Primary Cervical PNET mimicking as neurofibroma. A case report

P Ghodgaonkar, V Velho, R Mally, A Jaiswal

Citation

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Abstract

Primitive Neuro Ectodermal tumor (PNET) are the lesions commonly occurring in intracranial locations, more often in children and young adult. These are aggressive and highly malignant tumors which may spread through CSF to the spinal cord. Occurrence of primary spinal PNET is rare. In the cord these can arise at any level and may be intramedullary, extramedullary or extradural in location. The appearance on MR studies may mimic a neurofibroma with intra and extradural components. The management includes surgical excision, radiotherapy and adjuvant chemotherapy. Despite good surgical excision, due to the aggressive nature of the tumor recurrence is likely and the life expectancy remains poor.

An 18 year old right handed male presented with the complaint of pain in the neck and progressive weakness in both the upper and lower limbs more on left then right side of 4 month duration. Examination revealed wasting of the deltoid, supraspinatus and infraspinatus muscles on left side. Tone was increased in both lower limbs and the reflexes were brisk. Power was grade IV/V in RUL and RLL and III/V in LUL and LLL. Contrast MRI showed an extramedullary intradural mass of 3.4x1.2x1.7 cms in the spinal canal at C2 – C3 level compressing the cord and was extending to the C2 C3 neural foramen and thought to be neurofibroma with a major extradural component arising from the nerve root foramen. The patient was operated via C1-C4 laminectomy. Intra operative inspection revealed a large, vascular and firm tumor which had a major extradural component and was adherent to the left vertebral artery. The dura was opened and the intra and extradural components were excised. The intra foraminal portion of the tumor, which was adherent to the left vertebral was left. Histopathological examination revealed a highly cellular tumor which was infiltrating the surrounding meningeal and fibrocollagenous tissue with tumor cells arranged in small sheets suggestive of Primitive Neuro Ectodermal tumor (PNET). The patient did well after surgery and his power in upper and lower limbs improved to grade V on both the sides. A contrast CT scan of brain did not reveal any primary lesion in the brain thus ruling this out as a secondary lesion in the spine. The patient was advised adjuvant radiotherapy and chemotherapy, but was unable to take the treatment due to financial constraints. On follow up at the end of 4 months he was doing well. 6 months later he presented to us again with recurrence of progressive weakness in upper and lower limbs, as well as difficulty in breathing. A recurrence of the tumor was suspected and MRI of the cervical spine was repeated. This time it revealed two lesions; one at the previous (C2-3) level and the second lesion at the foramen magnum, which was compressing the cervico medullary junction anteriorly and encasing both vertebral arteries, which was a new development. The patient was re operated and both the lesions were excised by a posterior midline approach. The patient showed improvement post operatively with improvement in power and respiration. He was referred to the medical oncology department for chemotherapy.

Figure 1

Fig1. Preoperative post contrast sagital MR showing an intradural lesion with extradural extension at C2 C3 level

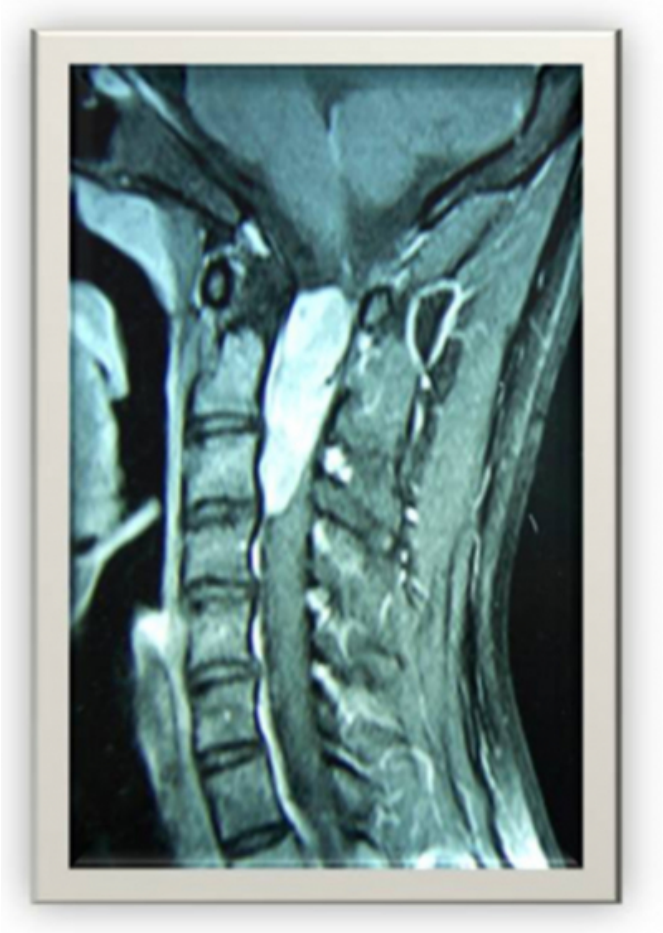


Figure 2

Fig 2. Post operative MR showing the residual tumor.

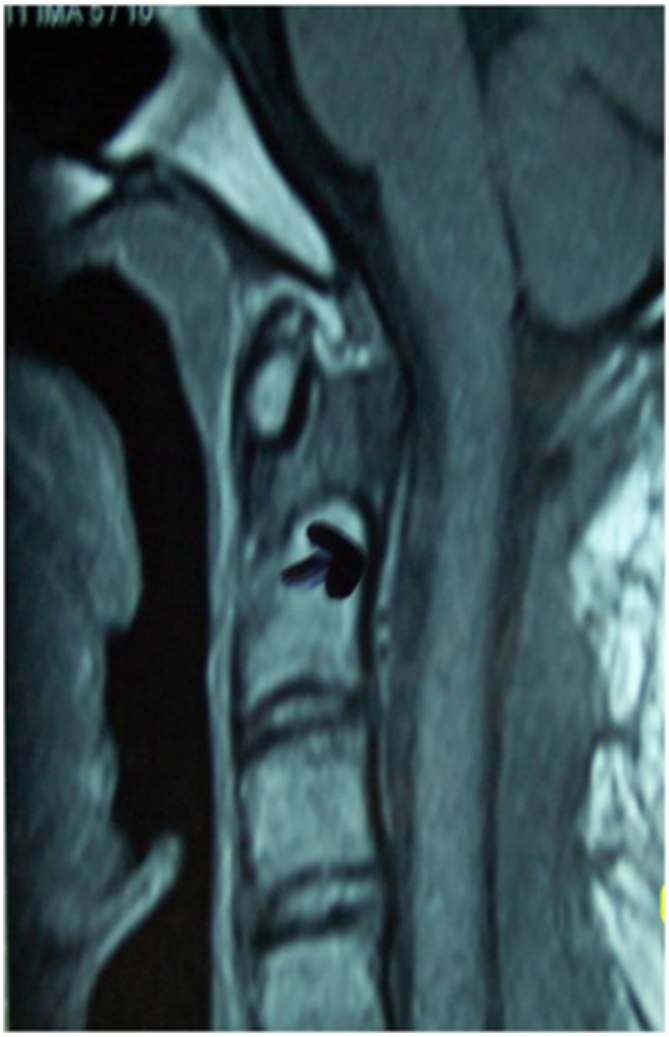
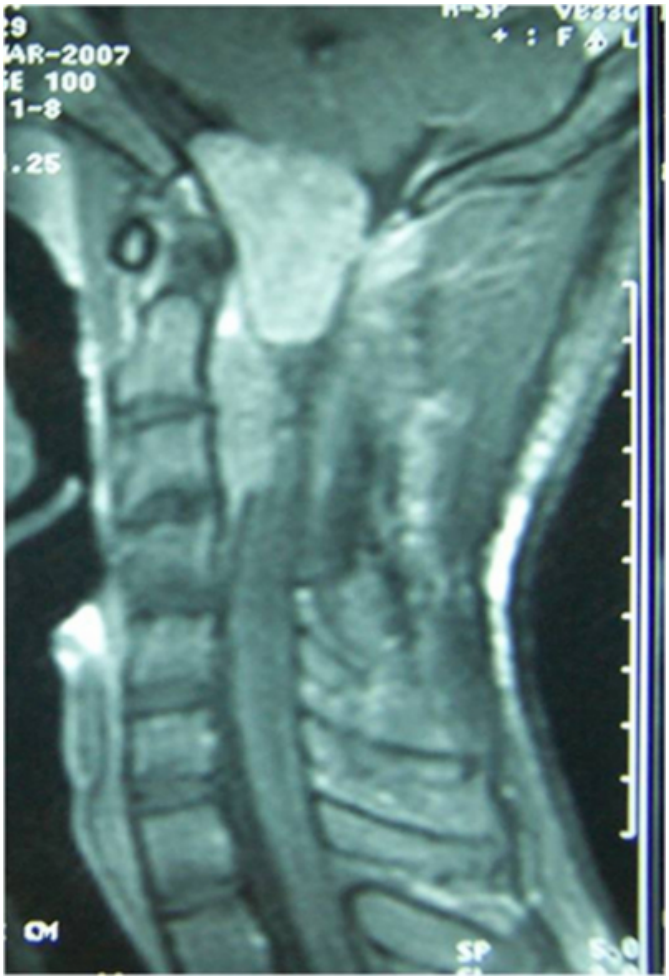


