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Case Report

Ewing Sarcoma of Neck - A Rare Case Report

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ABSTRACT

This case report details a rare presentation of cervical Ewing's sarcoma in a 4-year-old female, emphasizing its clinical presentation, diagnostic process, and treatment challenges. The patient initially presented with a rapidly growing neck mass and neurological symptoms; MRI confirmed a well-defined mass at the C5-C6 vertebral level, and histopathological examination verified the diagnosis of Ewing's sarcoma. Treatment involved neoadjuvant chemotherapy, followed by surgical resection with clear margins, and the patient is currently being monitored for recurrence with a guarded prognosis. This case underscores the need for a multidisciplinary approach, early intervention, and thorough surgical planning in managing rare and aggressive tumors in complex anatomical locations, contributing valuable insights to the limited literature on this uncommon malignancy.

Keywords: Ewing sarcoma, Cervical region, Neck tumor, Pediatric oncology

INTRODUCTION

Ewing's sarcoma is a rare and aggressive malignancy predominantly affecting children and young adults. It typically arises in the bones or soft tissues, with the most common sites being the long bones of the limbs, pelvis, and chest wall. The occurrence of Ewing's sarcoma in the cervical region is exceptionally uncommon, making such cases noteworthy for their rarity and the unique challenges they present.[1]

First described by James Ewing in 1921, Ewing's sarcoma is characterized by small, round, blue cells on histopathology and often involves a translocation between the EWSR1 gene and an ETS family transcription factor, most commonly FLI1.[2] This translocation results in the production of an abnormal protein that promotes tumorigenesis.

In the cervical region, Ewing's sarcoma can manifest as a neck mass, sometimes accompanied by pain or neurological symptoms due to the proximity to vital structures. Imaging studies, such as MRI and CT scans, are crucial for evaluating the extent of the tumor and planning treatment. Diagnosis is typically confirmed through a combination of histopathological examination and molecular studies to identify characteristic genetic translocations.

Treatment for Ewing's sarcoma generally involves a multimodal approach, including chemotherapy, surgery, and sometimes radiotherapy. Chemotherapy is often administered first to shrink the tumor, making surgical resection more feasible. In cases where the tumor is located in the neck, surgical intervention can be particularly complex due to the intricate anatomy and the need to preserve essential functions. Adjuvant therapies, such as radiotherapy, may be employed depending on the tumor's location and the presence of metastasis.[3]

Prognosis for Ewing's sarcoma varies based on factors such as the patient's age, tumor size, location, and the extent of disease at diagnosis. Advancements in treatment have improved survival rates over the years. However, due to the rarity of cervical Ewing's sarcoma, specific prognostic data for this location are limited. Close monitoring for recurrence and long-term follow-up is essential components of post-treatment care.[4]

This case report aims to contribute to the limited literature on cervical Ewing's sarcoma by detailing the presentation,

diagnostic process, treatment, and outcome of a 15-year-old male patient. By sharing this experience, we hope to enhance awareness and provide insights that may assist in the management of similar cases in the future.

CASE HISTORY

We report the case of a 4-year-old female, who presented with a rapidly growing neck mass, progressive weakness, and neck pain. Initial diagnostic imaging with MRI revealed a well-defined mass located between the L5-L6 vertebral level. The lesion extended to involve the adjacent spinal structures. The mass was suspicious for malignancy based on the imaging characteristics and patient presentation.

Histopathological examination confirmed the diagnosis of Ewing's sarcoma. The tumor was composed of small round cells with occasional rosette formation. Immunohistochemical staining was positive for Mic-2, NKX 2.2, and synaptophysin, confirming the diagnosis of Ewing's sarcoma. Further investigations ruled out distant metastasis.

Histopathological Findings: The biopsy showed malignant round cells with nuclear hyperchromasia, scant cytoplasm, and occasional mitotic figures. The tumor cells were arranged in sheets with necrosis present in certain regions. Immunohistochemical staining revealed positive markers for Ewing's sarcoma, including Mic-2, NKX2.2, synaptophysin, and desmin. The tumor was diagnosed as a round cell tumor, which was further classified as Ewing's sarcoma.

Imaging Findings: The MRI of the neck and cervical spine showed an extradural mass located between the L5-L6 vertebral level, with extension into the surrounding soft tissues. The sagittal and axial views revealed the mass causing mild compression of adjacent structures but without significant spinal cord invasion. The tumor demonstrated hypo intensity on T1-weighted images and hyperintensity on T2-weighted images, consistent with the appearance of Ewing's sarcoma [Figure-1].

Treatment and Outcome: The patient underwent a multimodal treatment approach, including neoadjuvant chemotherapy to shrink the tumor, followed by surgical resection. Given the location of the tumor in the neck, careful planning was essential to avoid damage to critical neurovascular structures. Post-operative radiotherapy was not indicated due to clear margins. The patient is currently undergoing regular follow-up to monitor for signs of recurrence, and her prognosis remains guarded due to the aggressive nature of the tumor.

