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A rare case of dorsal spine primitive Neuroectodermal Tumor (P.N.E.T) presenting with features of Pott's spine

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Abstract
Primary intraspinal primitive neuroectodermal tumor (PNET) is a type of round cell malignant tumor which is reported only above 100 in literature. We report a case of Dorsal Spine Primitive Neuroectodermal Tumor (P.N.E.T) Presenting with Features of Pott's Spine, & discuss its pathological features, radiology, and treatment options.

Keywords: CD99, EWS-FLI1 translocation, spinal epidural peripheral primitive neuroectodermal tumor, thoracic compressive myelopathy

Introduction
Malignant small round cell tumors are characterized by small, round, relatively undifferentiated cells in histopathology. These are also called small round blue cell tumors as the cells are blue, in the sense that they have large hyperchromatic nuclei and a thin rim of cytoplasm. Tumors that belong to this group are: desmoplastic small-round-cell tumor, Ewing's Sarcoma (ES)/primitive neuroectodermal tumors (PNET), neuroblastoma, medulloblastoma, rhabdomyosarcoma, synovial sarcoma, carcinoid tumor, mesothelioma, hybrid oncocytoma/chromophobe renal cell carcinoma, leiomyosarcoma, small cell lung cancer, Wilms' tumor, retinoblastoma, Small-cell lymphoma, hepatoblastoma, and Merkel cell carcinoma [1].
Differential diagnosis of small-round-cell tumors is particularly difficult due to their undifferentiated or primitive character from their morphology alone. Therefore, a multimodal approach using fine-needle aspiration cytology, immunohistochemistry (IHC) and immunophenotyping by flow cytometry, reverse transcriptase polymerase chain reaction, fluorescence in situ hybridization, and electron microscopy are employed. PNET is a type of round cell tumor, which is of neuroectodermal origin but has poor differentiation. It is classified into two types as follows: peripheral and central nervous system (CNS) PNET. Peripheral PNET (p-PNET) belong to Ewing's family of tumor, since they have similar histological and immunohistochemical characteristics. p-PNET sarcoma predominates in the second decade of life, and the pelvis and femur are most commonly affected sites. The term, "PNET" includes malignant small-round-cell tumors of the thoracopulmonary region (Askin's tumor), extraskeletal ES, peripheral neuroblastoma and peripheral neuroepithelioma. Primary spinal epidural p-PNET is rarely reported in the literature.

Case Report: A 17-year-old, healthy boy presented with the complaints of gradually progressive backache along with weakness of both lower limbs of 4 weeks duration. The progression of paraparesis to paraplegia was agile, over about a week. At the time of presentation, he was paraplegic, had absent sensation below D10 level bilaterally, bilateral plantar extensor, and bladder catheterized. Patient was evaluated for Tuberculosis and Metastasis. His HRCT Thorax showed Normal study & CSF analysis was negative for malignant cells or AFB & CEA was 1.80ng/ml. His Magnetic Resonance Imaging Thoraco-lumbar spine showed lytic lesion in D11 body with altered marrow signals in it & also in its neural arch appearing hyperintense in STIR Images & hypointense in T1WI. After contrast administration inhomogenous enhancement is seen. There is evidence of epidural soft tissue lesion in the spinal canal, mainly in posterior lateral aspect on left side extending from lower

D10 to upper D12 levels which is hypointense in T1WI with dense inhomogenous enhancement in it with extension in the paraspinal region on left side. [Figure 1]. It is compressing & displacing the thecal sac with hyperintense signals in T2WI in the spinal cord at D11-12 level which is isointense in T1WI [Figure 2]. Vertebrae at other levels are normal in alignment, morphology & signal intensity. No Marrow edema is seen.

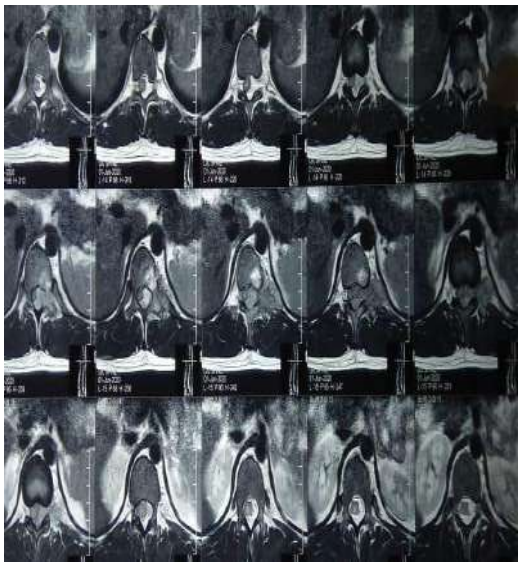

Fig 1: MRI D-L Spine (Axial) contrast

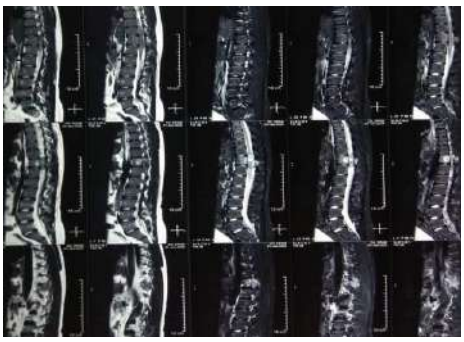

Fig 1: MRI D-L Spine (Saggital)

