

## CASE REPORT

# Primary Ewing's sarcoma of cervical vertebra: An uncommon presentation

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## ABSTRACT

Ewing's sarcoma is a malignant primary bone tumor primarily seen in the long bones. Primary Ewing's sarcoma of the cranium is quite uncommon occurring in 1% of the cases. We report the occurrence of this rare lesion in a 24-year-old male presenting with progressively increasing swelling in left mastoid region mimicking a mastoid abscess which was later diagnosed on Fine needle aspiration cytology (FNAC) as a small round cell tumor as Ewing's sarcoma. Contrast enhanced computed tomography (CECT) revealed a typical moth eaten appearance in the first and second cervical vertebra.

**Key words:** Ewing's sarcoma, primary cervical vertebra tumor, tumor of atlas vertebra

## Introduction

Primary tumors of the spine are relatively infrequent lesions compared with metastatic disease, multiple myeloma, and lymphoma. Primary Ewing's sarcoma of spine is quite uncommon and its location in the cervical spine has been seen very rarely.<sup>[1]</sup>

## Case Report

We report the case of a 24-year-old male presenting with a progressively increasing painful swelling in left temporo-occipital region over a period of one year. Physical examination revealed a firm, fixed, tender swelling measuring 9 × 8 cm over the left mastoid [Figure 1]. Neurological examination showed sensory and motor weakness in left upper limb.

On ultrasonography, a heteroechoic mass lesion of size 8 × 7 cm was seen in subcutaneous and muscle plane with deeper extension in relation to the cervical vertebra. The lesion showed an increased flow on color doppler. Contrast enhanced computed tomography (CECT) revealed destruction of C1, C2 vertebra (moth eaten appearance) with

associated enhancing soft tissue component extending into the extradural, paravertebral space, and foramen magnum, pushing the medulla to the right, and the internal carotid artery and internal jugular vein antero-laterally [Figure 1].

Fine needle aspiration of the swelling was done which revealed cellular smears showing small, round cells in loose cohesive clusters, rosettes and dispersed singly with round to irregular bland nuclei, abundant vacuolated cytoplasm positive for cytoplasmic glycogen [Figure 2]. The cells were interspersed with small dark cells with scanty cytoplasm. The cytological diagnosis of small round cell tumor, Ewing's sarcoma was made. A biopsy was performed which revealed sheets of round cells displaying perivascular arrangement at places in a necrotic background. Immunohistochemical evaluation showed diffuse cytoplasmic positivity for CD99, neuron specific enolase (NSE), and vimentin, whereas no expression was seen with LCA (CD45), synaptophysin, chromogranin, and desmin. Based on the imaging findings, primitive small round cell histology and characteristic positivity for CD99, NSE, and vimentin, the diagnosis of Ewing's sarcoma was established.

## Discussion

In 1921, James Ewing described a lethal primary bone tumor that affects children and young adults and most frequently originates in the long bone (47%), pelvis (19%), or ribs (12%).<sup>[2]</sup>

Ewing's sarcoma is the second most common bone tumor primarily seen in the long bones. This neoplasm is most frequently seen in children, with 75 % arising in patients under 20 years. The male-to female ratio is 1.6:1.<sup>[3]</sup>

Primary nonlymphoproliferative tumors of the spine represent less than 5% of all bone neoplasms. Ewing's and PNET are the most common nonlymphoproliferative primary malignant

