified by Turc Carel *et al*. in the year 1983. Peripheral PNET is mostly found in the extremities and the paravertebral areas. PNETs can also occur in organs such as the kidney, parotid gland, heart, lung, liver,rectum, pancreas, gall bladder, ovary, vagina, testis, uterus, cervix, urinary bladder, breast, it is an extremely rare tumor entity (Movahedi-Lankarani *et al*., 2002; Hyun *et al*., 2002). The kidney is the most common visceral organ involved by PNET (Marley *et al*., 1997; Pomara *et al*., 2004).

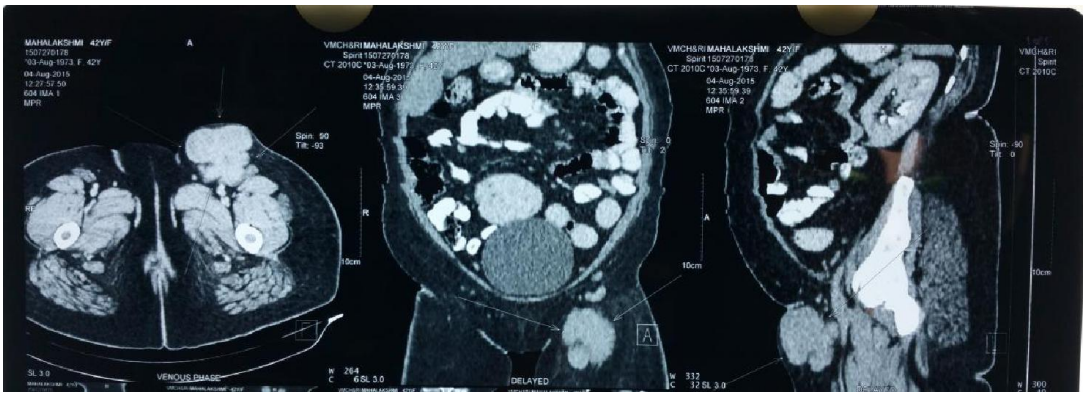

Figure 1. CT picture showing tumour

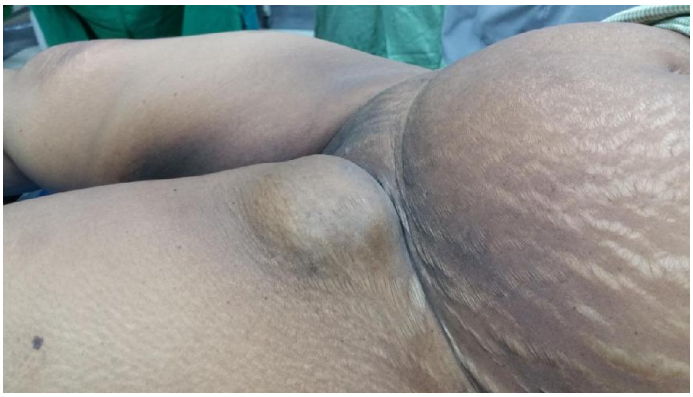

Figure 2. Pre-operative picture

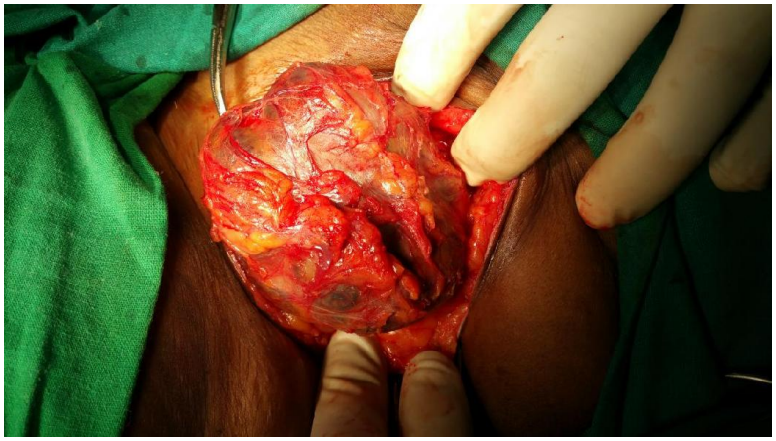

Figure 3. Intra operative picture

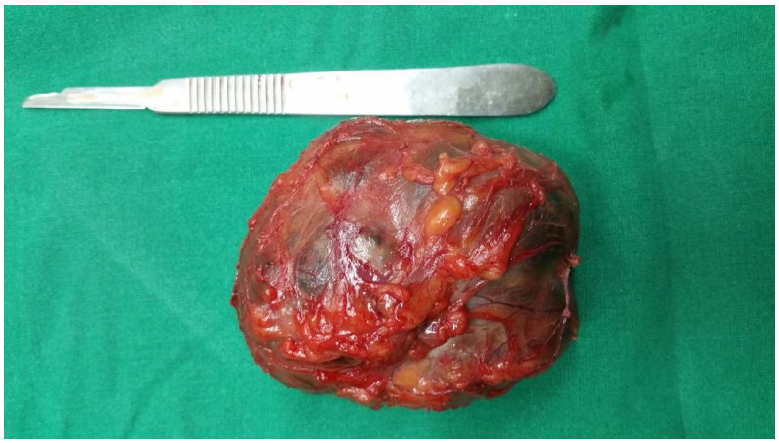

Figure 4. Tumour after excision

Investigations include histopathological examination and followed by panel Immuno Histochemical markers, which includes CD99 (MIC2) a cell surface glycoprotein responsible for cell adhesions (Tamura *et al*., 2007). But CD99 may be expressed in other tumors also, including metaplastic carcinoma of the breast, neuroendocrine carcinomas, lymphoblastic lymphoma, synovial sarcoma, and alveolar rhabdomyosarcoma (Milanezi *et al*., 2001). Cytokeratins, Chromogranin A, Synaptophysin, and LCA will be negative and will rule out other differential diagnoses like Small cell carcinoma, neuroendocrine carcinoma, and lymphoblastic lymphoma. EWS-FLI-1 Translocation study by RT-PCR or Flourescence in situ Hybridization (FISH) is a definitive diagnostic tool.

Treatment for PNET is surgical resection for local control followed by adjuvant chemotherapy and/or radiotherapy (Baldini *et al*., 1999). It has high propensity for metastatic spread. A regimen containing vincristine, adriamycin, cyclophosphamide, and actinomycin D is the standard first-line treatment for patients with localized disease (Paulussen *et al*., 2001). Radiotherapy as a primary treatment is used in about 10-15% in whom surgical resection is not possible. A 5-year survival of 58-61% with a median survival of 120 months has been noted (Smorenburg *et al*., 2007).

Conclusion

Peripheral PNET with unknown primary, this is the second such case to be reported in the literature after a case reported by Chaudhuri *et al*. (2013). The patient in this case report was treated with primary excision followed by chemotherapy. She is under regular follow up and there is no evidence of recurrence of the tumour till now.

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