He underwent D10 & D11 Left-sided Hemilaminectomy, and D12 Bilateral laminectomy and Sub-total excision of the lesion with Bilateral D10-D12 Transpedicular Screw Rod fixation. Intraoperatively, a whitish, soft elastic, moderately vascular with necrotic areas in between extradural tumor compressing the spinal cord on the left side found. Tumor was found to be involving both D10, and D 11 left intervertebral foramina & paraspinal soft tissues. Tumor was dumbbell-shaped.

Tumor was removed partially in a piecemeal fashion, and there was multiple gray-brown soft tissue within on the left side of thecal sac which was extending along the T10-T12 foramen paraspinally. There was a severe mass effect on the cord. The lesion was dissected meticulously from the dura and nerve roots to achieve a Sub-total resection. Whole Body PET-CT: Metabolically mildly active residual lytic lesion in D11 with paravertebral soft tissue mass. Metabolically active circumferential wall thickening in caecum & ileocaecal junction. No metabolically active other disease in rest of body. His colonoscopy showed Pseudomembranous Colitis & Biopsy from Rectum & sigmoid colon showed features of Chronic Colitis. Histopathological examination showed a tumour composed of small round uniform cells with scanty eosinophilic cytoplasm in a light cell & dark cell appearance. Some cells have Hyperchromatic nuclei and some have stippled chromatin withoccasional mild indentation and nucleoli. Mitosis is evident. Stroma is fibrous and show areas of haemorrhage. Section from bony fragments shows bony trabeculae with increased eosinophils in the marrow spaces. IHC was negative for CK, Synaptophysin, LCA, CD1a and WT-1. The tumour cells express CD99 & NKX2.2 (Diffuse, strong). The Mib-1labelling index is approximately 30%in areas of highest proliferative activity. Findings were suggestive of PNET. He was then referred to a radiation oncologist for further management. After 1 month of the review, he had only marginal improvement in symptoms.

Discussion: PNET is a small-round-cell malignancy arising in soft tissue and bone, predominantly in older children and adolescents with a male preponderance. They can arise either from central nervous system (cns- PNET) or from periphery (p-PNET). p-PNET has the histopathological characteristic similar to ES. Osseous ES, extraskeletal ES, pPNET are nowadays generally known as ES family tumors. p-PNET are tumors of adolescents, mostly in male in the second decade. They most commonly involve chest wall (askins tumor) pleura, pericardium, and soft tissues. The incidence of these tumors in the spinal cord is rare [2]. Review of previous cases reported in the literature suggests that p- PNET may arise from all levels of the spine and can be intradural-intramedullary or extramedullary (most common site being cauda equina), or extradural. Literature also suggests intradural extramedullary and intramedullary location to be almost equal in frequency [3].

Although p-PNET/ES mostly arises from peripheral soft tissues and bone, they have also been reported from CNS. The tumor is most frequently located at lower spinal levels, in lumbar and lumbosacral regions. Their occurrence in thoracic spinal epidural space is very rarely reported in the literature. On histopathology, the cytoplasm is scanty, eosinophilic, and usually contains glycogen, which is detected by periodic acid–Schiff stain and is diastase degradable. The nuclei are round, with finely dispersed chromatin, and one or more tiny nucleoli. Depending on differentiation, tumor cells may also express neuroendocrine proteins (synaptophysin and chromogranin A), neural crest-derived protein (S 100), neuron-specific enolase. The mesenchymal markers such as intermediate filaments (cytokeratin, vimentin, neurofilament, desmin, and glial fibrillary acid protein) though nonspecific can be positive. These tumors share the chromosomal translocation, t(11:22)(q24:q12). This genetic anomaly leads to the creation of a fusion protein consisting of EWS and FLI-1 gene products. The FLI-1 protein, the gene product of FLI-1, t(11:22), is positive in 85% of all EWS/PNET cases. The microneme protein 2 (MIC2) gene is a pseudoautosomal gene, located on the short arms of the sex chromosomes. ES and pPNET cells express glycoprotein CD99 in very high amounts and a highly selective manner, which help to differentiate from other malignant round cell tumor [3]. Surgery followed by craniospinal irradiation and chemotherapy with cyclophosphamide or ifosfamide, cisplatin or carboplatin, and vincristinebleomycin have shown to benefit [4]. However, despite all treatment mean survival rate is around 3 years. A very high index of suspicion with the good immunohistochemical analysis is required for the diagnosis of primary intraspinal p-PNET. Awareness about this condition is necessary for early diagnosis and appropriate management.

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