World Journal of Advanced Research and Reviews
eISSN: 2581-9615 CODEN (USA): WJARAI
Cross Ref DOI: 10.30574/wjarr
Journal homepage: https://wjarr.com/

(CASE REPORT)

Childhood spinal extradural EWING'S sarcoma: A case report

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World Journal of Advanced Research and Reviews, 2022, 14(03), 228–231

Publication history: Received on 19 April 2022; revised on 31 May 2022; accepted on 02 June 2022

Article DOI: https://doi.org/10.30574/wjarr.2022.14.3.0466

Abstract

Extraosseous, extradural and extramedullary Ewing's sarcoma is a rare tumor that usually affects young people. We report a 10 years old child operated for Inguinal Hernia at the age of 4 years which presents a lumbar spinal syndrome for 4 months, the second is a 3 years old child with a history of falls on stairs 4 months ago, presented for 14 days before his admission a right leg paresis at ⅗ on the muscular testing, associated with fever. Both patients underwent magnetic resonance imaging that showed a compressive extradural process from L2 to L5 for the first case and a compressive extradural process from L4 to S2 for the second case. The patients benefited from macroscopic total resection by laminectomy. Histology confirms the diagnosis of Ewing's Sarcoma. Both patients were referred at discharge to the pediatric oncology department for complementary chemotherapy management. The long-term follow-up at three years was good clinically with no recurrence on imaging. Spinal Ewing's Sarcoma extradural without bone involvement remains rare and its overall survival at 5 years is still poor.

Keywords: Spine; Ewing's Sarcoma; Extradural; Childhood

1. Introduction

Ewing's Sarcoma (ES) is a highly malignant bone tumor (1). It can occur in any part of the skeleton but the most common sites are the ilium and the diaphyses of the femur and tibia (2). These are highly undifferentiated, primitive, malignant, small round cell neoplasms, which usually occur in children and young adults (3).

Ewing's sarcoma is a rare tumor, accounting for 10% of all primary malignant bone tumors in children and 3% of all malignant tumors in childhood (4). Symptoms are dominated by pain which is present in 94-100% and neurological deficit in 50-94% due to spinal cord compression (5-8), however, it is rarely posted traumatically neglected with intradural and extra-spinal imaging features.

2. Case Description

2.1. Clinical Presentation

2.1.1. Case 1

A 10-year-old boy was operated on 4 years ago for an inguinal hernia and presented a progressive lumbar spinal syndrome with back pain for 4 months without sensory-motor deficit. MRI of the spinal cord showed an intradural tissue lesion from L2 to L5 (Figure 1). The blood workup was normal.

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Case 2: A 3-year-old boy with no notable history, presented with progressive monoparesis of the right lower limb following a fall from the stairs in a febrile context (T 39°). Neurological examination at admission found the patient to be conscious GCS 15 with a 3/5 deficit of the right lower limb. A spinal cord MRI showed a tumor process straddling the lumbosacral canal (L4-S2). The blood workup was normal except the chain reaction Protein was increased to 47.5.

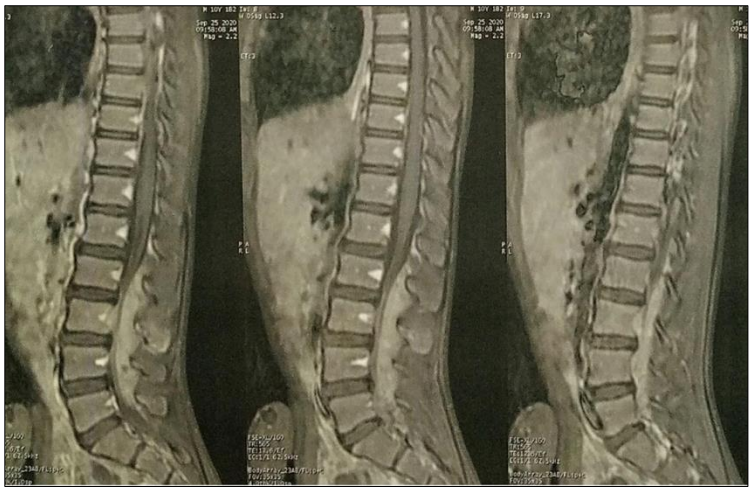

Figure 1 MRI shows the presence of an intracanal lesional process, extended from L2 to L5, this process is extra-dural, encompassing the roots of the cauda equina and filling the right and left foramina in L3-L6 and L4-L5

2.2. Management

The first one benefited from a laminectomy L3-L5 laminectomy with total macroscopic excision of an extradural process whose histology was performed in favor of Sarcoma. The second patient underwent an emergency for L4-S1 laminectomy with total excision of a compressive epiduritis process whose pathology was in favor of Sarcoma.

