



## Case Report

# Primary Ewing's sarcoma of the C2 vertebra with progressive quadriparesis: Report of a rare case and review of the literature

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## ABSTRACT

**Background:** Ewing's sarcoma is a malignant primitive neuroectodermal tumor (PNET) of childhood and adolescence. Primary Ewing's sarcoma of the spine is uncommon, and even more rarely involves the C2 vertebra.

**Case Description:** A 14-year-old patient was admitted with a history of chronic neck pain, which exacerbated after playing contact sports 3 weeks before presentation. On initial examination, he had pain radiating into the left upper extremity plus spasticity in all the four limbs. The cervical X-rays revealed a mixed sclerotic-lytic lesion involving the C2 vertebral body. The CT bony and soft-tissue windows documented predominant left-sided tumor invasion of the posterior elements, pedicles, and body of C2 along with extension into the spinal canal resulting in severe cord compression with peritumoral soft-tissue edema. The angiogram revealed a patent left vertebral artery entirely surrounded/encased by tumor. The PET-CT scan demonstrated no other spinal or systemic lesions. Due to his rapid neurological deterioration, the patient underwent an emergent biopsy of the tumor with posterior decompression and occipitocervical stabilization. The biopsy demonstrated a PNET (e.g., positive CD 99 MIC2 marker for Ewing's sarcoma). Following subsequent chemotherapy and radiation, the patient rapidly improved over a period of 3 months.

**Conclusion:** Primary Ewing's sarcoma involving the C2 vertebra is exceedingly rare and warrants surgical decompression with pathological confirmation to provide additional multi-modal/multi-disciplinary adjunctive radiation/chemotherapy.

**Keywords:** C2 vertebra, Cervical spine, Ewing's sarcoma, Primitive neuroectodermal tumor

## INTRODUCTION

Ewing's sarcoma is a malignant, primitive neuroectodermal tumor that occurs in the pediatric or young adult population. It involves more males than females, and has a peak age incidence of 15.<sup>[9]</sup> Primary Ewing's sarcoma involving the spine is uncommon, and only rarely involves the cervical spine (e.g., in this case C2).<sup>[10]</sup>

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## CASE DESCRIPTION

A 14-year-old male, who sustained a sports injury 3 weeks ago, acutely presented with progressive neck pain and left upper limb radiculopathy and quadriparesis. On examination, he had tenderness over the upper cervical spine, and exhibited a Nurick Grade III myelopathy characterized by weakness (Medical Research Council Grade 4/5) in all four extremities with spasticity in all four extremities.

### Radiographic studies

Radiographs of the cervical spine showed a predominantly sclerotic lesion with multiple lytic foci involving the C2 vertebra [Figure 1].

The MR revealed an expansile lesion involving the C2 vertebral body; the tumor infiltrated the laminae, pedicles, transverse processes, and spinous process [Figure 2]. There was also an enhancing intraspinal component resulting in cervical cord compression. The CTA (e.g., CT-angiography) revealed tumor encasing the left vertebral artery, without



Figure 1: Radiograph anteroposterior and lateral view showing sclerotic and lytic lesion involving C2 vertebra.

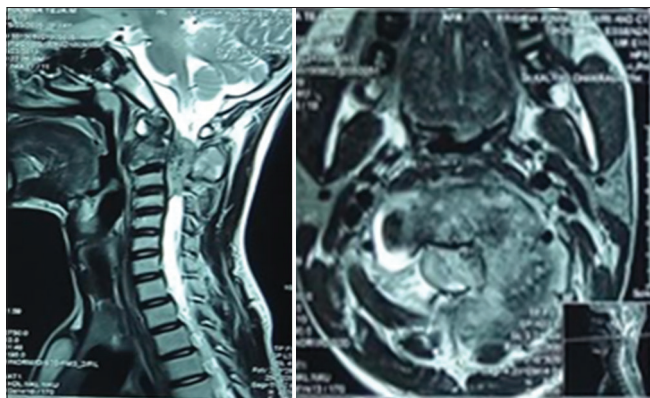


Figure 2: T2 weighted MRI showing an expansile lesion involving C2 vertebra with cord compression.

significantly narrowing the lumen. The PET-CT scan demonstrated no metabolically active additional spinal or systemic lesions.

### Clinical and surgical course

Over 2 days, the patient neurologically deteriorated, and lost sphincter functions. He, therefore, underwent a posterior spinal decompression (e.g., laminectomy C2/C3) with biopsy of the lesion and occipitocervical stabilization (C2-C4). Intraoperatively, the lesion appeared highly vascular with grayish white cheesy material destroying the neural arch, especially on the left side.

Surgical excision required; partial removal of the C2 vertebra (e.g., full resection was not deemed feasible because of encasement of the left vertebral artery), posterior excision of tumor involving the C2 lamina, and C3 superior dome laminectomy. Stabilization required placement of an occipital plate with pedicle screws placed at C3, and lateral mass screws at C4.

