Primary Cervical Extraosseous Ewing's Sarcoma in a Pediatric Patient: A Case Report

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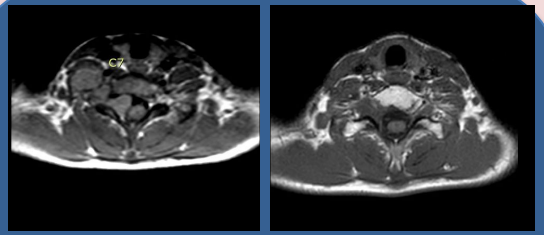
Patient History:

A 13-year-old female presented with a nine month insidious onset of diffuse, intermittent weakness and numbness of her right upper extremity (RUE).

Initial physical exam findings were significant for diffuse cervical paraspinal muscle tenderness with no associated midline tenderness and diminished sensation in her RUE. Initial X-ray imaging showed no acute osseous abnormalities. However, MRI imaging showed a localized extra-osseous mass of the lower cervical spine located in the right epidural space and extending into the C6-C7 and C7-T1 intervertebral foramen. Histopathological analysis showed evidence of Ewing's sarcoma.

The patient subsequently underwent surgical exploration which showed tumor involvement of the right brachial plexus that proved to be not fully resectable. Patient was treated with a combination of chemotherapy per the COG Protocol AEWS1031 and a total of 25 fractions of intensity-modulated radiation therapy. Patient completed treatment in March 2019 and has since been in clinical remission with no complications.



Left: Soft tissue lesion centered in the right epidural space extending through the intervertebral foramina at C6-7 and C7-T1 is noted.
Right: Foramen on the right at C6-7 and C7-T1 demonstrate signal asymmetry from the left side with no associated pathologic enhancement. No evidence of disease recurrence.

Discussion:

Primary extraosseous Ewing's sarcoma of the cervical spine is a rare diagnosis with few reported cases in the last decade. In this case, X-ray imaging revealed no acute osseous abnormalities, making MRI necessary for the patient's diagnosis. This highlights the importance of acquiring additional imaging and maintaining a high index of suspicion. Given this infrequent presentation and poor tumor prognosis, it is important to note the role of combined chemotherapy, radiotherapy, and surgery in the management of this patient's Ewing's sarcoma.

Conclusion:

Primary extraosseous Ewing's sarcoma of the cervical spine is a rare diagnosis with few reported cases in the last decade. In this case, X-ray imaging revealed no acute osseous abnormalities, making MRI necessary for the patient's diagnosis. This highlights the importance of acquiring additional imaging and maintaining a high index of suspicion. Given this infrequent presentation and poor tumor prognosis, it is important to note the role of combined chemotherapy, radiotherapy, and surgery in the management of this patient's Ewing's sarcoma. Conclusion: Although the incidence of primary cervical extraosseous Ewing's sarcoma is low, it should remain a possible diagnosis in adolescents with similar presentations. Imaging is a crucial factor in making the diagnosis given that there are no characteristic signs or symptoms. Due to the aggressive nature of these tumors, a multidisciplinary approach involving chemotherapy, radiotherapy, and surgery is often necessary.