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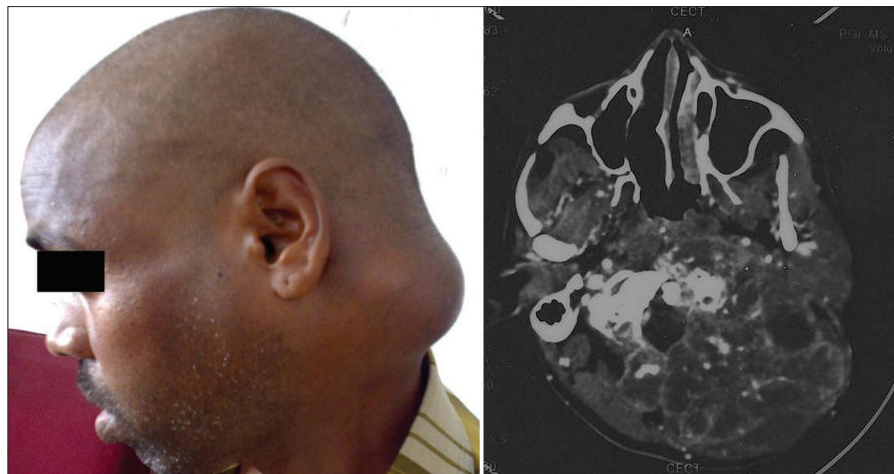
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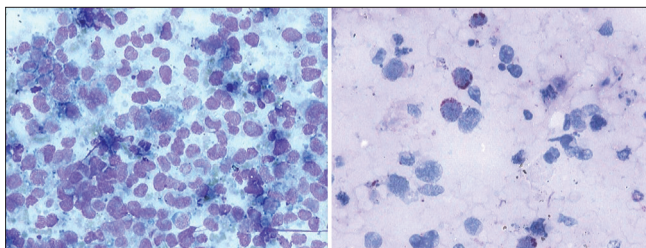
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**Figure 1:** (L)-Firm, fixed, tender swelling measuring 9x8 cm over the left mastoid, (R)-(CECT) revealing destruction of C1, C2 vertebra (moth eaten appearance)



**Figure 2:** (L)-FNAC smear showing small, round cells in loose cohesive clusters, rosettes and dispersed singly (MGG, x200), (R)-FNAC smear showing small, round cells in loose cohesive clusters and dispersed singly with positivity for cytoplasmic glycogen (PAS, x400)

tumors of the spine in children. Lesions of the spine account for 3%-10% of all primary sites of Ewing's sarcoma and PNET. However, metastatic foci of Ewing sarcoma involving the spine are much more common than primary lesions. Patients with both lesions usually present between the ages of 10 and 30 years of age. Clinical symptoms are pain and neurological changes.<sup>[4]</sup> The most common location of Ewing sarcoma in the spine is sacrococcygeal region, followed by the lumbar and thoracic segments, and only rarely the cervical spine. The lesions are centered in the vertebral body, although extension into the posterior elements is not uncommon.<sup>[5]</sup> On roentgenograms, Ewing's tumors tend to be extensive, sometimes involving an entire bone. The lesion is usually a permeative one with destructive process, with multiple tiny areas of destruction involving the bone. Typically, Ewing's tumor produces a pronounced reactive new bone formation of the periosteum, giving rise to an onion-skin appearance.<sup>[4]</sup>

The differential diagnoses considered were malignant lymphoma, metastatic embryonal rhabdomyosarcoma, and metastatic neuroblastoma. Malignant lymphomas usually affect older patients and, on immunohistochemistry, most lymphomas are positive for leukocyte common antigen. Likewise, with metastatic embryonal rhabdomyosarcoma, specific immunohistochemical studies reveal positive

expression by actin, desmin, and myoglobin which are negative in the Ewing's sarcoma. To rule out metastatic neuroblastoma was not a problem in the absence of primary elsewhere. Moreover, with a clearcut rosette formation and a neurofibrillary background, the diagnosis of neuroblastoma is obvious and those which do not show such differentiation can be diagnosed by immunohistochemistry.

Before the advent of chemotherapy, survival of patients with spinal Ewing sarcoma was dismal because these axial lesions could not be completely resected. However, radiation therapy and chemotherapy are the current mainstays of treatment for spinal lesions, with results approaching 100% for local control and 86% for long term survival for patients with nonsacral tumors. Patients with evidence of instability or neurologic compromise may still require surgical decompression and stabilization.<sup>[1]</sup> However, Ewing's sarcoma originating from the axial system is qualified as with high risk and has a worse prognosis compared with those at the extremities because of the high tumor volume, frequent locoregional recurrence and distant metastasis, and difficulties in radical surgical intervention.<sup>[6]</sup>

In the present case, the patient was treated with partial resection of the tumor mass followed by chemotherapy cycles and local radiotherapy avoiding exposure to the spinal cord, to which he responded well. He was advised to follow-up for five years for the risk of recurrence, and is tumor free for 18 months. Left upper limb power is 4/5.

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