2.3. Outcomes and Follow-up

The clinical evolution was marked by a progressive neurological recovery, no fever, and normalization of the biological inflammatory parameters. The patients were sent to the pediatric oncology department, they received chemotherapy treatment. After six months of follow-up, the first one recovered his deficit and the second one relieved his pain. The MRI did not show any recurrence at 6 months after surgery and under chemotherapy (Figure 2), and they were both in good clinical and imaging outcomes without recurrence at 3-year follow-up.

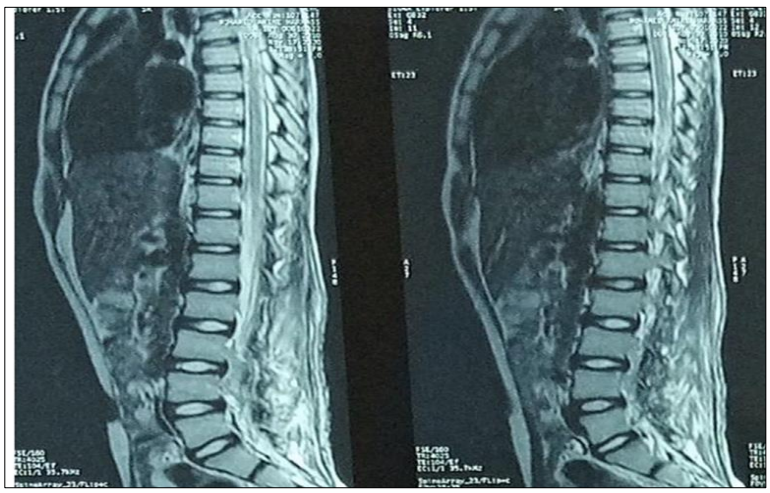

Figure 2 Postoperative MRI showing total regression of the lesional intracanal epidural process L2-L5, the respect to spinal statistics, and the persistence of significant infiltration of the lumbar paravertebral soft tissue related to postoperative changes

3. Discussion

We reported two rare cases of spinal Ewing's Sarcoma in childhood which we managed surgically with good clinical and imaging outcomes under chemotherapy.

ES occurs in the second decade of life under the age of 30 years, its occurrence after the age of 30 is rare (4, 5). They were 03 years and 10 years old boys in our cases.

The clinical presentation includes radicular syndrome with progressive sensory and motor function disturbances followed by the inflammatory syndrome. Besides, ES affected the lung bones and rarely the spinal (5%) (2, 3).

The mainstay of treatment is complete resection of the tumor followed by chemotherapy or radiotherapy, If metastasis was not present at the time of diagnosis combined treatment with radiotherapy and chemotherapy after surgery are recommended for good outcomes with fewer recurrences (2-5).

The authors suggested that the first-line treatment of ES of the spine should be induction chemotherapy before surgery even in a case of major neurological deficit, even in case of major neurological deficits (9). In their series, Event Free Survival (EFS) was lower in patients who were submitted to primary surgery than their patients who had initial chemotherapy (P = 0.011), Also the 5-year OS and EFS in their series were respectively 44.0% and 37.0%. However, the prognosis of ES is still poor (3, 6).

In our present case, the tumor was located in the extra bony, extradural, part of the cauda equina surrounding neural plates, they were treated with surgery and chemotherapy. They had no recurrence after 3 years of follow-up.

4. Conclusion

Ewing's sarcoma is a malignant bone tumor, with extraosseous, spinal extradural locations being relatively rare, the diagnosis is based on histology. The 05 years OS is significantly improved by chemotherapy. The management is still multidisciplinary between neurosurgeons and medical oncologists.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest.

Funding

The authors did not receive any funding for the preparation of this case report.

Statement of informed consent

Informed consent and verbal permission were obtained from the patient and her family before the submission of this article. In addition, this article follows both the Consensus-based Clinical Case Reporting Guideline and the Recommendations for the Conducting, Reporting, Editing, and Publication of Scholarly Work in Medical Journals.

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