Primary Intramedullary Primitive Neuroectodermal Tumor Of Spine– A Rare Clinical Presentation

Sweety Gupta1, Shashank Srinivasan2, Rashi Agarwal3, Sandeep Agarwal4, Sudarsan De5, Malay Nandy6

1 Radiation Oncology Dept Of Oncology Galaxy Cancer Institute, Pushpanjali Crosslay Hospital, Ghaziabad – U.P

ABSTRACT

Primitive neuroectodermal tumors (PNETs) are a group of malignant neoplasms which were initially defined by Hart and Earle in 1973. PNETs most commonly occur in the cerebellum but can arise in the pineal gland, cerebrum, spinal cord brain stem, and peripheral nerves. Intraspinal PNET is extremely rare and histologically indistinguishable from other neural axis PNETs. A a multitude of therapeutic strategies have been employed with varying success. We report a case of Primary intraspinal PNET which was rare location in the intramedullary region with review of literature.

Key words: Intramedullary, Primitive neuro ectodermal tumors, lumbosacral

Corresponding author address: Dr. Sweety Gupta, B-1 503, Olive County, sector-5, Vasundhara, Ghaziabad (U.P.) India. Pin code – 201012 E-Mail: drsg2411@yahoo.co.in
M: 9891981332

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INTRODUCTION

Primitive Neuroectodermal tumor (PNET) are a heterogenous group of neoplasms initially defined by Hart and Earle [1] in 1973 occur mostly in childhood and early adulthood. According to WHO[2], they are embryonic tumors composed of undifferentiated or poorly differentiated neuroepithelial cells that display divergent differentiation along neuronal, astrocytic, ependymal, muscular or melanotic lines. The most common PNET involving the spinal cord are drop metastases from primary intracranial tumors which disseminate via CSF. Primary Intraspinal PNETs are rare and constitute less than 1% of primary spinal tumors. We report a case of

Primary intraspinal PNET which was rare location in the intramedullary region with review of literature.

CASE REPORT

A 33 year old male presented with lower backache for three months with progressive lower limb weakness alongwith bladder and bowel involvement. On clinical examination, he was conscious, oriented. Power in bilateral lower limbs was 3/5 with sensory loss below L3.Contrast Enhanced MRI Spine reported intradural oblong mass with well defined margins occupying the spinal canal at L5 level extending upto L4 level that measured 5.5cm in long axis and 1.5x1.8 across approximately. Patient underwent L3-S1 Laminectomy and tumor excision with duraplasty. Histopathological Examination reported Small nests of tumor cells with clear cytoplasm, round to angulated nuclei with brisk mitosis suggestive of PNET. Immunohistochemistry revealed CD 99 strongly+, NSE +, Ki 67-80% GFAP, Synaptophysin and Chromogranin negative (Fig: 1).Post operative Craniospinal MRI revealed residual intramedullary soft tissue lesion in the lumbosacral region with deposit at D12 vertebrae(Fig: 2). Patient then received post operatve radiotherapy to whole brain and spine for a dose of CTV 36Gy/20fr followed by boost 9Gy/5fr to gross lesion. Whole body PET CT Scan (Fig: 3) done after radiation was suggestive of absence of FDG avid metabolically active disease. He then received three cycles of adjuvant combination chemotherapy (Vincristine,Adriamycin, Cyclophosphamide alternating with Ifosfamide and Etoposide). Patient then defaulted. He survived for nine months.

Fig:1 Small nests of tumor cells with clear cytoplasm, round to angulated nuclei with brisk mitosis (Hematoxylin and eosin,40x view))

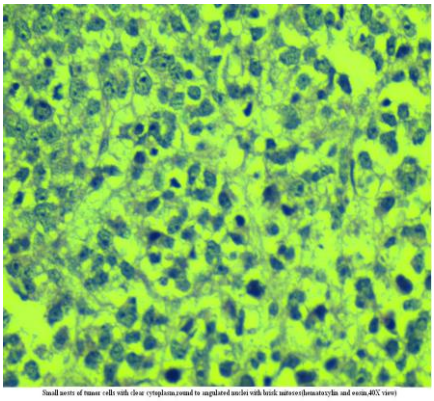


Fig 2: MRI spine showing intramedullary lesion at L4-5 level,isointense on T1W images and hypointense on T2W images)

