



**SPINAL EXTRADURAL PRIMITIVE NEUROECTODERMAL TUMOUR IN A
CHILD A CASE REPORT**

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Abstract :

Primary spinal cord extradural primitive neuroectodermal tumors are rare . These belong to a category of aggressive childhood malignancies . They pose a significant challenge in the management . Primary spinal extradural PNET most commonly occur in children and young adults . So far around fifty cases have been reported in the literature . This case is presented for its rarity and also for its typical presentation . These tumors are highly aggressive and have a rapid growth . This 6 year female child underwent gross total excision of the extradural tumor . Review of the literature shows that the overall prognosis is very poor even with surgery , chemotherapy and radiotherapy .

Keyword :

Spinal cord tumors , primary , extradural tumor , pediatric, PNET

Introduction:

Primitive neuroectodermal tumors (PNETs) , term coined by Hart and Earle is a group of malignant neoplasms derived from the primitive neural crest, are highly malignant and mainly exist in the central nervous system (CNS), chest wall, lower extremities, trunk, kidney, and orbit , but rarely in the spine. These tumors are characterized by neuroectodermal derivation and anatomic distribution. Currently, Ewing sarcoma (ES), primitive neuroectodermal tumors (PNETs), and malignant small round-cell tumors of the thoracopulmonary region are suggested to be different manifestations of a single tumor family. However, these tumors are rare and often

occur in childhood or adolescence. Only a few series have been described in the past. We review a case with the same pathology. Though multidisciplinary treatments have been proposed the standard therapy for PNETs, is the complete excision followed by craniospinal irradiation.

CASE REPORT

A 6 years old female child apparently normal before 4 months presented with progressive weakness of both lower limbs . The child was not able to perceive the sensations below the level of umbilicus . The weakness was getting worse day by day and child had difficulty in rolling over in bed and sitting up .There was continuous dribbling of urine and the child was taken to a nearby hospital where she was catheterised. Child had an history of fall from bicycle 2 months before for which she was treated conservatively and discharged . Child had no history of seizures or vomiting . There was a no history of evening rise of temperature and loss of weight. Child was conscious and taking feeds normally. Examination of cranial nerves was normal .Bulk was normal in all limbs. On initial examination there was hypotonia of both lower limbs. On examination of power, the child was not able to move both lower limbs . The superficial reflexes in the lower abdomen and plantar was not elicitable and deep tendon reflexes were absent in the lower limbs There was no bladder sensation as evidenced by clamping the urinary catheter .The child underwent MRI of the spine which revealed a mass which was hypointense on T1W, hyperintense on T2W, and enhanced heterogeneously from C 6 – T 3 levels with cord compression.

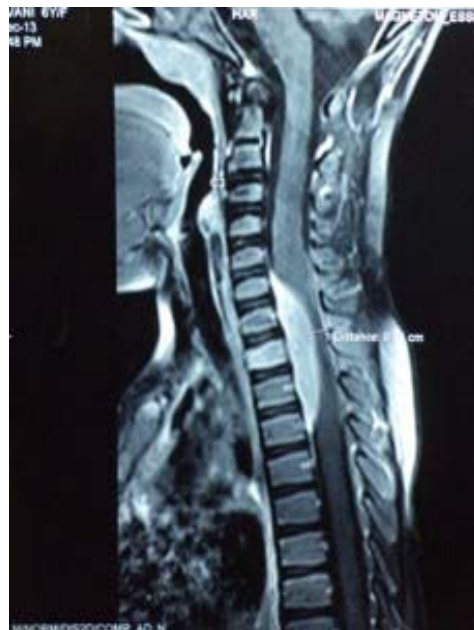
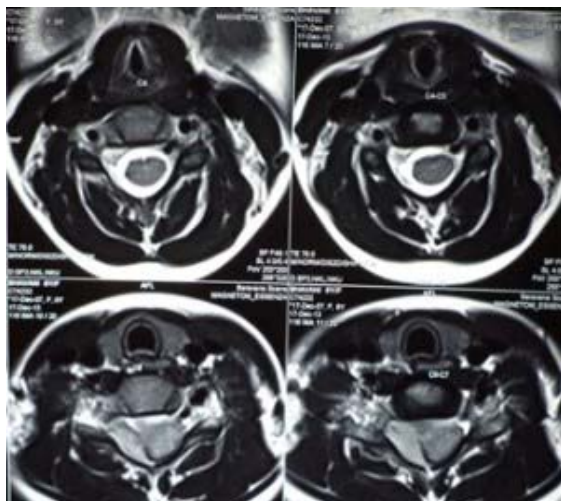


Fig 1 . T 1 contrast sagittal view showing C6 to D3 lesion Fig . 2 T1 sagittal view
Fig 3 . T2 sagittal view showing C 6 to D 3 level lesion



Child was taken up for surgery. C 5 to T 3 laminotomy was done. Pinkish soft friable granulation like tumor was found from C 6 to T 3 with cord compression. C7 root was engulfed by the tumor on right side . Gross total removal of the tumor was done. Tissue was sent for HPE. Wound was closed in layers after hemostasis. Tissue specimen represented a cellular neoplasm composed of small round cells with reticular nuclei and finely developed chromatin cells arranged in dense sheets of peritheliomatous and papillary pattern. Areas of hemorrhage and necrosis was seen .Areas of micro calcification were present. CD99 was positive , synaptophysin was negative , NSE was positive and PAS negative .Diagnosis of a small round cell tumour consistent with PRIMITIVE NEUROECTODERMAL TUMOUR was arrived.

Fig 4 . Axial cuts showing an extradural lesion

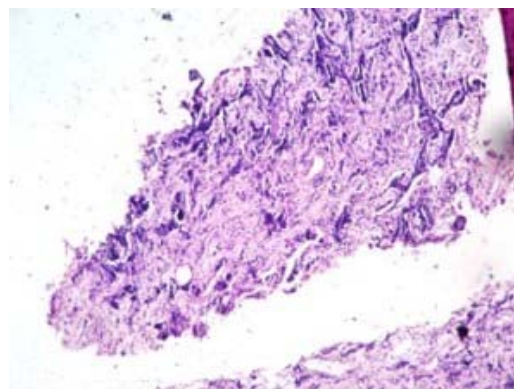


Fig 5 . HPE High power PNET
Fig.6 HPE Low power PNET

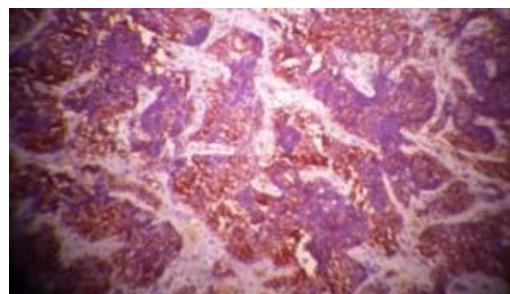
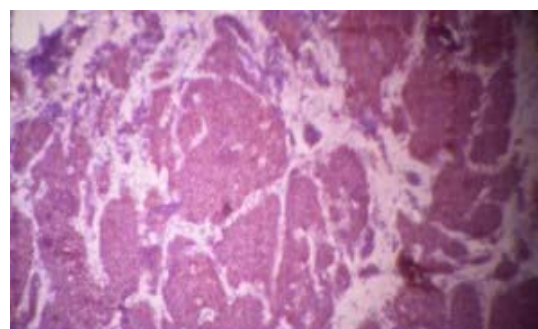


Fig 7 . CD 99 and NSE positive IHC

Fig 8 . CD 99 and NSE positive IHC

Postoperative period was uneventful. Wound healed well and sutures were removed on the tenth day. Physiotherapy was done regularly. The child showed improvement of the power and was able to stand with support .The child showed improvement of the power and was able to stand with support . Bladder sensation showed significant improvement and at the time of discharge the

child was discharged with an indwelling urinary catheter . Parents were educated regarding the bladder care and was advised for a regular follow up . There was no respiratory difficulty .The child was evaluated for any intracranial lesion .MRI of the brain showed no intracranial lesion. Screening imaging for thorax and abdomen did not show any lesion .Tc 99 MDP bone scan was negative for metastatic bone disease. After discussion with the radiation oncologist , post op radiotherapy and If osfamide and etoposide based chemotherapy was given . The child is being followed up regularly .

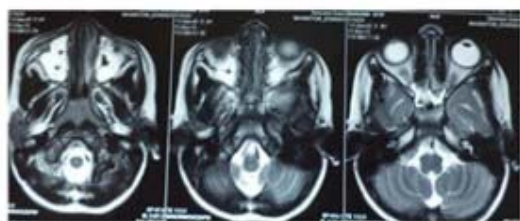


Fig 9 Brain screening showing no intrac-



Fig 10 Technitium 99 MDP bone scan – negative for metastatic bone disease

Discussion: Primitive neuroectodermal tumour (PNETs) first described by Bailey and Cushing in 1925, were also called spongioblastoma cerebelli. PNETs can occur outside the brain and throughout the body, as peripheral neuroblastomas and Ewing sarcomas. PNETs eventhough found in both children and adults, and they are more common in children. The mean age is between 5 and 77 years, and 80% of tumours occur in less than 15 years. There is male predominance; it is 1.4 to 4.8 times more common in males than in females. Usage of maternal folate, iron, and multivitamin supplementation reduces incidence of medulloblastomas. Several syndromes are associated with a familial increased incidence of medulloblastoma. They are Gorlin's syndrome ,Li-Fraumeni syndrome, and Turcot's syndrome. Intramedullary spinal cord tumors (IMSCTs) of the PNET type are usually rare.

The onset of symptoms occurs in months . Frequently identified after a trivial trauma. Diagnosis is by radiographic techniques. Majority of the tumors are benign in nature and have insidious growth pattern. The most common complaint is weakness. Other complaints are pain , gait disturbances, dysesthesia ,and spinal deformity . Other symptoms may be of sphincter disturbance and sensory deficits. The clinical picture is of myelopathy is seen in most patients. Symptoms and signs can vary according to the level of the spinal cord involved with tumor. Tumours of the cervico thoracic spine have a more insidious onset. Usually manifests with pain and

progressive scoliosis. They may also present with respiratory complaints. Examination may show paraspinal spasm and evidence of myelopathy. Sensory findings and bowel and bladder dysfunction occurs late. MRI is the investigation of choice. MRI provides excellent soft tissue imaging within the spinal column, and any intramedullary tumor, edema, and cysts can be visualized. Also the extent of the solid portion of the lesion, differentiation between tumoral cysts from nontumoral cysts can also be obtained. Computed tomography and plain radiography are reserved for the evaluation of associated spinal deformities/instabilities. T1-weighted sequence, with and without gadolinium enhancement, and a T2-weighted sequence in the axial and sagittal planes are studied. A gradient echo sequence and a fluid-attenuated inversion recovery (FLAIR) gives information about hemosiderin deposits (gradient echo) or subtle intramedullary lesions (FLAIR). In children, a mass lesion within the spinal cord is probably an astrocytoma, ganglioglioma/glioma neuronal tumor, or ependymoma. A hemangioblastoma or primitive neuroectodermal tumor is a rare possibility that can be considered when an unusual clinical presentation or imaging is obtained. Due to the aggressive behavior of the neoplasm and its great potential to metastasize, treatment should be multimodal, involving radical surgical resection, radiotherapy, and chemotherapy. Initial management of these tumors is almost always surgical since tissue biopsy can be obtained at surgery, which also serves the purpose of debulking of the tumor. 80% resection or more gives a 5-year event-free survival rate higher than 70%. Aggressive resection has been shown to

improve survival in children with primary spinal PNETs. These tumors need radiation therapy but that there is a risk of injury to the growing spine and the risk of induction of second malignancies always exist. Radiation therapy is a critical component in the management of newly diagnosed spinal PNETs. The possibility of primitive neuroectodermal tumor to disseminate along the neuraxis supports prophylactic craniospinal irradiation with an involved boost to the primary site. The utility of chemotherapy in the management of PNET remains unclear. The usual agents used included ifosfamide, vincristine, methotrexate, cisplatin, and lomustine. This combination gives 49.3% 3-year progression-free survival rate with optimal surgery followed by chemoradiation. The use of temozolomide has yet to meet significant success.

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