

## Ewing Sarcoma Cervical Spine Rare Case Report With Review of Literature



### Medical Science

KEYWORDS :

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### ABSTRACT

*Ewing sarcoma of cervical spine is a rare presentation.. These rapidly growing tumors rarely present as cervical mass before neurological symptoms of myelopathy or radiculopathy. In this report, we discuss a primary cervical spine Ewing sarcoma in a 15-year-old child who presented with a 2 week history of left cervical mass . Imaging revealed a cervical vertebral mass.*

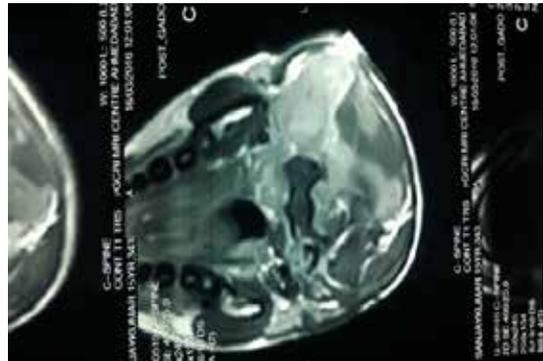
### INTRODUCTION

Ewing's sarcoma is a primary bone malignancy with the highest incidence in the second decade of life. Although it mostly affects the metaphyseal region of long growing bones, involvement of spine is not very uncommon especially the sacrum. Nonsacral spinal Ewing's sarcoma is rarer and often mimics a benign condition before spreading extensively. They present with neurologic deficits due to spinal cord compression, but acute onset paraplegia has not been previously reported. A high index of clinical suspicion can clinch the diagnosis early in the course of the disease. A prompt intervention is required to keep neurological damage to a minimum, and a correct combination of surgery, chemotherapy, and radiotherapy is required for better long-term patient outcome. We report a 16-year-old female who presented with acute paraplegia and had an excellent postoperative outcome after radical excision of a D9 Ewing's sarcoma.

Primary malignant sarcomas of the spine are rare and they account for only 3.5%–14.9% of all primary bone sarcomas.(1) Ewing's sarcoma is the second most common primary bone tumor in pediatric patients accounting for approximately 4% of pediatric malignancies.[2] Its incidence is highest in the second decade of life and most commonly involves the long bones of the extremities and the pelvis.Primary involvement of the nonsacral spine represents approximately 0.9% of all cases(5).In this cervical spine involvement is even more uncommon (4). A high index of suspicion is essential for diagnosis, especially in a young patient. We report a rare case of cervical spine Ewing's sarcoma that presented with left side cervical swelling followed by quadriparalysis.

### CASE REPORT

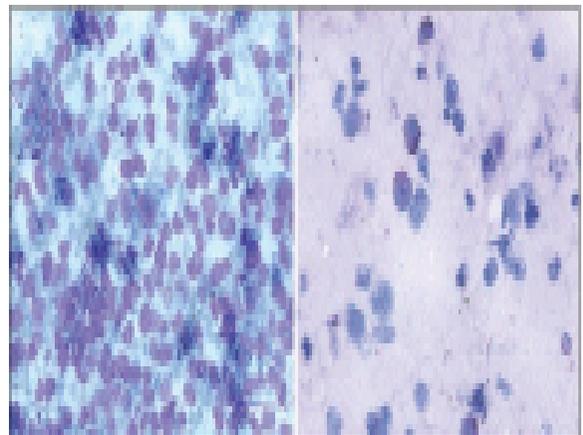
We report the case of a 15 year-old male presenting with a progressively increasing painful swelling in left lower cervical region from last 2 weeks. Physical examination revealed a firm, fixed, tender swelling measuring 8× 7 cm over the left lower cervical region[Figure 4]. Neurological examination showed sensory and motor weakness in left upper limb.



**FIGURE 1,2 MRI revealing paraspinal mass involving of C2to C4 vertebra**

On magnetic resonance imaging, a mass lesion of around 7 × 4 × 6 size lesion present in left paraspinal lesion at level C1 to C5 vertebra, hypointense on T1W and hyper intense on T2W image, involving left half body of C2to C4 vertebra and extend into spinal cord.

A true cut biopsy was performed The cytological diagnosis of small round cell tumor, Ewing's sarcoma was made, 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.





**FIGURE 3.4. biopsy smear showing small, round cells in loose cohesive clusters, rosettes and dispersed singly (MGG,  $\times 200$ ), (showing small, round cells in loose cohesive clusters and dispersed singly with positivity for cytoplasmic glycogen .**

### 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%).[2]

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.(3)

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 tumors of the spine in children. Lesions of the spine account for 3%-5 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.(4) 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., Ewing's tumors tend to be extensive, sometimes involving an entire bone.. Typically, Ewing's tumor produces a pronounced reactive new bone formation of the periosteum, giving rise to an onion-skin appearance on roentgenograms.(4)

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

In the present case, the patient was treated with chemotherapy cycles , to which he responded well.

### REFERENCES

1. Murphey MD, Andrews CL, Flemming DJ, Temple HT, Smith WS, Smirniotopoulos JG. From the archives of the AFIP. Primary tumors of the spine: Radiologic pathologic correlation. *Radiographics*. 1996;16:1131-58. [PubMed]
2. Bawa SS, Hegde KK. Primary ewings sarcoma of the temporal bone- a case report. *Indian J Radiol Image*. 2006;16:623-5.
3. Seçkin H, Varan A, Demirhan B, Altinok D, Kars HZ. Primary ewings sarcoma of the cranium: A case report. *Turkish Neurosurg*. 2001;11:130-3.
4. Subbarao K, Jacobson HG. Primary malignant neoplasms. *Semin Roentgenol*. 1979;14:44-57. [PubMed]
5. Grubb MR, Currier BL, Pritchard DJ, Ebersold MJ. Primary Ewing's sarcoma of the spine. *Spine (Phila Pa 1976)* 1994;19:309-13. [PubMed]