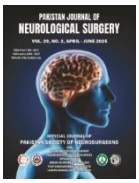

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Original Research

A Rare Presentation of Extradural Ewing Sarcoma in the Lumbar Spine: A Case Report

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ABSTRACT

Introduction: Ewing sarcoma (ES) is the most prevalent malignant bone tumor in children and adolescents, mainly impacting the axial skeleton and long bones. Extradural manifestations of Ewing sarcoma, particularly in the lumbar spine, are rare and may pose diagnostic challenges. This manuscript describes an unusual case of an epidural lumbar region, Ewing sarcoma, presented with acute neurological symptoms.

Case Presentation: A 14-year-old male patient presented with a history of a fall resulting in paraparesis and urinary incontinence for the last 1 week. MRI revealed an extradural lesion compressing thecal sac at the L4-L5 level. An extradural hematoma or an ependymoma were the initial differentials, and intravenous steroids were commenced, resulting in partial relief of symptoms. Later on, a core needle biopsy and immunohistochemical staining were performed, confirming Ewing sarcoma with positive CD99 and vimentin. Surgical excision of the tumor was performed, achieving clear margins, and there was significant improvement in patient symptoms.

Discussion: Lumbar epidural Ewing sarcoma in the lumbar spine is unusual and can result in acute neurological symptoms, creating a diagnostic dilemma. This case report testifies to the significance of maintaining high suspicion and emergency action for extradural spinal tumors. A complete surgical excision confirmed the diagnosis and improved the patient's neurology.

Conclusion: Recognising a spinal tumor and swift treatment is vital to patient improvement, especially in acute cases. A delay can result in permanent neurological harm to the patient.

Keywords: Ewing sarcoma, extradural lesion, surgical excision, extradural spinal tumors.

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INTRODUCTION

Ewing's sarcoma was first described by James Ewing in 1921. It is the most common malignant bone tumor in the initial decade of a patient's life.[1] During the twenties, it ranks second to

osteosarcoma.[2] Ewing sarcoma is now considered part of the family of Ewing sarcomas, which encompasses the Askin tumor, primitive neuroectodermal tumor, extraosseous Ewing sarcoma, and Ewing sarcoma.[3] Tefft et al, originally described extra-skeletal Ewing sarcoma in 1969.[4] The paravertebral muscles, chest wall, retroperitoneal region, extremities, and pelvis represent several regions where it may be observed.[5]

There are only a handful of documented cases of primary extradural Ewing sarcoma located in the lumbar spine that causes compression of the neural elements. A young patient could present with quickly exacerbating paraplegia along with bowel and bladder incontinence, creating a challenging diagnostic scenario. Diagnosing lumbar epidural Ewing's sarcoma requires a strong suspicion. This case study illustrates a relatively rare manifestation of the disease and the patient's recovery after receiving appropriate surgical intervention.

Case Presentation

A 14-year-old male patient arrived with a one-week history of weakness in both lower limbs and urinary incontinence following a 10-foot height. On examination, he had 1/5 power in both dorsiflexion and plantarflexion of the ankle. The sensations were also reduced in L4, L5, and S1 dermatomes. There was also saddle hypoesthesia. Magnetic resonance imaging (MRI) was performed, keeping in view the trauma history and positive neurology. The imaging results indicated an extradural lesion at the L4-L5 vertebral level, exhibiting hypo-isointense characteristics on T1-weighted images and hyperintensity on T2-weighted images. This lesion extends into the spinal canal and exerts pressure on the thecal sac, as demonstrated in the accompanying MRI images (Figure 1). Furthermore, the computed tomography (CT) scan of the chest, abdomen, and pelvis yielded unremarkable findings.

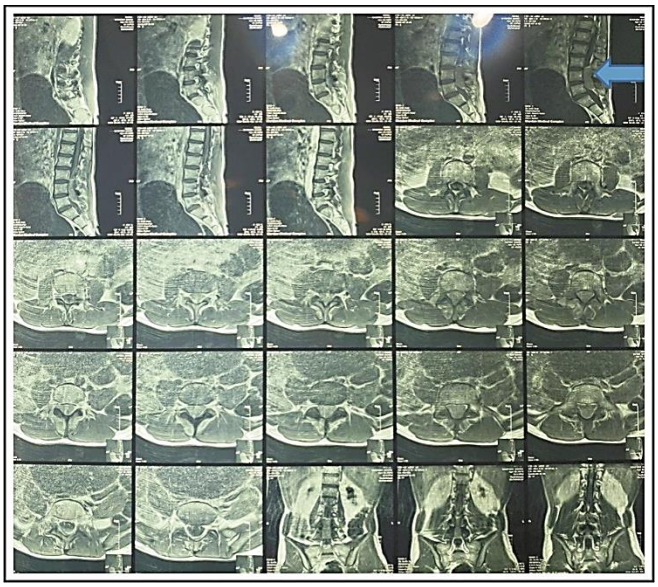

Figure 1: *MRI of the Patient (after informed consent) showing the spinal tumor.*

It was suspected to be either an extradural hematoma or an ependymoma. The patient was initially treated with intravenous steroids (dexamethasone 4mg i/v TDS for 5 days), which led to some improvement in symptoms. Nevertheless, a core needle biopsy indicated the presence of a minor, blue-cell-rounded neoplasm, consistent with a primitive neuroectodermal tumor. The tumor exhibited positive staining for CD99 and vimentin.