DISCUSSION

Ewing's sarcoma, though primarily affecting the long bones and pelvis, occasionally presents in atypical locations such as the cervical region, as seen in this case. Cervical involvement is rare, constituting a unique challenge in terms of diagnosis, treatment planning, and prognosis. The atypical location requires a high index of suspicion, especially in pediatric patients presenting with neck masses. In our case, the patient, a 4-year-old female, demonstrated typical symptoms including a rapidly growing mass and pain, but the cervical location added complexity to the diagnostic process.[5]

Histopathological examination plays a crucial role in confirming the diagnosis of Ewing's sarcoma, which is characterized by small round blue cells and a specific immunohistochemical profile. In this case, the presence of markers such as Mic-2 and synaptophysin, along with genetic testing confirming the EWSR1 translocation, solidified the diagnosis. This molecular confirmation is consistent with other cases reported in literature, where genetic mutations are integral in establishing an accurate diagnosis.[6]

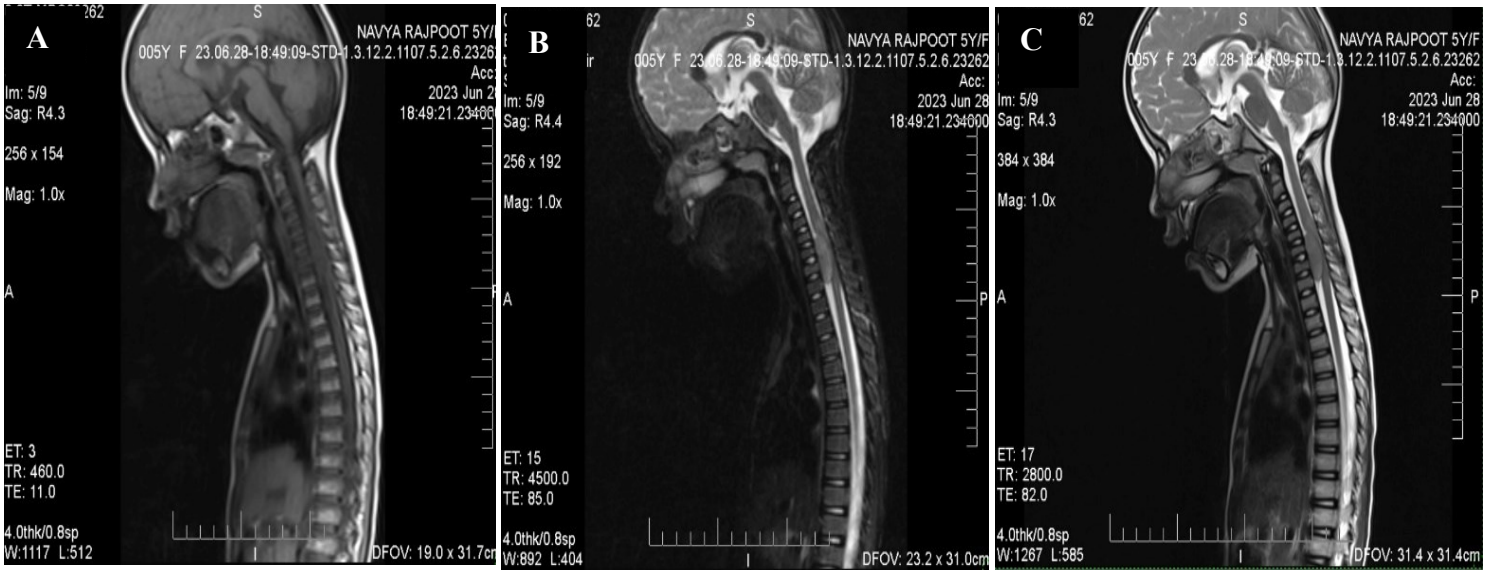
The treatment of cervical Ewing's sarcoma, like other locations, involves a multimodal approach. Neoadjuvant chemotherapy is typically the first step to reduce tumor size, followed by surgical resection. In our case, the tumor's proximity to critical neurovascular structures in the neck required meticulous surgical planning. Literature suggests that the complexity of surgery in cervical Ewing’s sarcoma often limits complete resection due to the vital anatomical structures involved. However, the surgical team achieved clear margins in this case, obviating the need for adjuvant radiotherapy, which is typically recommended in cases with incomplete resection or positive margins.[7]

Prognosis in Ewing’s sarcoma depends heavily on factors such as the size of the tumor, response to chemotherapy, and the presence or absence of metastasis. Cervical Ewing’s sarcoma, due to its rarity, has less established prognostic data compared to more common locations. However, the aggressive nature of Ewing’s sarcoma necessitates close follow-up to monitor for recurrence. In this patient, while initial treatment outcomes have been favorable, the guarded prognosis is reflective of the high recurrence rates associated with Ewing’s sarcoma in rare anatomical locations.[8]

In comparison to similar cases, the early detection and comprehensive treatment in this case reflect the critical importance of a multidisciplinary approach in managing complex cases of Ewing’s sarcoma. Future reports and studies on cervical Ewing’s sarcoma are essential to better understand the long-term outcomes and optimize treatment protocols for such rare presentations.[9-10]

This discussion highlights the rarity of cervical Ewing's sarcoma and underscores the challenges in management, while emphasizing the importance of a coordinated approach for successful outcomes.

Figure-1: MRI - a well-defined IDEM mass lesion in cervical region (opposite to C5-T1 vertebral bodies) which is hypointense on T1 (Figure A) , isointense T2 (Figure B), iso to hyper intense on T2 STIR (Figure C), showing homogenous enhancement on post contrast (Gd) and causing severe spinal cord compression



CONCLUSION

Ewing's sarcoma of the neck is an exceptionally rare and aggressive malignancy, posing significant diagnostic and therapeutic challenges due to its atypical location and proximity to vital structures. Early diagnosis, multimodal treatment involving chemotherapy and surgery, and careful surgical planning are critical for achieving optimal outcomes. In this case, the successful management of the tumor with clear surgical margins highlights the importance of a multidisciplinary approach. However, close follow-up is essential due to the high risk of recurrence associated with this aggressive tumor.

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