CASE REPORT

A primary dorsal osseous Ewing's sarcoma hidden as acute paraplegia in a 7-year child: A Case report

How to Cite This Article: Sameer Yahya F, Hieder A, Al- Shami H, A primary dorsal osseous Ewing's sarcoma hidden as acute paraplegia in a 7-year child: A Case report. Iran J Child Neurol. summer 2022; 16(3): 219-222

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Received: 05-Nov-2020
Accepted: 16- Jan-2021
published:16- Jul-2022

Abstract

Ewing's sarcoma is a highly malignant bone tumor commonly affecting the diaphysis of long bones and pelvic bones. It belongs to a spectrum of neoplastic diseases known as the Ewing sarcoma family of tumors (EFT) with the same histologic, immunohistochemical characteristics and chromosomal translocation. It commonly affects children between the ages of 5-15 years. Spinal involvement might be primary or metastatic. Primary spinal involvement is infrequent. We report a 7-year female with acute paraplegia caused by dorsal osseous Ewing's sarcoma that her presentation mimics other causes of acute lower limb weakness in children. Early detection of this rare case and prompt treatment will ensure better outcomes for the patient.

Keywords: Ewing's sarcoma; spinal compression; acute paraplegia; bone tumor; pediatrics

DOI: 10.22037/ijcn.v16i2.32783

Introduction

Primary bone tumors form 6% of childhood malignancies. Ewing's sarcoma is the second most common bone tumor in children after osteosarcoma. It belongs to a spectrum of neoplastic diseases known as the Ewing sarcoma family of tumors (EFT) with the same histologic, immunohistochemical characteristics and chromosomal translocation. It peaks in late teenage groups with a slight male predominance. It arises commonly from the diaphysis of long bones and pelvis. Spinal Ewing's sarcoma might be primary or metastatic. Primary spinal Ewing's sarcoma is rare (8%) (1,2).

Ewing's sarcoma might be presented as a localized painful mass or constitutional symptoms like fever, weight loss, and elevated serum inflammatory markers.

Acute paraplegia in children is an ominous symptom; it may

result from various causes, including traumatic, infectious, inflammatory, demyelinating, or malignant causes. Malignant causes, although they are rare in children, every pediatrician should put a high index of suspicion in dealing with a child presented with lower limb weakness, mainly if associated with signs of the upper motor lesion.

Case Report

A 7-year-old female with previous healthy life was referred as a case of acute paraplegia over a few days duration, followed by a history of urine retention that required catheterization. She denied any history of trauma or fever except for dull backache one month before admission to ER that was managed by oral analgesics. On examination, she was fully conscious with pallor, and her vitals were normal with slightly low blood pressure (90/70 mmHg). Cranial nerves and neurological examination of the upper limbs were normal. In contrast, examination of lower limbs revealed decreased reflexes at early presentation due to spinal shock that changed to increased knee reflexes and bilateral up planter reflexes a few days later. Sensory examination was non-conclusive due to an uncooperative child. The rectal tone was weak. Her initial labs were normal except for mild anemia. The immediate differential diagnoses were Guillain-Barre syndrome, transverse myelitis, and malignant spinal cord compression. MRI of the spine was ordered and revealed evidence of well-defined oval mass at D1 & D2 with the extradural location associated with vertebral bodies wedging and subsequently mass effect on thecal sac and an upper dorsal cord suggesting osteolytic extradural space-occupying lesion (Fig.1).

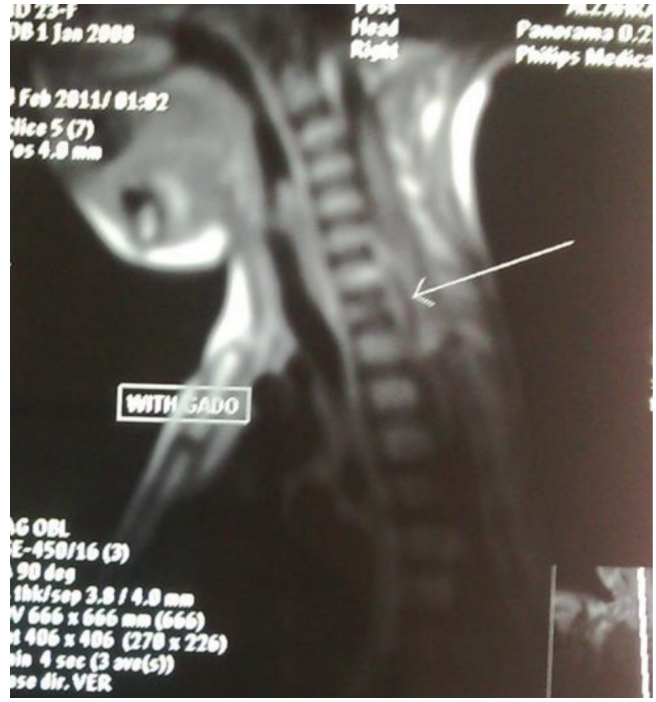

Figure 1. *Spinal MRI shows evidence of well-defined oval mass (Arrow) at D1 & D2 with the extradural location associated with vertebral bodies wedging and subsequent mass effect on the dural sac and upper dorsal cord suggesting osteolytic extradural space-occupying lesion.*

Metastatic workup and staging were started, PET/CT total body was ordered and was negative for neoplastic involvement. Due to the rapid progression of the mass and cord compression effect, she was prepared for emergency spinal decompression, tumor debulking, and dorsal fixation with unilateral pedicle and screws. The tumor was totally resected. The patient had a good postoperative recovery. The neurological assessment showed mild residual deficit with increased muscle strength. The histopathology report for the biopsy was suggestive of a small round blue cell tumor confirmed in an immunohistochemistry study as Ewing's sarcoma. Two weeks after surgery, she received courses in radiotherapy and chemotherapy. Chemotherapy regimens involve cycles of doxorubicin, vincristine, and cyclophosphamide alternating with cycles of ifosfamide and etoposide. During this period, she also had courses in physiotherapy and

rehabilitation. Initially, she was responding well to chemotherapy. Unfortunately, after four months of diagnosis, we lost the patient due to extensive pulmonary and bone metastasis.

Discussion

Primary malignant osseous tumor of the spine is rare in children forming only 1% of all spine, and spinal cord tumors combined 3. Ewing's sarcoma is a poorly differentiated primary bone tumor. It is a part of the Ewing sarcoma family of tumors (EFT). It occurs in the pediatric age group between the ages of 5-15 years3. It most often arises from long bones in the extremities and pelvic bones. Spinal Ewing's sarcoma is extremely rare in the pediatric age group. Sacral bones are the most common location for this lesion, followed by lumbar and dorsal spines, and less often, the cervical spines.

Acute paraplegia is a severe condition in children. Differential diagnoses include many traumatic, inflammatory, infectious, and neoplastic causes. In children, more than 50% of malignant spinal tumors present with neurological symptoms. Malignant spinal tumors like lymphoma or Ewing's sarcoma can be preceded by constitutional symptoms like fever, weight loss, and night sweat which are misleading, especially in young children. Acute neurological symptoms indicate a rapidly growing tumor. Deformities like kyphosis or scoliosis are seen in 25% of patients with malignant spinal tumors, but they are unusual first presenting signs. Management of spinal Ewing's sarcoma needs a multimodality approach (4,5). En-bloc resection with tumor-free margins is a surgical goal to avoid micro-metastasis, especially in rapidly progressing cord compression symptoms. Although it is an aggressive neoplasm, it responds well to radiotherapy. The Current treatment plan involves surgery followed by multidrug chemotherapy and radiotherapy, particularly in a low-grade tumor. A high-grade tumor biopsy followed by chemotherapy and radiotherapy is recommended (3,4).

Conclusion

Although primary malignant spinal tumors are rare in children, it is a fatal condition. A high index of suspicions is needed in evaluating a young child with acute paraplegia, particularly in the era of polio eradication, especially at the early presentation of paraplegia when neurological signs are not clear and non-conclusive. MRI of the spine is the best modality for evaluating a young child with acute paraplegia, especially if cord compression is highly suspected or if the neurological examination findings are deceiving. As a role, early diagnosis with prompt treatment modality will improve outcomes and life expectancy of the patient.

Collected the data and helped in data analysis, wrote the manuscript.

Author's Contribution

Sameer Yahya , Conception and design, Supervised cognitive and behavioral assessments, Hieder A, Al- Shami helped in data analysis, helped in writing We are authors of manuscript (A primary dorsal osseous Ewing's sarcoma hidden as acute paraplegia in a 7-year child) We confirm that we have no funding and competing interest statements.

Acknowledgement

None

Conflict of interest

None

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