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CASE REPORT

Primitive Neuroectodermal Tumor (PNET) at the Level of Thoracic Spine: A Case Report

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Abstract

Introduction Primitive neuroectodermal tumors (PNET) are malignant tumors thought to arise from the neural ectoderm and comprise of undifferentiated small round cells that affect soft tissue and bone. These tumors occur primarily in children and adolescents (thirteen to sixteen years old), with a tendency to recur quickly and metastasize especially to lungs, bone marrow, brain and lymph nodes. PNET are uncommon malignancies that are increasingly reported in the literature and located in the spine are extremely rare. In this study we present a case of 14 years old girl with PNET of the thoracic spine which recur after initial resection. Case Report a 14 years old girl presented with low back pain and progressive weakness of both of lower limbs of three months duration. On physical examination, motor and sensory function was lost, motoric strength in the lower limbs was grade 1/5 with hypoesthesia below L1 level. Magnetic resonance imaging (MRI) of the spine showed burst pathologic fracture of the 12th thoracic spine with suspicious malignant lesion in the 12th thoracic spine. Initially the condition was treated with laminectomy, decompression, stabilization and open biopsy. Histopathology and immunohistochemistry of the lesion revealed the diagnosis of PNET. During the early postoperative course, she had instance neurological recovery and was able to walk unassisted. But, 1 month later the neurologic deficit recurred and she was treated with extended laminectomy with tumor debulking. Then the patient received chemotherapy and planned for radiotherapy. Conclusion Primitive neuroectodermal tumors (PNET) are extremely rare and are aggressive with poor outcomes. Good modalities play a significant role in the identification of the feasibility of surgical excision of the tumors, detection of distant metastasis, and evaluation of the efficacy of therapeutic interventions.

Keywords: *Primitive neuroectodermal tumor (PNET), Malignant Tumor, Rare Tumor, Spinal Tumor.*

Introduction

Primitive neuroectodermal tumors (PNET) are malignant tumors thought to arise from the neural ectoderm and comprise of undifferentiated small round cells that affect soft tissue and bone. These tumors occur primarily in children and adolescents (thirteen to sixteen years old), with a tendency to recur quickly and metastasize especially to lungs, bone marrow, brain and lymph nodes.

PNETs can be classified as central PNETs (cPNETs) or peripheral PNETs (pPNETs) depending on the site of presentation. It was first coined for a group of embryonal tumors located in the central nervous system (cPNET). More recently, the PNET concept has been expanded to include histologically similar, peripherally located tumours, referred to as peripheral PNET's (pPNET's). Among the cPNET's are medulloblastoma, pineoblastoma, cerebral neuroblastoma, ependymoblastoma, medulloepithelioma, primary rhabdomyosarcoma, and atypical teratoid / rhabdoid tumour. The pPNET is part of the Ewing's sarcoma family of tumours, which includes Ewing's sarcoma of bone, extraosseous Ewing's tumour and primitive neuroectodermal tumour. Various synonyms including peripheral neuroepithelioma, peripheral neuroblastoma, and Askin tumour have been used to describe peripherally occurring primitive neuroectodermal tumours. PNET are uncommon malignancies that are increasingly reported in the literature and located in the spine are extremely rare. In this study we present a case of 14 years old girl with PNET of the thoracic spine which recur after initial resection.

Case Report

A 14 years old girl presented with low back pain and progressive weakness of both of

lower limbs of three months duration. She was bedridden before the admission. On physical examination, motor and sensory function was lost, motoric strength in the lower limbs was grade 1/5 with hypoesthesia below L1 level. Magnetic resonance imaging (MRI) of the spine showed burst pathologic fracture of the 12th thoracic spine with suspicious malignant lesion in the 12th thoracic spine.

Initially the condition was treated with laminectomy, decompression, stabilization and open biopsy. Histopathology and immunohistochemistry of the lesion revealed the diagnosis of PNET. During the early postoperative course, she had instance neurological recovery and was able to walk unassisted. But, 1 month later the neurologic deficit recurred and she was treated with extended laminectomy with tumor debulking. Then the patient received chemotherapy and planned for radiotherapy. Treatment of the patient is continuing with a multidisciplinary approach with collaboration of orthopedics, pediatric and radiologic departments.

Discussion

Peripheral primitive neuroectodermal tumour (PNET) are small round cell tumours occurring in bone and soft tissues, characterized as a group by the presence of the typical translocation and its variants. Pathological characteristics ; routine hematoxylin and eosin (H-E) staining and light microscopy manifested uniform, small round tumor cells with little cytoplasm. These cells mainly had round or oval nuclei and were arranged in a pattern of Homer-Wright Rosettes.

PNETs can be classified as central PNETs (cPNETs) or peripheral PNETs (pPNETs) depending on the site of presentation. Peripheral PNET and Ewing sarcoma are closely related, and the distinction between these two entities has become blurred. PNET / Ewing sarcoma is the most common tumor of the chest wall in children and adolescents. PNETs can be distinguished from Ewing's sarcoma due to their neural differences detected by immunohistochemistry. pPNETs express at least two neural differentiation antigens, whereas Ewing's Sarcoma expresses only one antigen or sometimes no antigens. In addition, Homer-Wright Rosettes can be found in pPNETs but not in Ewing's sarcoma using light microscopy. The literatures hows that congenital and neonatal tumors are rare, accounting for only 2% of all childhood malignancies.

Specifically, chest wall tumors are also a rare occurrence, comprising only 1.8% of all solid childhood tumors. There are multiple causes of these chest wall tumors, but the most common is the PNET / Ewing sarcoma / Ask in tumor family, which comprises 71%. Primitive neuroectodermal tumors (PNET) are devastating malignancies that appear to be more common than has previously been reported.

The clinician should give consideration to the possibility of this diagnosis, especially in the setting of a young adult or child with an intraspinal mass. While there are no widely accepted standards for the management of spinal PNETs, it is advocated that complete resection is performed when possible with the goals of neurological stabilization and obtaining sufficient tissue for accurate diagnosis. This should then be followed by an individualized combination of chemotherapy and/or radiation.

Conclusion

Primitive neuroectodermal tumors (PNET) are extremely rare and are aggressive with poor outcomes. The imaging features included osteolytic bone destruction and an apparent soft-tissue mass, whereas little evidence exists of periosteal reaction, calcification, or ossification. Diagnosis requires histopathological examination and immunohistochemistry such as NSE, S-100, LCA and CD 99. In addition, these modalities play a significant role in the identification of the feasibility of surgical excision of the tumors, detection of distant metastasis, and evaluation of the efficacy of therapeutic interventions.

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