Journal of Paediatric Surgeons of Bangladesh (2015) Vol. 6 (2): 64-67
Official organ of the Association of Paediatric Surgeons of Bangladesh

Journal of Paediatric Surgeons of Bangladesh

Case Report

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

AKMA MORSHED[1], S ISLAM[2] S ANWAR[3]

Abstract:

Primitive neuroectodectodermal 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. Initialy it was diagnose as TB vertebrae and treated accordingly but no improvment 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 invasssive 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 [1]. 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]

1. AKM Amirul Morshed, Associate Professor and Head, dept of Pediatric Hematology and oncology, Dhaka medical College, Dhaka
2. Shahnoor Islam, Associate Professor, dept of Pediatric Surgery, Dhaka medical College , Dhaka
3. Sayeeda Anwar, Professor and Head,Dept of Pediatric Nephrology, Dhaka medical College , Dhaka

Correspondence to : Dr. AKM Amirul Morshed, Associate Professor and Head, dept of Pediatric Hematology and oncology, Dhaka medical College , Dhaka, E-mail: amirul_morshed63@yahoo.com

Case Report:

Laboni Akhter, 11years 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 lumber 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 lumber region.Inaddition to that she had features of upper motor neuron lesion as increased tone and deep reflexes exaggerated in

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 niddle 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 sevre pain and a swelling on left lumber region. Secopnd MRI done where report was - altered marrow signal intensity and heterogeneous contrast enhancement suggestive of marrow infiltrative disease. Biopsy was taken fron the swelling and revealed that primitive neuroectodermal tumor (PNET). She was given standard protocol .

