Abstract:
Primary spinal primitive neuroectodermal tumours (PNETs) are a rare entity. Primitive neuroectodermal tumours (PNET) are aggressive childhood malignancies and offer a significant challenge to treatment. Most of them occur in children and young adults. To date, 47 cases of primary spinal PNET have been reported in the literature. We present two cases of primary spinal extradural PNET. These tumours are highly aggressive with rapid growth as evidenced by the short history in both of our cases. Both cases underwent gross total removal. Review of the literature shows that the overall prognosis of PNETs of the spinal cord is very poor even with adequate surgery, radiotherapy and chemotherapy.

Keyword: Spinal cord tumours, Extradural SOL, Primitive neuroectodermal tumor, 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 two cases of same pathology. Though multidisciplinary treatments have been proposed the standard therapy for PNETs, is the complete excision followed by craniospinal irradiation.
Case report 1:
A 2 years old female child apparently normal before 2 months presented with progressive weakness of both lower limbs. Mother noticed the child not perceiving sensation below the level of umbilicus when she nursed the child. The weakness was getting worse day by day and child had difficulty in rolling over in bed and sitting up. Mother also noticed continuous dribbling of urine. Child had no history of seizures, vomiting or weight loss. Child was alert active cry was good and taking feeds normally. Examination of Cranial nerves was essentially normal Bulk was normal in all limbs. We noticed hypotonia of both lower limbs. On examination of power child was not moving both lower limbs spontaneously or to painful stimuli. The superficial reflexes in the lower abdomen and plantar was not elicitable and deep tendon reflexes were absent in the lower limbs. There was continuous dribbling of urine noted. The child underwent MRI of the spine which revealed a mass which was hypointense on T1W, hyperintense on T2W, and enhanced heterogeneously from D11-L2 levels with cord compression. The cord was pushed anterolaterally.
Axial section showing tumour with cord pushed to one side
Child was taken up for surgery.D9 to L3 open door laminotomy was done. Pinkish soft friable granulation like mass was found from D11 to L2 with cord compression. Gross total removal of the mass 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 extensive hemorrhage and necrosis. Areas of microcalcification were present. CD99 was positive. Diagnosis of a small round cell tumour consistent with PRIMITIVE NEUROECTODERMAL TUMOUR was arrived.

MRI Brain without any demonstrable pathology.

Case report 2:
A 23 years old male apparently normal before 3 months presented with progressive weakness of all 4 limbs. The spastic weakness becoming worse day by day and he had difficulty in rolling over in bed and sitting up. Examination of Cranial nerves was essentially normal. Bulk was normal in all limbs. On examination of the bulk of all limbs were normal, tone – increased, power ranged from 3 to 4- and deep tendon reflexes were exaggerated. MRI of the spine which revealed a mass which was hypointense on T1W, hyperintense on T2W, and enhanced heterogenously from C3-D2 levels with cord compression. The cord was pushed anterolaterally. Patient was taken up for surgery.C4 to D1 laminectomy was done. Greyish soft friable granulation like mass found from C4 to D1 with cord compression. Gross total removal of the mass was done. Tissue was sent for HPE. Wound 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 peritheliomatous and papillary pattern. Areas of extensive hemorrhage and necrosis. Areas of microcalcification present. Evidence of neuronal proliferation. CD 99 not done. Diagnosis of a small round cell tumour consistent with PRIMITIVE NEUROECTODERMAL TUMOUR was arrived. Postoperative status uneventful. Wound healed well and sutures removed on the tenth day. Physiotherapy was done regularly. Patient showed improvement of the power, evaluated for any intracranial lesion. MRI of the brain showed no intracranial lesion. Now patient is being referred for Chemotherapy and Radiotherapy.

Axial section showing extradural SOL with cord pushed to one side

MRI Brain without any demonstrable pathology.

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) are usually rare. The onset of symptoms occurs in months. Frequently identified after a trivial trauma. Diagnosed 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 in most patients. Symptoms and signs can vary according to the level of the spinal cord involved with tumor. Tumors of the thoracic spine have a more insidious onset. Usually manifests with pain and progressive scoliosis. 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 lesions, edema, and cysts can be visualized. Also the extent of the solid portion of the lesion, to differentiate tumoral cysts from nontumoral cysts. 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 subtile intramedullary lesions (FLAIR).

Children, a mass lesion within the spinal cord is probably an astrocytoma, ganglioglioma/gliial neuronal tumor, or ependymoma. A hemangioblastoma or primitive neuroectodermal tumor is a rare possibility that can be considered when an unusual image or clinical state is present. 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 obtained at surgery, the usefulness 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 SPNETs.

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 SPNETs. 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.

References:


