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PRIMITIVE NEURECTODERMAL TUMOUR IN YOUNG ADULTS – A REPORT OF 2 RARE CASES AND REVIEW OF LITERATURE

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ABSTRACT

Primitive Neurectodermal tumours are a group of malignant neoplasms of presumed neural crest origin previously termed as "Neuroepithelioma".They are small round blue tumours which are sub classified based on differentiation and occurs mostly in brain and very rarely in the spinal cord. We report two cases of primitive neurectodermal tumour in young adult female patients. One is in the spinal cord of a young female and another is a primitive neurectodermal tumour with neuroglial differentiation. The diagnosis was made based on the morphological and immunohistochemical examination. These tumours have a highly aggressive clinical course with a poor outcome in spite of multimodality treatment options. The other case was a primitive neurectodermal tumour with neuroglial differentiation which imparts a poor prognosis.

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INTRODUCTION

A group of malignant neoplasms of presumed neural crest origin previously termed as "Neuroepithelioma" (Tommasi, 2006), were termed as primitive Neurectodermal tumour (PNET) by Hart and Earl in 1973. They are small round blue tumours which are sub classified based on differentiation and occurs mostly in brain (Ba, 2014), and very rarely in the spinal cord. These tumours are more common in children than in adults. Here we report two cases of primitive Neurectodermal tumours in young adult females.

Case 1

A 28 year old female patient presented to the hospital with complaints of neck pain radiating to left upper limb associated with left upper limb weakness for duration of 4 months. She had left sided flaccid weakness and left sided sensory impairment at C5-C7 level.

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MRI cervical spine revealed an anteriorly placed small extra dural lesion at C6-C7 level with involvement of vertebral body on the left side with extra spinal extension and minimal contrast enhancement. Anterior cervical discectomy and biopsy of extradural granulation tissue was done. Histopathological examination was inconclusive of any definite pathology. Four months later she presented with torticollis to the right side with pain increasing in severity. On examination she had lower motor neuron weakness at C5-C7 in left upper limb and upper motor neuron weakness right upper limb and both lower limbs. Her erythrocyte sedimentation rate (ESR) was elevated. The ancillary tests done for Myeloma and tuberculosis were negative. Repeat MRI showed a minimally contrast enhancing left sided antero lateral extra dural lesion extending from C2-D1 level with maximum involvement around C4-C6 level with displacement and compression of cord involving the bone. Left sided C4-C6 hemi laminectomy was done and it revealed a yellowish, fleshy, moderately vascular tumour plastered to the dura, extending to the neural foramina and involving the bone. Microsurgical excision of the tumour was done.

Histopathological examination of the mass revealed an infiltrating and necrotic tumour destroying the bone, cartilage and muscle fibers. The tumour was composed of closely packed small round cells with scant cytoplasm, round nuclei with stippled chromatin, mitosis and pseudo rosettes. Periodic acid Schiff (PAS) positive globules were seen in the cytoplasm of few of these cells. The cells showed negative Immunohistochemical staining for Leukocyte common Antigen (LCA) and exhibited strong positivity for CD 99.

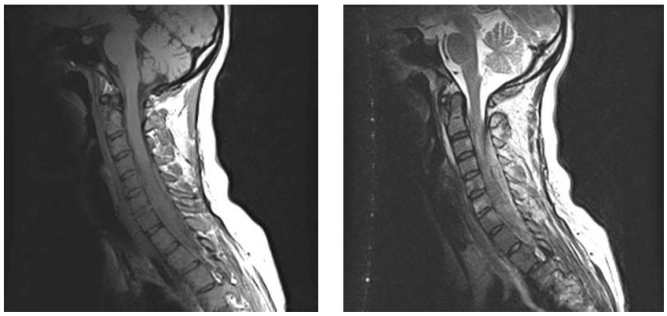


Fig. 1. MRI cervical spine minimally contrast enhancing, left sided anterolateral extra dural lesion extending from C2 – D1 level with maximum involvement around C4- C6 level with displacement and compression of cord involving the bone

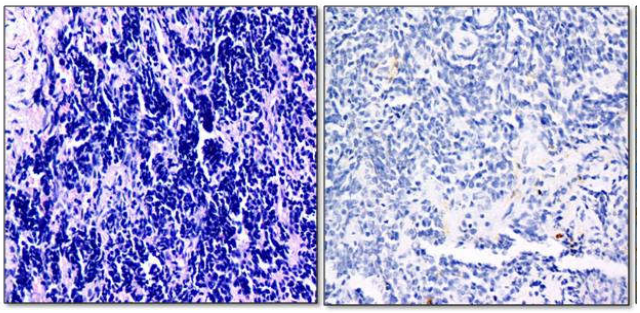


Fig 2 (a) Fig 2 (b)

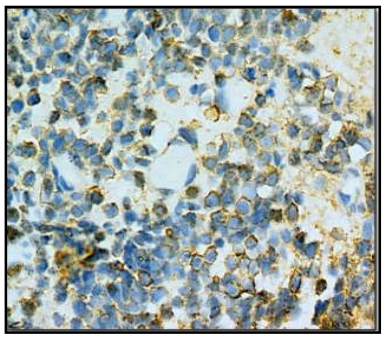


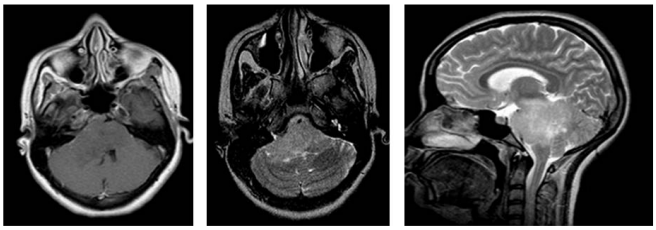
Fig. 2(c).

Fig. 2 (a). closely packed small round cells with scant cytoplasm, round nuclei with stippled chromatin, mitosis and pseudo rosettes, H&E, 400X. (b) Immunohistochemical staining for Leukocyte common Antigen (LCA) showing tumour cells are negative. (c) Tumour cells expressing strong membrane staining for the immunohistochemical marker CD 99

The patient was referred for radiotherapy and chemotherapy. On follow up, there was significant neurological improvement and no evidence of recurrence after three months.

Case 2

21 years young female came to the neurosurgery outpatient department with complaints of head ache in occipital region - 2 yrs. She also complained of decreased sensation in the right half of face associated with slurring of speech and decreased taste sensation. She had weakness of left upper & lower limbs and nasal regurgitation of fluids since one week. She denied history of hearing deficit, double vision and projectile vomiting. There was no preceding trauma or tuberculosis. On examination, patient's higher mental functions were normal. Bilateral papilloedema was present. She had right sided V, VI, VII, IX and X cranial nerve palsy and left hemiparesis. The cerebellar signs were positive on the right side. The working clinical diagnosis of a right sided cerebello pontine lesion was made. MRI brain showed a large lesion involving mid brain, pons & upper medulla with extension through cerebellar peduncles. The lesion was hypo intense in T1 weighted image and hyper intense in T2 weighted images.



Axial cut T1W Axial cut T2W Sagittal cut T2W

Fig. 3. MRI brain showing lesion involving Mid Brain, Pons & upper Medulla with extension through cerebellar peduncles

Through right retro mastoid – retro sigmoid approach the tumour was debulked after placing an external ventricular drain. Per operatively, the lesion was grayish pink, soft to firm in consistency, moderately vascular and seen in relation to pons and medulla, infiltrating into the right side cerebellum and growing around the cranial nerves. Histopathology of the lesion showed a highly cellular tumour involving the cerebellum composed of sheets of relatively uniform round to oval cells with scant cytoplasm, hyper chromatic to vesicular nuclei, stippled chromatin, occasional mitotic figures. Perivascular pseudo rosettes and Homer Wright rosettes were seen. Thin walled capillaries devoid of endothelial proliferation, hemorrhage & necrosis were noted in a fibrillary background.

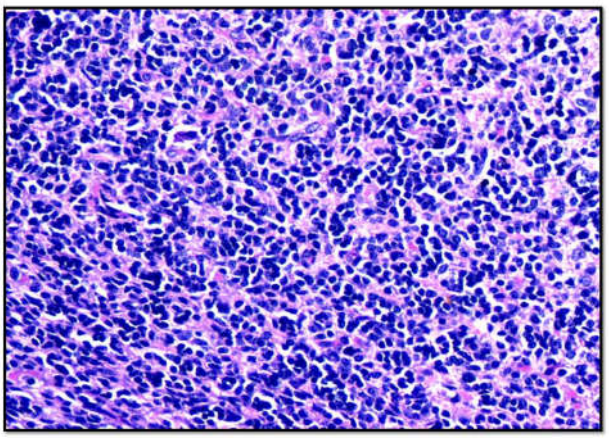


Fig. 4. Highly cellular tumour with rosettes in a fibrillary background, (H & E x 400)

Immunohistochemistry using a panel of markers revealed that the tumour cells showed strong membrane staining for CD 99, focally positive for Synaptophysin and negative staining with leukocyte common antigen (LCA). There were many cells showing peri nuclear cytoplasmic staining with Glial fibrillary acidic protein (GFAP) marker.

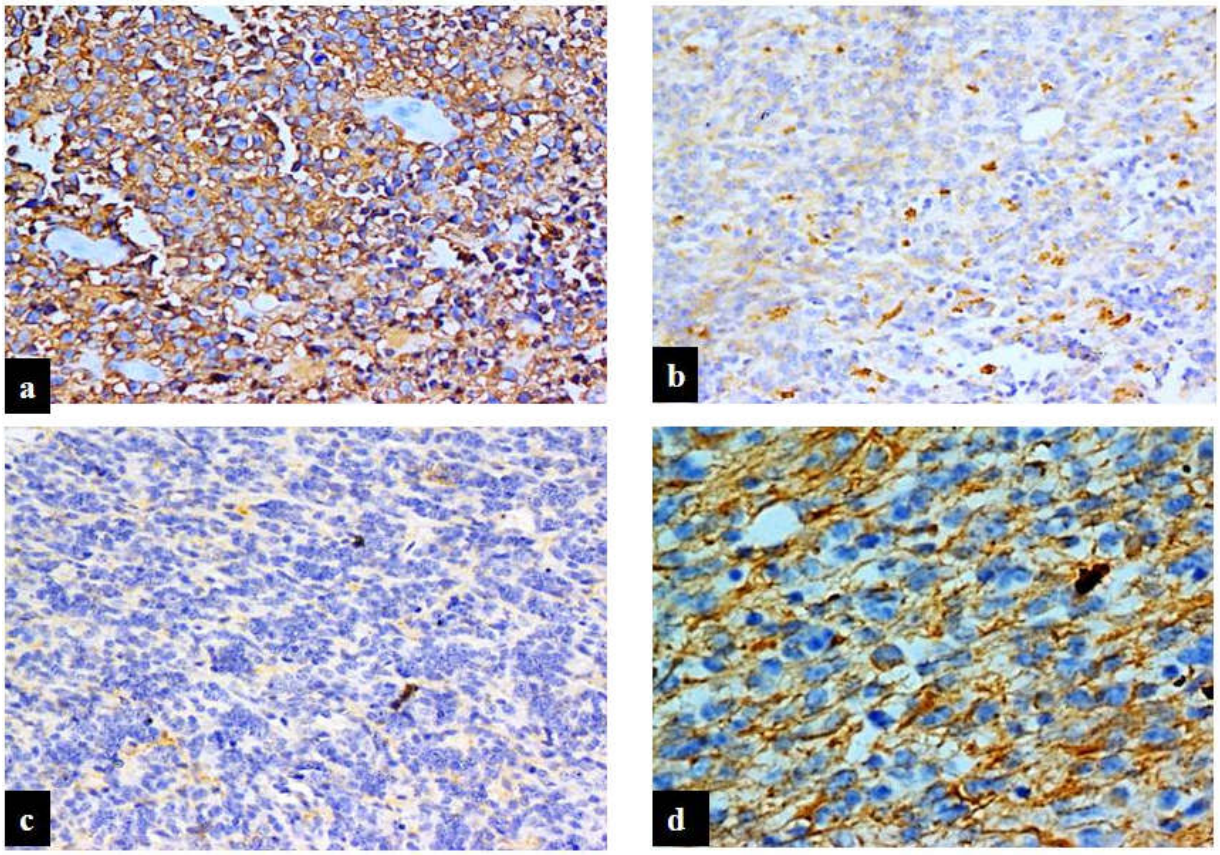


Figure 5 (a). strong membrane staining with CD 99, (b) focal positivity with Synaptophysin (c) negative for Leukocyte common antigen (d) peri nuclear cytoplasmic staing with Glial fibrillary acidic protein (GFAP)

A diagnosis of primitive Neurectodermal tumour with Neuroglial differentiation was made and patient was referred for chemotherapy.

DISCUSSION

Primitive Neuro Ectodermal Tumor (PNET) are the embryonal neoplasms outside the cerebellum, but morphologically similar to medulloblastomas (Chavit, 2001). These tumors that have chromosomal translocations identical to Ewing's sarcoma. They are highly aggressive embryonal tumors, manifesting preferentially in children. PNETs are derived from neoplastic transformation of common progenitor cells in the sub ependymal matrix layers. They most commonly occur in the cerebellum (medulloblastomas) but can arise in the pineal gland, cerebrum, spinal cord brain stem, and peripheral nerves (Michelle, 2006). Primitive neuroectodermal tumors frequently metastasize via the CSF pathways to the spinal and cranial subarachnoid spaces and are highly malignant both histologically and clinically. These tumors are composed of primitive small round blue cells forming Homer Wright & Peri vascular pseudo rosettes (Chen Jingyu, 2009). They have a capacity for divergent differentiation into neuronal, astrocytic, ependymal or retinal photo receptors (Kumar, 2007). Undifferentiated variety of PNETs carries a better prognosis than the differentiated ones. Expression of GFAP in PNETs has prognostic power comparable with the most significant clinical factors currently used to predict clinical outcome (Chavit Chantranuwatm, 2001). A diagnosis of Primitive Neuro Ectodermal tumor is usually made based on the morphological and immunohistochemical examination. PNETs in children and adults following chemotherapy, including alkylating agents (Marion, 2014), and radiotherapy as treatment for leukemias or lymphomas have been published. Other risk factors for PNET/EWS include a possible genetic predisposition. Those individuals with Li-Fraumeni syndrome with germline p53 mutations have an increased risk for primitive neurectodermal tumour. Spinal PNETs are mostly due to metastasis via CSF pathways from cranial origin. Primary spinal primitive Neurectodermal tumours are relatively rare (Michelle, 2007). They may be intramedullary, extramedullary or extradural in location. Extradural location is rare; only 4 such cases have been reported. Radical Surgery with combined radiotherapy and chemotherapy is the preferred treatment. In spite of multimodality management, the prognosis is poor. The causes of death in these patients include pneumonia, metastatic disease, aggressive local spread of the disease and progressive spinal cord involvement (Tommasi, 2006). Some early reports had suggested that adults with Ewing's sarcoma/PNET have a less favorable outcome than children, but a study of adult patients performed in the Department of Cancer Therapeutics, Institute of Cancer Research, Sutton, United Kingdom (Volkan Aydin, 2004), has suggested that adults may have a similar outcome to children.

Conclusion

Primitive neurectodermal tumours are rare but have a highly aggressive clinical course with a poor outcome in spite of multimodality treatment options (Chen Jingyu, 2009). PNETs in adults are extra ordinarily rare. Histopathological, immuno histochemical and molecular studies are important for correct diagnosis. Our case 1 was a primary extra dural spinal primitive neurectodermal tumour which has been rarely reported in the literature. So spinal PNET has to be thought of

as a differential diagnosis for any spinal lesion with rapidly progressive course (Janss, 1996). Our case 2 was a primitive neurectodermal tumour with neuroglial differentiation which imparts a poor prognosis. This case report emphasizes the many faces of this aggressive tumour and the need for research towards a better theranostic approach (WHO classification of CNS tumours, IARC 2016).

REFERENCES

Ba, L., Tan, H., Xiao, H., Guan, Y., Gao, J., Gao, X. 2014. Radiologic and clinicopathologic findings of peripheral primitive neuroectodermal tumors. Acta Radiol 55(6): 515-523.

Chavit Chantranuwatm, A. 2001. Peripheral Primitive Neuroectodermal Tumor with Neuronal and Glial Differentiation: Report of a Case Arising in Suprarenal *Region. J Med Assoc Thai* 84(4): 584.

Chen Jingyu, Song Jinning, Meng Hui, Feng Hua. Intraspinal primitive neuroectodermal tumors: Report of four cases and review of the literature. *Neurol India* 57 (5): 661-668, 2009

Janss, A.J., Yachnis, A.T., Silber, J.H., Trojanowski, J.Q, Lee, V.M., Sutton, L.N., Perilongo, G., Rorke, L.B., Phillips, P.C. 1996. Glial differentiation predicts poor clinical outcome in primitive neuroectodermal brain tumors. Ann Neurol 39(4):481-9.

Kumar, R., Reddy, S.J., Wani, A.A., Pal, L. 2007. Primary spinal primitive neuroectodermal tumor: case series and review of the literature. Pediatr Neurosurg 43(1):1-6.

Marion Cole, E. F,G,Shobha Parajuli,,E,F,Douglas Laske,Lori Goldstein, E, Peripheral primitive neuroectodermal tumor of the dura in a 51-year-old woman following intensive treatment for breast cancer. *Am J Case Rep*., 10; 15: 294-9, 2014

Michelle Scurr, Ian Judson. How to Treat the Ewing's Family of Sarcomas in Adult Patients. The Oncologist 11; 65–72, 2006

Tommasi, A., De Tommasi, C., Occhiogrosso, G., Cimmino, A., Parisi, M., Sanguedolce, F. and Ciappetta, P. Primary intramedullary primitive neuroectodermal tumor (PNET) – case report and review of the literature. *European Journal of Neurology*, (13), 240–243, 2006

Virani, M.J., Jain, S. 2002. Primary intraspinal primitive neuroectodermal tumor (PNET): a rare occurrence. Neurol India 50(1),75-80.

Volkan Aydin. M. 2004. Primary Primitive Neurectodermal tumor within the spinal epidural space: report of a case and review of the literature. Neurological Research, 26(7),774-777(4).

WHO, Classification of Tumours of the Central Nervous System. IARC 2016. Edition: 4; The International Agency for Research on Cancer; Edited by D.N. Louis, H. Ohgaki, O.D. Wiestler, W.K. Cavenee.

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