Case Report

SPINAL PRIMITIVE NEUROECTODERMAL TUMOR IN CHILDREN–A RARE CASE REPORT AND LITERATURE REVIEW

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Abstract:
Primitive neuroectodermal tumor is a rare tumor in childhood. It is aggressive childhood malignancies and offer a significant challenge to treatment. An eleven year old female baby presented with fever, pain and neurologic manifestation. Initially it was diagnose as TB vertebrae and treated accordingly but no improvement rather new metastatic swelling appear after few days. Diagnosis was made by MRI and local tissue biopsy and immunohistohistochemistry. As it is rare and highly invasive malignancy with poor out come the case was presented along with literature review.
Key words: Primitive neuroectodermal tumor(PNET), Children, Spine.

Background: Primary spinal primitive neuroectodermal tumors (PNETs) are rare tumors. Most of these tumors occur in children and young adults. These tumors are highly aggressive with rapid growth. Review of the literature shows that the overall prognosis of PNETs of spinal cord is very poor even with adequate surgery, radiotherapy and chemotherapy.2 PNETs even though found in both children and adults but 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 PNET.2-4 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. Tumours 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.5

Case Report:
Laboni Akhter, 11 years old immunized girl was admitted in DMCH with the problems of fever for 2 months which was initially low grade for nearly 21 days but high grade for 9 days. Moreover she also complain excruciating pain in the left side of the back along with pain in the left lower limb. In addition to fever there was a swelling over left side of lower thoracic and upper lumbar region.) On examination patient was febrile, mildly pale, comfortable in right lateral position with left knee and left hip flexed. There was a tender swelling (4x2.5 cm) in the left lower thoracic and upper lumbar region. In addition to that she had features of upper motor neuron lesion as increased tone and deep reflexes exaggerated in

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lower limbs with bilateral planter extensor. Joint tenderness was grade ¾ in left knee and hip joints, range of movement was restricted in extension of left hip and knee joints, muscle power in left lower limb was 3/5. On investigation CBC was normal except moderate anemia an ESR was 34 mm at 1st hour. Blood CS, Montoux test, USG of abdomen and paravertebral region and CXR were normal. MRI and Bone scan could not identified any malignancy. Fine needle Aspiration cytology (FNAC) from 8th vertebrae shows feature suggestive on Chronic granuloma. With this evidence anti TB drug was given. But after fifteen days she again developed severe pain and a swelling on left lumber region. Second MRI done where report was - altered marrow signal intensity and heterogeneous contrast enhancement suggestive of marrow infiltrative disease. Biopsy was taken from the swelling and revealed that primitive neuroectodermal tumor (PNET). She was given standard protocol.

Fig1,2,3: CXR, Dorsolumber vertebrae and MRI on 1st MRI of vertebrae

Fig-4,5,6: Bone scan, Swelling left lateral abdomen and 2nd MRI
Discussion:
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 is required. 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. 

Sundar Venkatarama and colleagues report a 2 years old girl with same type of presentation and they treated with surgery and was event free after 1 year. Our case also was a female but 11 yrs old and she was diagnosed after metastasis so surgical treatment was not swaetable for her. She initialy diagnosed as a vertebral TB which was not occur in other case repoorts. Patnaik etal 1 reported two cases of childhood PNET which was presented like neurofibroma and both was treated by Surgery, Radiotherapy and chemotherapy but one died within 4 month of therapy. Our patient presented like Pott’s disease not neurofibroma. In contrast to their case our patient did not treated with surgery or radiotherapy, they treated with only Chemotherapy. Yavuz AA et al reported a case of 18 year old female with lession in the Lumber 4-5 region with compression of sacral nerve root, in our case it was at thoracic level T8. Perry R and colleagues reported two cases of spinal PNET with 16 and 27 years old persons who presented tumor at L4 and L5 level, among them one survived for 72 months with remission and relapse.

Conclusion:
Early diagnosis and treatment are the key point of management of childhood malignancy. A meticulous history, proper physical examination and judicious investigation is essential for fighting against pediatric cancer. Highly aggressive tumor like PNET needs special attention and rapid intervention investigation are essential for proper diagnosis and treatment. Chemotherapeutic agents are becoming more popular than radiotherapy. Comined effort of Pediatric surgeon, Oncologist and Radiotherapist should work together for conquering PNET.

References:


