



PARASPINAL NEUROBLASTOMA IN ADOLESCENT GIRL- A CASE REPORT

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ABSTRACT

A 17 year old Indian female presented with lower backache since 2 months. MRI was done which revealed intradural extramedullary space occupying lesion at D12- L1 level with extension into neural foramina causing its widening, marked cord compression and compressive myelopathic changes. Histopathology of excision biopsy revealed histological diagnosis of Neuroblastoma. Immunohistochemistry confirmed the diagnosis as Synaptophysin and NSE were positive. Bone marrow aspiration and Chest X Ray did not reveal metastases. Generally neuroblastoma is a localized disease but in adults it is more aggressive. Neuroblastoma is the most common malignancy in children less than one year of age, but is rare in adults. In adults, classical features are lacking.

KEYWORDS: Adolescent, paraspinal, neuroblastoma.

INTRODUCTION

Neuroblastoma originates from embryonal neural crest cells involving the peripheral sympathetic nervous system, is a common, extracranial malignant tumor in children. 90% neuroblastoma are seen in children younger than 5 years age.^[1,2] It is rarely seen after age of 20 years. In adults it presents in abdomen and paravertebral region. It can arise anywhere in the sympathetic nervous system and frequently occurs in the adrenal gland or sympathetic ganglia in a paraspinal location in retroperitoneum or chest. It metastasizes to regional lymph node, liver, skin, brain, bone and bone marrow. Neuroblastoma are morphologically heterogeneous tumors with some tumors undergoing spontaneous differentiation.^[3,4,5] Approximately 45% of neuroblastomas are high risk with 6.1% over 20 years of age.^[6]

CASE REPORT

A 16 years Indian female presented with abdominal and back pain since 2 months duration with weakness in left lower limb since 7 days. There was tenderness at D12-L1 level. MRI was done which revealed large ill defined heterogeneously enhancing intradural extramedullary space occupying lesion, measuring approximately 5.3 x 6.7 x 7 cm in lower dorsal/upper lumbar region extending from D12 to L1 / 2 levels. The lesion was hyperintense on T2/TIRM. The lesion was causing marked compression and right antero-lateral displacement of dorsal cord with intramedullary T2 hyperintensity-compressive myelopathic changes. Laterally, the lesion was extending into the left lateral

recess / neural foramen of D12/L1 vertebrae, causing its expansion and involving left para vertebral region. There was no evidence of vertebral body scalloping. [Fig1].



Fig 1: MRI showing intradural extramedullary lesion at D12 – L1 level.

Excision Biopsy was done and subjected for histopathological examination. Grossly it showed a greyish white soft tissue mass with encapsulation. Histologically, the section showed small round cells with hyperchromatic to vesicular chromatin nucleus, scanty

fibrillary cytoplasm forming pseudorosette (Homer-Wright rosettes). Background showed fibrillary material. Histopathological diagnosis of Malignant Round Cell tumor of neural origin – Neuroblastoma was made. [Fig 2]. Paraffin Block was subjected for Immunohistochemistry for confirmation of diagnosis.

IHC of tumor showed NSE (neuron Specific Enolase) [Fig 3], and Synaptophysin [Fig 4] positivity, while tumor was negative for CD-99 [Fig 5], CD-45 [Fig 6], and WT-1. Thus histological diagnosis of Neuroblastoma was established by IHC.

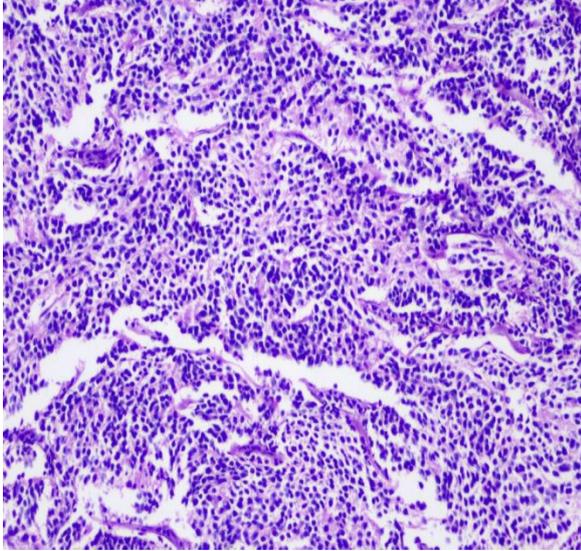


Fig 2. H & E stain, showing round cells in rosettes in fibrillary background.