Figure 3

Fig 3. Contrast MR at 10 months shows recurrent tumor at C2 C3 level and a new lesion at cervico medullary junction.



DISCUSSION

Primary Neuro Ectodermal tumors commonly occur intracranially and are seen more in children and young adults (3). These are aggressive tumors and have a rapid rate of growth (1). They tend to spread throughout the central nervous system via the cerebrospinal fluid. Primary intra spinal primitive Neuro Ectodermal tumors are not common and very few cases have been reported in literature (1 & 3). These tumors may arise at any level in the spinal cord and could be intramedullary, intradural extramedullary or extradural in location (2 & 3). This patient had a tumor at C2-3 level which mimicked a neurofibroma with an extradural and intradural component on MR studies. Within 10 months of the previous surgery the patient developed a recurrence at the previous site as well as a new lesion in intradural extramedullary location with intramedullary component encasing the vertebral arteries and the medulla. Of the 14 cases mentioned in literature 6 cases showed origin from the Cauda equina region (1, 2, 4 & 5). In this case the tumor was in the high cervical region without any primary involvement of the brain.

PNET arise from neoplastic transformation of primitive neuroepithelial cells in subependymal zones (5). Subependymal zones are present in all areas of the central nervous system and may explain the presence of PNET at locations other than the cerebellum (1). Spinal PNET appear to be more common in adults rather than in children in contrast to intracranial PNETS that predominate in children.

Management of PNET includes surgery, radiotherapy and adjuvant chemotherapy (3). Our patient did not receive radiotherapy & chemotherapy, in spite of advice for personal reasons and thus resulted in an early recurrence with a new lesion within a short span of 10 months. Total excision is at times difficult as these tumors encase blood vessels and get their vascularity from these major vessels. The prognosis for PNET is poor with a life expectancy period of 2 years. Death usually results from metastatic disease, aggressive local spread of tumor and progressive spinal cord involvement.

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Author Information

Pranav Ghodgaonkar, M Ch
Assistant Professor, Department of Neurosurgery, Grant Medical College and Sir J J Group of Hospitals

Vernon Velho, M Ch
Associate Professor, Department of Neurosurgery, Grant Medical College and Sir J J Group of Hospitals

Rahul Mally, M Ch
Assistant Professor, Department of Neurosurgery, Grant Medical College and Sir J J Group of Hospitals

Amit Jaiswal, M Ch
Assistant Professor, Department of Neurosurgery, Grant Medical College and Sir J J Group of Hospitals