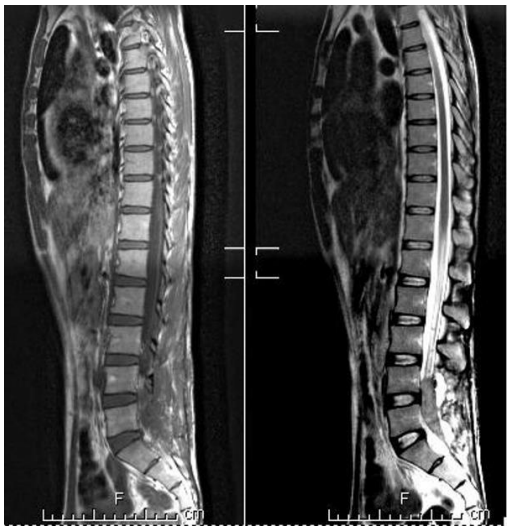
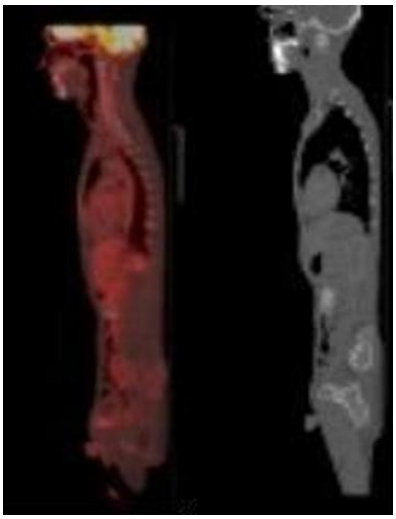


Fig: 3 Post Radiotherapy PET CT Scan showing absence of metabolically active disease)



DISCUSSION

Primitive neuroectodermal tumors (PNETs) are a group of malign neoplasms which were initially defined by Hart and Earle in 1973. PNETs most commonly occur in the cerebellum but can arise in the pineal gland, cerebrum, spinal cord brain stem, and peripheral nerves. Intraspinal PNET is extremely rare and histologically indistinguishable from other neural axis PNETs. The

earliest recognized case report of a spinal PNET is from a paper by Smith et al. published in 1969.[3] Eighty two cases have been reported since 1969. It has been postulated that PNETs arise from neoplastic transformation of primitive neuroepithelial cells in subependymal zones. PNETs may arise at all levels of the spine and can be intramedullary, intra - and extramedullary, extramedullary or extradural. 10 cases of primary intramedullary PNETs have been reported till date. Pure IPNETs are tumors presenting mainly in children and young adults(mean age 12.9 yrs) and they have a predilection for thoracic spine(77.8% of all cases). Cranial symptoms are not a feature of primary intraspinal PNET and this distinguishes primary intraspinal PNET from primary intracranial PNET with spinal metastasis. The cranial PNET frequently disseminates via the cerebrospinal fluid and rarely metastasize outside the CNS whereas IPNET may disseminate to distant sites. The characteristics histological features are poorly differentiated small, round/spindle-shaped cells, densely packed or in sheets or nests and positive neuron-specific enolase, CD99, synaptophysin, glial fibrillary acidic protein (GFAP) and S-100. As indicated by recent studies expression of MIC2 glycoprotein by immunocytochemical staining (CD99) can help to differentiate between central and peripheral PNET [6]. Surgery excision is the primary treatment. Craniospinal irradiation has been shown to be of benefit. Chemotherapy regimens include combinations of cyclophosphamide or ifosfamide, cisplatin or carboplatin, and vincristine [7]. Primary spinal PNETs are uncommon malignancies and a multitude of therapeutic strategies have been employed with varying success. Early detection, tumor identification and surgical removal and aggressive neuraxis radiation, offers hope of long term and good quality survival.

CONCLUSION

Primary spinal PNETs are uncommon malignancies and a multitude of therapeutic strategies have been employed with varying success. Early detection, tumor identification and surgical removal and aggressive neuraxis radiation, offers hope of long term and good quality survival. Cases seen in day to day practice needs to be reported to understand the management strategies which are the most beneficial for the patient.

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