### Pathology

The frozen section diagnosis favored a round cell tumor. The final histopathology showed a densely cellular tumor consisting of small round cells with clear cytoplasm [Figure 3a]. The immunohistochemistry showed that the tumor was strong positivity for vimentin-clone V9 and CD99 MIC2- clone 12E7 [Figure 3b]; these findings were consistent with the diagnosis of Ewing's sarcoma.

### Postoperative course

Postoperatively, the patient underwent physiotherapy, rehabilitation, and chemotherapy (e.g., received vincristine, adriamycin, cyclophosphamide, ifosfamide, and etoposide according to National Comprehensive Cancer Network guidelines).<sup>[7]</sup> In addition, a total dose of 50 Gy radiation was given using the 3DCRT protocol. One year later, he fully recovered neurological function, and radiographs confirmed stability of the surgical construct [Figure 4]. Further, the PET-CT scan showed no evidence of any other lesions.

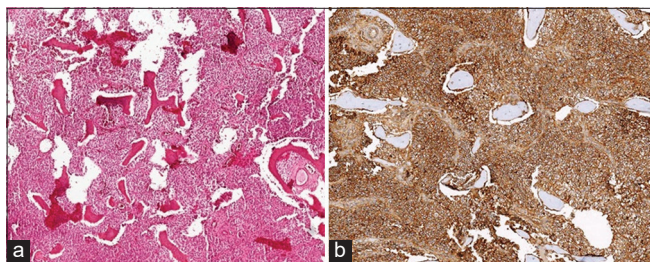
## DISCUSSION

### Frequency of spinal Ewing's sarcomas

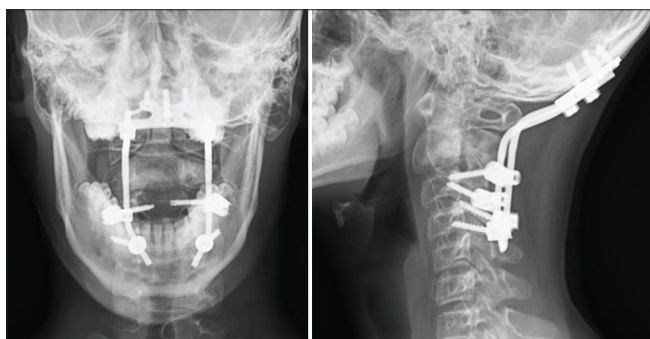
Ewing's sarcoma involving the cervical spine, particularly involving the atlantoaxial junction, is rare, and constitutes approximately 3.2% of all spinal Ewing's sarcomas.<sup>[5]</sup>

### Differential diagnoses

The differential diagnoses of cervical Ewing's sarcomas include giant cell tumor, aneurysmal bone cyst, Langerhans cell



**Figure 3:** (a) Histopathology shows densely cellular tumor consisting of small round cells with clear cytoplasm. (b) Immunohistochemistry shows positivity for vimentin-clone V9 and CD99 MIC2-clone 12E7.



**Figure 4:** One year post operative radiograph shows well maintained sagittal alignment with implant *in situ*.

histiocytosis, vertebral hemangioma, chordoma, malignant lymphoma, metastatic embryonal rhabdomyosarcoma, neuroblastoma, and bacterial infection.<sup>[2]</sup>

### Treatment options

The treatment of cervical Ewing's sarcomas must be individualized based on the age, location, and size of the tumor, neurological deficits, reconstruction options, and experience of the surgeon. Postoperative chemotherapy is paramount in eliminating the residual tumor, and reducing the likelihood of metastasis. Giakoumettis *et al.*<sup>[6]</sup> reported a case of Ewing's sarcoma involving C2 vertebra in a 3.5-year-old girl without neurological deficits; she was successfully treated with chemotherapy alone. Aggarwal *et al.*,<sup>[1]</sup> in an 11-year-old female who presented with myelopathy and a lesion involving C2, performed an anterior decompression, followed by an occipitocervical fusion for neoplastic mass; the patient was initially treated with empirical anti-tuberculous therapy that was appropriately changed to chemotherapy once the biopsy revealed Ewing's sarcoma.

### Surgical options

En-bloc excision at the occipitocervical junction presents a tremendous technical challenge. Liu *et al.*<sup>[8]</sup> utilized a 3-D printed, customized self-stabilizing artificial vertebral body to treat primary Ewing's sarcoma involving the C2 vertebra in a

12-year-old male following complete C2 excision. However, there are limited data regarding the long-term efficacy of such implants, and this was not an option in our case as the procedure was emergent, and the tumor encased the vertebral artery which we were not willing to sacrifice.

Although the axial location of a Ewing's sarcoma is a poor prognostic factor,<sup>[4]</sup> the combined use of chemotherapy and radiotherapy has improved the overall prognosis/survival in the past two decades.<sup>[3]</sup>

### CONCLUSION

Primary Ewing's sarcomas involving the C2 vertebrae are exceedingly rare. Optimal treatment typically includes adequate radiological evaluation, biopsy of the lesion to obtain immunohistopathological confirmation of the diagnosis, decompression where indicated, and postoperative multimodal adjunctive chemotherapy.

### Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Nil.

### Conflicts of interest

There are no conflicts of interest.

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