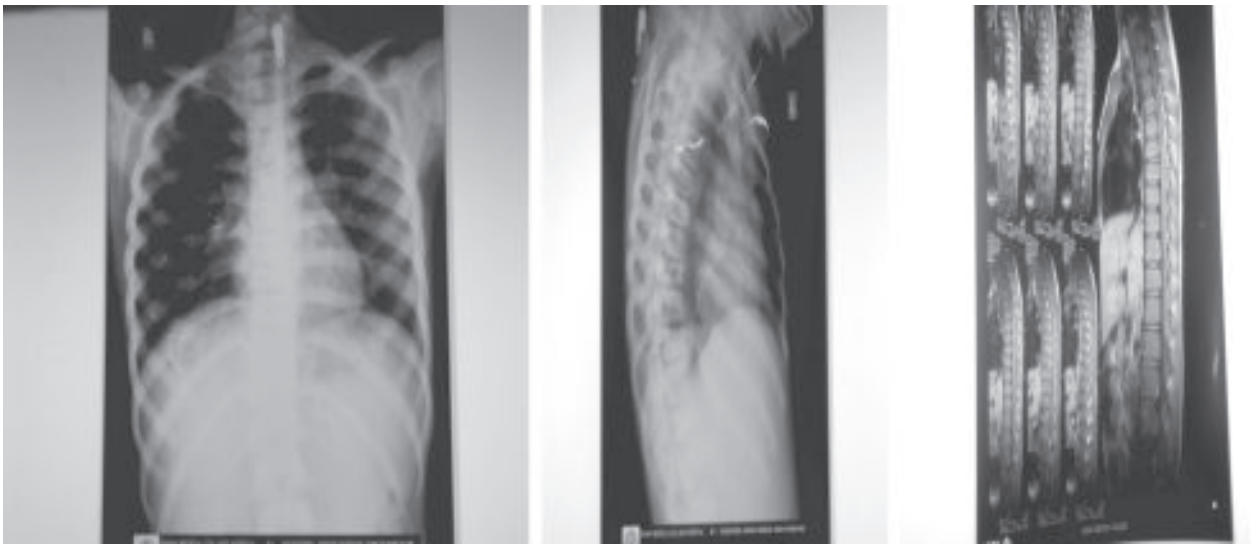

Fig1,2,3: *CXR , Dorsolumber vertebrae and MRI on 1st MRI of vertebrae*

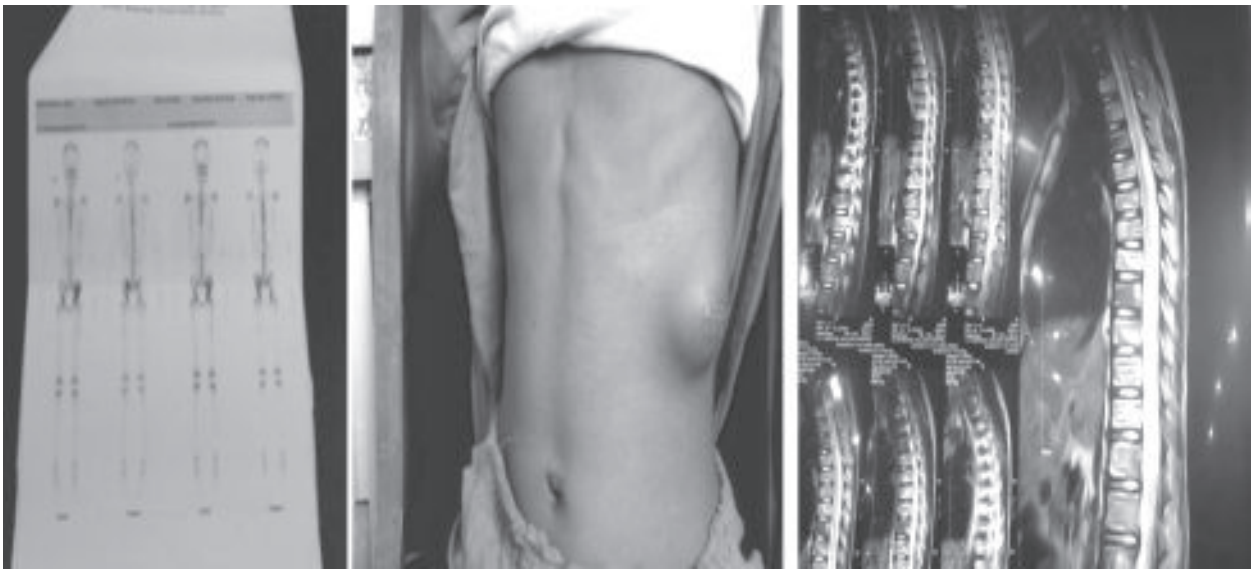

Fig-4,5,6: Bone scan, Swelling left lateral abdomen and 2nd MRI

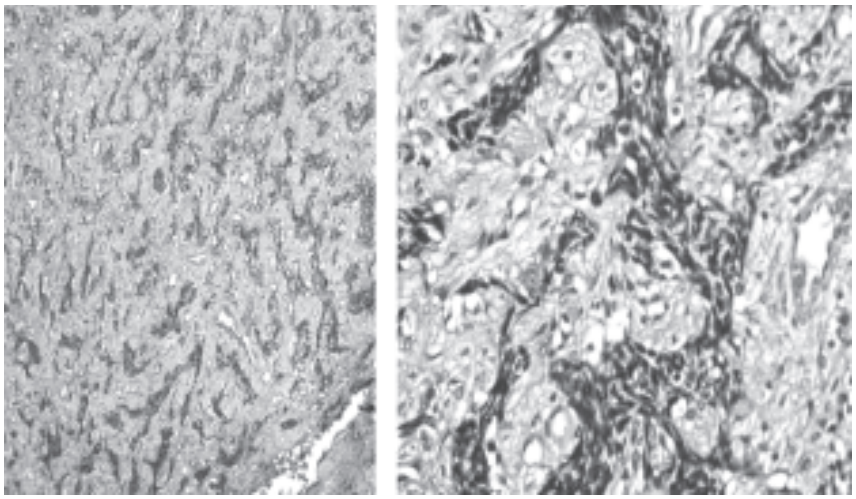

Fig 7,8: *histopathology and Immunostaining*

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 tumours 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 tumours 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 neuro ectodermal tumour to disseminate along the neuraxis supports prophylactic craniospinal irradiation with an involved boost to the primary site.[6]

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 .[6] Our case also was a female but 11 yrs old and she was diagnosed after metastasis so surgical treatment was not sweatable for her. She initialy diagnosed as a vertebral TB which was not occur in other case repoprts. 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[7] 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 colleages reported two cases of spinal PNET with 16 and 27 years old persons who presnted tumor at L4 and L5 level , among them one survived for 72 months with remission and relapse.[8]

Conclusion:

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

References:

1. Ashis Patnaik, Sudhansu S. Mishra, Sanjib Mishra & Rama C. Deo. Review of spinal neuroectodermal tumor.B J Neurosurgery. 2013; 27: 2-6
2. Akgun B, Ates D, Kaplan M. Ganglioneuro-blastoma of the thoracic spinal cord: a very rare case report. ActaMedica (Hradec Kralove) 2012;55(1):50–52.

3. Coumans JV, Walcott BP, Nahed BV. et al. Multimodal therapy of an intramedullary cervical primitiveneuroectodermal tumor in an adult. J ClinOncol. 2012;30(2)

4. Parikh D, Short M, Eshmawy M. et al. Surgical outcome analysis of paediatric thoracic and cervical neuroblastoma., Brown R. Eur J Cardiothorac Surg. 2012;41(3):630–634.

5. Ellis JA, Rothrock RJ, Moise G. et al. Primitive neuroectodermal tumors of the spine: a comprehensive review with illustrative clinical cases. Neurosurg Focus. 2011; 30(1)

6. Sundar Venkataraman, Chelladurai Pandian, Suresh A. Kumar. Primary spinal primitive neuroectodermal tumour – a case report. Ann Neurosci. 2013 Apr; 20(2): 80–82.

7. Yavuz AA[1], Yaris N, Yavuz MN, Sari A, Reis AK, Aydin F. Primary intraspinal primitive neuroectodermal tumor: case report of a tumor arising from the sacral spinal nerve root and review of the literature.Am J Clin Oncol. 2002;25(2):135-9.

8. Perry R, Gonzales I, Finlay J, Zacharoulis S .Primary peripheral primitive neuroectodermal tumors of the spinal cord: report of two cases and review of the literature.J Neurooncol. 2007 F;81(3):259-64.