ORIGINAL RESEARCH PAPER
VOLUME-6 | ISSUE-7 | JULY-2017 | ISSN No 2277 - 8179 | IF : 4.176 | IC Value : 78.46
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

PRIMARY INTRASPINAL PRIMITIVE NEUROECTODERMAL TUMOR (PNET): A RARE OCCURRENCE IN ADULT FEMALE-CASE REPORT

Medical Science

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ABSTRACT

Primitive neuroectodermal tumors (PNETs) are malignant, poorly differentiated neoplasms derived from the neural crest. Only a few cases of primary intraspinal PNETs have been reported in children and in adults. Here, we present a case study of a 44-year-old female with L3 intradural PNET grade IV, including details of her examination, surgical procedures applied, histological findings and her subsequent treatment.Post operative neurological improvement was minimal. Primary intraspinal PNETs are rare tumors and carry a poor prognosis.

KEYWORDS:

Spinal cord, Primitive neuroectodermal tumor (PNET)

Introduction

As for the World Health Organization (WHO) classification of brain tumors, all undifferentiated (primitive) tumors are derived from common neuroepithelial cells and are thus classified under primitive neuroectodermal tumors (PNET).1 Hart and Earle were the first to introduce the term PENT in 1973.2 These tumors are more common in children. There is considerable confusion and controversy in the classification of PNETs.3

Central PNETs (cPNET) develop from the central nervous system (CNS) and peripheral PNETs (pPNET) putatively from the neural crest. Both cPNET and pPNET, are aggressive tumors and have similar survival rates. However they differ in their clinical presentation and spread pattern.4 They also have distinct immunohistochemical profiles and clinical evolution and treatment protocols also differ. PNETs are mainly intracranial, predominantly in the cerebellum. 5Intraspinal PNETs are extremely rare. A review of the literature shows that only 19 cases of primary intraspinal PNETs have been reported to date and the present case is intratradural in location.6 In this article, we present a rare adult patient with a primary intradural PNET and describe his treatment regimen and outcome.

Case report

A 44 years old female from Bihar, India presented to the neurosurgery department, VPIMS,lucknow in December 2016 after experiencing radicular pain of both lower limbs for last 6-7 months along with progressive increasing weakness with difficulty in ambulation since 2 months. She became bedridden since 1.5 months. Numbness in both lower limbs, urinary straining and constipation was present since last 1.5 months.

Neurological Examination

Tone: Normal, Power : Upper Limb: B/L 5/5, Lower Limb: Hip (Abd/add-1-2/5), Knee (Flex/Exten-0/5), Ankle (DF/PF:1/5), Plantar: B/L mute. Sensory: Decreased pinprick sensation in L2-3 distribution by 10-15%, saddle hypoesthesia present. Cranial nerve examination was normal.

Investigations

All the routine blood investigation was within normal limit.
MRI findings revealed well defined intradural intramedullary mass lesion measuring 16x1x13 mm in size seen at lumbar canal at L3 vertebral level. The lesion showed heterogenous enhancement in post contrast images. Diagnosis ? ependymoma, meningioma,nerve sheath tumor was given (figure 1)

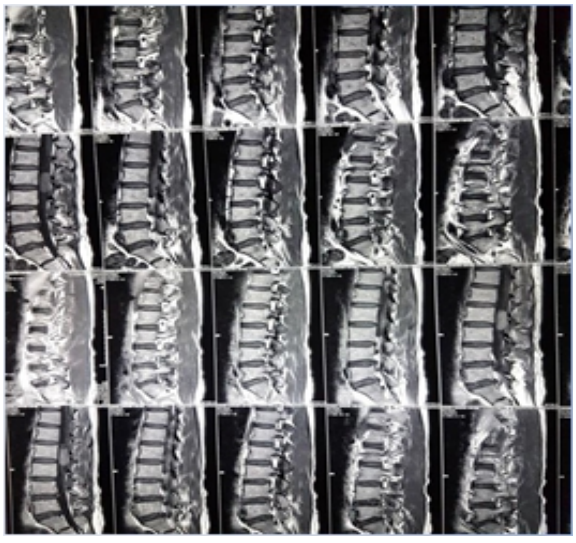

Figure 1: MRI Shows well defined intradural extramedullary tumor at the level of L3- l4 Vertebrae. On T1- weighted Image hypointense and on contast Heterogenously enhance.

Surgery

L3-L4 laminectomy with gross total excision of tumor was done .Soft greyish white, non vascular, partly suckable, ill defined tumor was seen engulfing multiple roots below conus. Tissue sent for HPE. Wound closed in layers after hemostasis.

Pathology

Tissue specimen revealed sheets of undifferentiated -appearing cells closely arranged in cords, nests and sheet. Cells are highly pleomorphic with dense hyperchromatic nuclei, irregular nuclear membrane and an ill-defined cell borders. Radial arrangement of tumor nuclei around small tangles of fibrillar eosinophilic material is seen (Homer-Wright rosette). Both typical and atypical mitotic figures and numerous apoptotic bodies are evident. There are massive areas of necrosis. Final diagnosis of PNET grade IV was made (figure 2a and 2b)

Postoperative status

Postoperative status was uneventful. The patient showed little improvement in radicular pain and numbess of both lower limbs. Motor status was same as preoperative period. Physiotherapy was done regularly. Radiotherapy was adviced ..

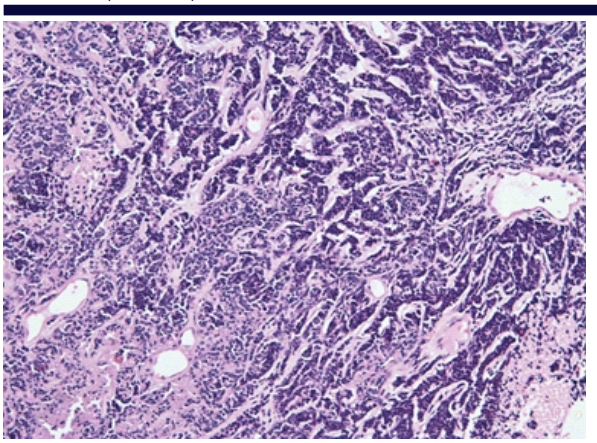

Figure 2a: CNS PNET with neuronal differentiation ; nodular architecture with streaming of tumor cells (H&E 10X)

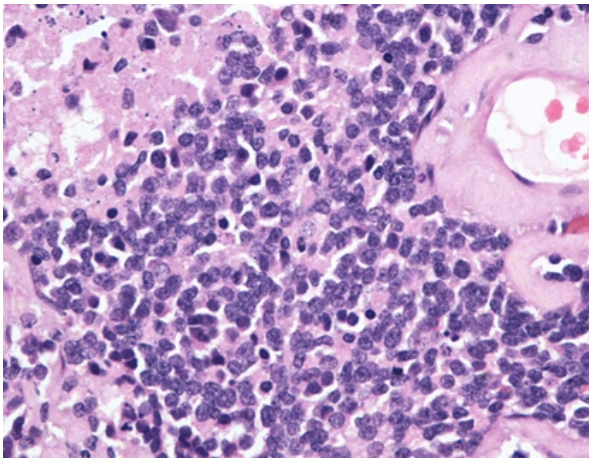

Figure 2b: Cells with round to oval hyperchromatic nuclei, indistinct cytoplasmic margins (H&E 40X)

Discussion

PNETs was first described by Bailey and Cushing in 1925, and were also called spongioblastomacerebelli. PNETs can occur outside the brain and throughout the body, as peripheral neuroblastomas and ewing sarcomas. PNETs even though found in both children and adults are more common in children.[6]

With regard to PNET involving the spinal cord, the most common are drop metastases from primary intracranial tumors, which disseminate via cerebrospinal fluid.7Primary intraspinal PNETs are rare. These neoplasms can be extradurally, extramedullary, intra-extramedullary and purely intramedullary.[7]

In 1992, a literature review by Ogasawara et al 8 revealed 13 cases of spinal PNET. These tumors tend to occur in young males (approximately 3:1 male:female ratio, with 50% under 15 years and 85% under 30 years of age). The sites involved include the cauda equina and lower lumbar region (8/14 or 57%), the cervical cord (3/14 or 21%), and the thoracolumbar area (2/14 or 14%). Our case was unusual in that it occurred in a woman, was located at the lumbar cord level, and, of particular interest, was predominantly extramedullary in location.The differential diagnosis of a solid lumbar intradural, extramedullary mass consists of nerve sheath tumors (ie, schwannoma, neurofibroma), meningiomas, and metastases .

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.9

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%.[6]

After surgical excision, the role of adjuvant treatment modalities such as radiotherapy and chemotherapy is unknown. However, primary spinal cord PNETs have a propensity to metastasize along the subarachnoid space as well as to distant sites, including the bone marrow, lungs, and pleura. Therefore, entire neuroaxis irradiation should be included as part of the initial treatment strategy.[10]

The role for adjuvant multiagent chemotherapy is unclear. In one review of the reported primary intraspinal PNETs to date, chemotherapy as part of initial therapy did not appear to affect subarachnoid dissemination, distant spread, or survival; however the number of patients was too small to be conclusive.[11]

As regard to new treatment strategies are concerned,role of peripheral blood stem cell transfusion(PBSCT) is suggested in chemosensitive tumors or in cases where the patient has remissions. PBSCT after remissions prevents relapse. A trial has been conducted at Hinduja hospital, Mumbai, India, where PBSCT was employed in 21 year old male with PNET of chest wall-stage-IV. More studies are required to explore the role of PBSCT in improving the survival in these patients.[12]

In conclusion, purely intramedullary PNETs (IPNETs) are uncommon tumors affecting children and young adults.They are characterized by recurrence, progression or intracranial dissemination. Outcome is dismal: most patients die within two years in spite of surgical resection followed by radiotherapy and chemotherapy. Every case contributes to our understanding of their treatment and prognosis.

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