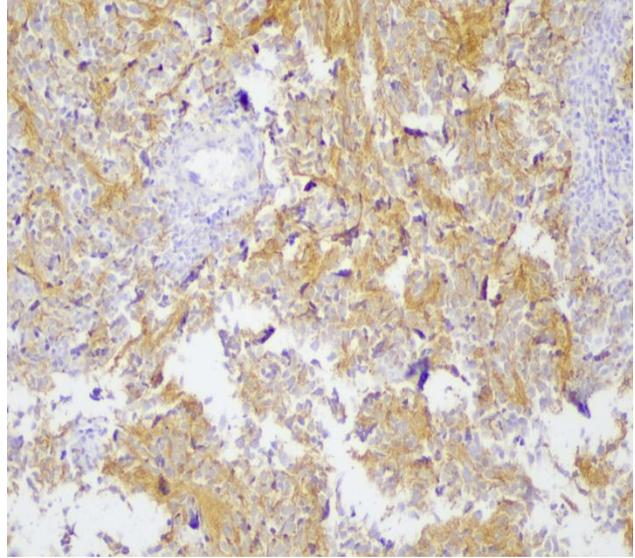


Fig 3. NSE, 40 X, showing positivity.

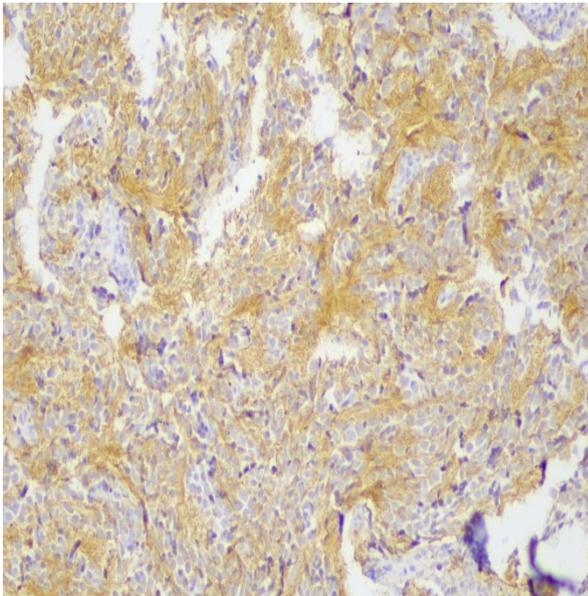


Fig 4. Synaptophysin, 40 X showing positivity.

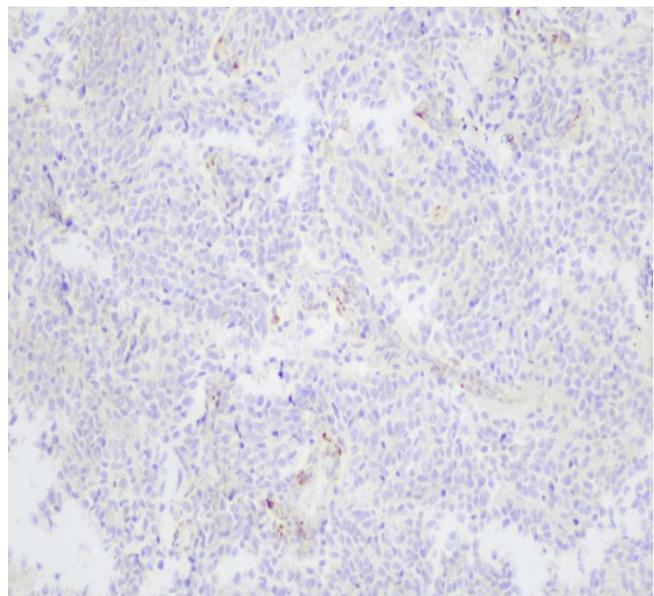


Fig 5. CD99, 40 X showing negativity.

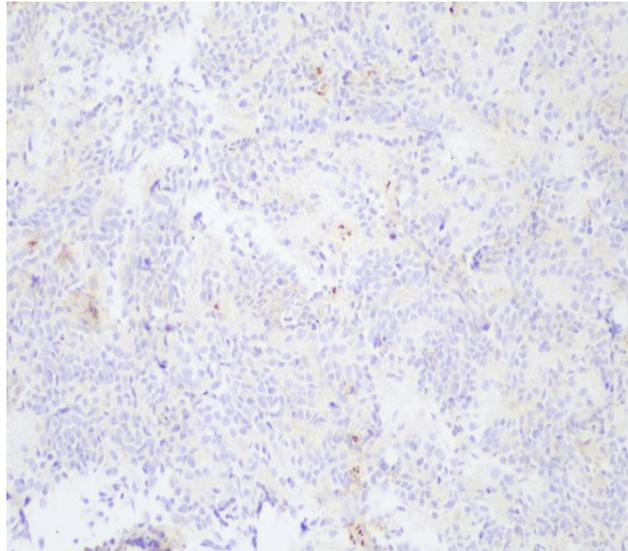


Fig 6. CD 45, 40 X showing negativity.

CT scan of chest and abdomen were done which showed lungs, liver, other organs were free from tumor. Bone marrow aspiration also did not reveal presence of tumor cells. Chemotherapy was started.

DISCUSSION

Neuroblastoma primarily affects children, 90% diagnosed under age of 05 years.^[7] Spinal involvement of neuroblastoma constitutes 20-30% of all pediatric extradural spinal tumors^[7] while in adolescents and adults spinal involvement of neuroblastoma is extremely rare with limited number of cases.^[8,9,10] In children neuroblastoma can develop within the adrenal medulla or sympathetic ganglion. In adults, it is more frequently seen in the abdomen, pelvis, thoracic, spinal cord, mediastinum and olfactory region.^[10,11] Tumors along the spinal cord can result in compression, paraplegia and pain. It metastasizes to bone, bone marrow, lung, brain, breast, lymph nodes, liver and pleura.^[10,12] Adolescent Neuroblastoma have poor prognosis.^[8,13] The most important positive prognostic factors are low age group and the stage of the disease at the time of diagnosis. It spreads through vascular and lymphatic route. In adults brain and lung metastases is frequent. There is no standard treatment for Neuroblastoma in adolescents and adults. Complete resection combined with chemotherapy and radiotherapy should be administered. Paraspinal tumors in the thoracic, abdominal and pelvic regions are able to invade the spinal canal through the neural foramina (dumbbell tumor) causing spinal cord compression and cause symptoms related to compression of nerve roots and spinal cord. Symptoms include paraparesis, paraplegia, bladder or bowel dysfunction or radicular pain. Although Neuroblastoma is rare in adults and adolescents, it is important to differentiate from small round blue cells tumors. Differential diagnosis includes lymphoma, Ewing's Sarcoma/PNET. Rhabdomyosarcoma, Osteosarcoma. CD 99 differentiates Neuroblastoma from PNET, CD 45 is used to differentiate from lymphoma. Neuroblastoma

histology is classified according to International Neuroblastoma Pathologic Committee Classification (INPCC) which ranks tumor histology as favourable or unfavourable. Based on differentiation of tumor cells as characterization of stroma and mitosis-karyorrhexis index.^[14,15] Unfavourable histology has poor prognosis. Our patient had differentiated tumor with favourable histology.

CONCLUSION

Paraspinal Neuroblastoma is rare in adolescents and adults. Histopathological diagnosis has 100% accuracy in diagnosing Neuroblastoma. It should be kept in Differential Diagnosis of Paraspinal lesions in adolescent and adults.

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