The patient underwent surgical resection with an adequate margin, decompression, and L4-5 laminectomy. The lesion was completely removed with clear margins, as confirmed by the surgical pathology report. The operative picture of the lesion is shown below after permission from the patient (Figure 2).

The histopathology report came out to be extradural Lumbar Ewing Sarcoma.

Due to the high risk of metastasis, the patient received adjuvant chemotherapy and radiation therapy prescribed by the oncologist as per the standard guidelines. The patient's motor strength gradually recovered to 4/5 in the affected

myotomes, and there was also sensory recovery after a 2-month follow-up; the spinal biomechanics remained stable. There has been no tumor recurrence in the last five months of follow-up.

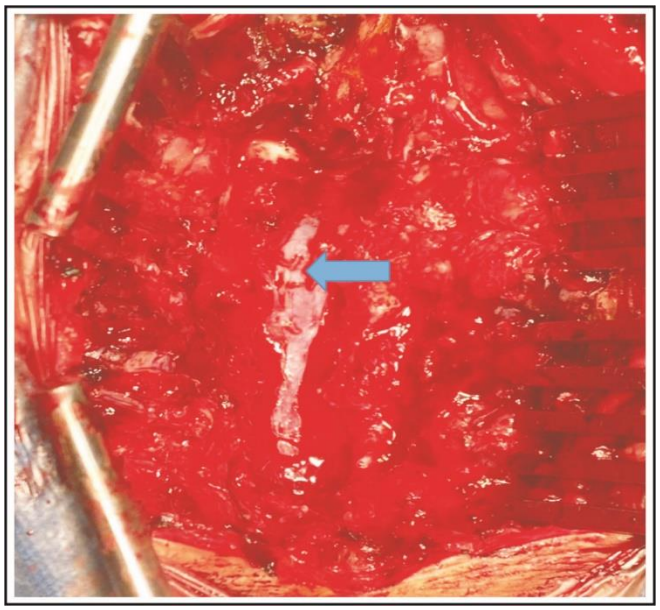

Figure 2: *Intra-operative image (with the informed consent of the patient) of the spinal tumor (Ewing Sarcoma).*

DISCUSSION

Ewing sarcoma (ES) constitutes a rare and aggressive malignant bone tumor that predominantly impacts children and young adults. This particular case is noteworthy due to its involvement of an extradural manifestation of Ewing sarcoma in the lumbar spine. The lumbar spine is an unusual location for Ewing sarcoma. Nevertheless, it is an important differential diagnosis in a patient presenting with acute onset of neurological deterioration and thecal sac compression.[6]

Diagnosing Ewing sarcoma in the lumbar spine is challenging owing to its rarity in this location and the vagueness of the neurological symptoms. Lower limb weakness, paraplegia, and bowel or bladder incontinence can present with a variety of lumbar spine pathologies. In this young patient, the initial differentials suggested by the history (of falls) and MRI was epidural hematoma or ependymoma (the most common tumor at this location). Ewing sarcoma was diagnosed only after a core needle biopsy followed by immunohistochemical analysis was performed. The positive staining for CD99 and vimentin was vital to making the diagnosis as these are typical markers for Ewing sarcoma and its wider family.[7,8]

Due to the acute onset and rapid progression of neurological deficit, complete surgical excision of the tumor was planned to achieve decompression of the cal sac. Enbloc resection was confirmed intraoperatively and through subsequent biopsy reports. The surgical excision not only reaffirmed the diagnosis of Ewing sarcoma but also resulted in immediate post-operative improvement in the patient's neurology. Hence, swift and accurate decision-making in cases of rapid onset of neurological deficit and MRI-confirmed compressive pathology is vital to the best possible outcome for these patients.

The management of this patient underscores the significance of exhibiting a high index of suspicion and swift action in suspected epidural spinal lesions. Delaying the diagnosis and subsequent treatment may result in permanent neurological compromise to the patient.[9,10] It is also crucial to seek thorough histopathological analysis and immunohistochemistry to diagnose rare pathologies.

A literature search reveals only a few cases of Ewing sarcoma reported in the lumbar spinal region, as it usually occurs in the chest wall, femur, and pelvic bones.[11,12] The presentation and management of the patient reported in this manuscript contribute to properly managing tumors occurring at unusual locations. It also provides insight to further research into this rare tumor of the lumbar spinal region and devising further management strategies for a better prognosis of the tumor.

CONCLUSION

Ewing sarcoma of the lumbar spine is rare but a critical differential, especially in the pediatric age group, particularly in young individuals presenting with cauda equina syndrome or acute neurological deficit of the lower limbs. A multidisciplinary strategy involving the radiologist, pathologist, neurosurgeon, and oncologist, along with a physiotherapist, is crucial to diagnose and manage the tumor promptly. Oncological input is also paramount in deciding whether chemotherapy or radiotherapy can achieve a complete cure.

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Additional Information

Disclosures: Authors report no conflict of interest.
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Human Subjects: Consent was obtained by all patients/participants in this study.
Conflicts of Interest: In compliance with the ICMJE uniform disclosure form, all authors declare the following:
Financial Relationships: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work.
Other Relationships: All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

AUTHORS CONTRIBUTIONS

Sr.#	Author's Full Name	Intellectual Contribution to Paper in Terms of:
1.	Syed Nasir Shah	1. Study design and methodology.
2.	Musawer Khan	2. Paper writing.
3.	Waseem Sajjad	3. Data collection and calculations.
4.	Naeem-ul-Haq	4. Analysis of data and interpretation of results.
5.	Shehzad Sadbar, Zeeshan Ali	5. Literature review and referencing.