

Case Report

A rare case of acute paraplegia: primary Ewing's Sarcoma of dorsal spine

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ABSTRACT

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. Non-sacral 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 11-year-old female who presented with acute paraplegia and had an excellent postoperative outcome after radical excision of a C7-D2 Ewing's sarcoma.

Keywords: Ewing's sarcoma, Paraplegia, Primary spine tumor, Spinal cord compression

INTRODUCTION

Primary malignant sarcomas of the spine are rare, and they account for only 3.5%-14.9% of all primary bone sarcomas. Ewing's sarcoma is the second most common primary bone tumor in pediatric patients accounting for approximately 4% of pediatric malignancies.¹ Its incidence is highest in the second decade of life and most commonly involves the long bones of the extremities and the pelvis, presenting primarily as swelling with pain. Primary vertebral Ewing's sarcoma has been divided into sacral and nonsacral types based on the differences in the treatment responses and survival rates. Primary involvement of the nonsacral spine represents approximately 0.9% of all cases. Low back pain is the most common symptom followed by a palpable swelling. Spinal cord compression can produce neurological deficits depending on the tumor location but is often a delayed presentation. Rapidly progressing paraplegia is

uncommon and a high index of suspicion is essential for diagnosis, especially in a young patient. We report a rare case of dorsal spine Ewing's sarcoma that presented with acute onset paraplegia and improved with radical tumor decompression. Ewing's sarcoma in the sacral spine is common but in the non-sacral spine is very rare accounts to 0.9%.

CASE REPORT

A 11-year-old female child presented with a 1 month history of back pain (mid dorsal) for which she was treated with analgesics at a local hospital. She presented to us with rapidly progressing weakness and numbness of both lower limbs over 1 week. She was bed bound for 3 days. She had bowel and bladder dysfunction for 1 week. There was no antecedent history of trauma / head injury / bony injury. There was no history of developmental delay. Child immunized till date. On CNS examination –

higher functions normal, decrease in sensation in both lower limbs, Power – 0/5 in both lower limbs, Atonia, Bladder catheterised, Perianal sensation absent. CT-Chest and Abdomen showed no other lesion in the patient. Contrast Magnetic resonance imaging revealed homogenously enhancing vertically oblong extradural lesion extending to 4 dorsal levels in posterior and lateral epidural compartments with D2 vertebral posterior bony element involvement (suggestive of neoplasm) In view of the rapid progression of weakness, the patient was considered for emergency Laminectomy C7-D4 and dorsal spine extradural tumour excision done.



Figure 1: Pre-op picture.

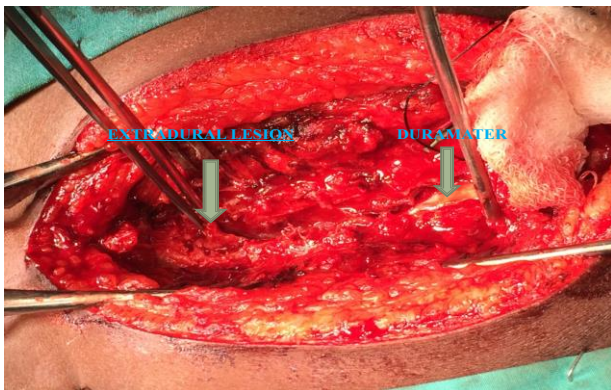


Figure 2: Intraop –laminectomy with excision of the extradural lesion.



Figure 3: Intraop – complete excision of the extradural lesion.



Figure 4: Contrast MRI STUDY showing dorsal spine extra-dural lesion.

Intraoperatively, Extradural lesion compressing the spinal cord was soft, vascular, jelly like in consistency excised from C7- D4. The neurological outcome of surgery was good. Power in the lower extremities gradually increased by 1/5, and within 2 weeks after the surgery, the patient was ambulatory. She was referred for adjuvant systemic chemotherapy and local radiotherapy. Histopathology report was suggestive of a small round blue cell tumor that was confirmed as Ewing's sarcoma of dorsal spine by immunohistochemistry.

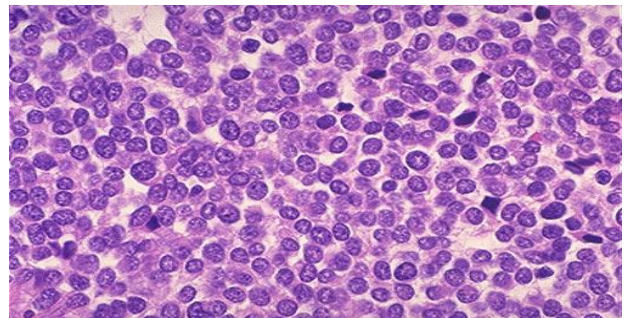


Figure 5: HPE- Ewings sarcoma of dorsal spine.

DISCUSSION

The complex neuromusculoskeletal development of the spine may account for a spectrum of malignant tumors with distinct biological behaviours.¹ The differential diagnosis of a small round cell bone tumor includes Ewing's sarcoma, neuroblastoma, primitive neuroectodermal tumor of bone, malignant lymphoma, and rhabdomyosarcoma.²

Ewing's sarcoma mostly occurs in childhood when the bones are growing.³ It commonly affects the metaphyseal plates of long bones and rarely involves the spine where the most common location is the sacrum.⁴ Although there have been reports of nonsacral spinal involvement but these are rare.⁵

Ewing's sarcoma is often a multifocal disease, and a proper pre-staging workup including bone scan is required before institution of therapy and for follow-up

Systemic symptoms may also be present. Involvement of nonsacral spine usually presents with features of spinal cord compression which is often late in the course of disease.⁶ The expansile nature of the lesion makes local swelling and pain as the most common presentation in cases of long bone involvement. Presentation as acute paraplegia without significant localized pain and swelling has not been reported in the literature.

Early diagnosis and treatment in Ewing's sarcoma can result in favorable outcomes. The presence of benign musculoskeletal symptoms often leads to a delay in diagnosis with many patients being misdiagnosed and treated for disc disease. Unlike other malignant spinal lesions that cause progressive and continuous pain that increases with recumbency, in majority of nonsacral spinal Ewing's sarcoma pain is often intermittent, without nocturnal exacerbation.⁷ This intermittent progression versus an expected rapid course is a reason for further delay in diagnosis. Thus, it is clear that diagnosis of nonsacral spinal Ewing's sarcoma will require a high index of suspicion.

A detailed patient history and a careful physical examination supplemented by imaging are essential to minimize the delay.

When deciding about the treatment of Ewing's sarcoma of the mobile spine, the most important determinant is the presence of neurological deficits, which once present are often rapidly progressive. In such circumstances, only a prompt surgical decompression can provide maximum chance of recovery.⁸ The approach is defined by the type of involvement. Anterior decompression is warranted in cases where cord compression is due to extension from the body.

Ewing's sarcoma often tends to invade the spinal canal from the paravertebral soft tissue component through the intervertebral foramen, compressing the cord circumferentially. This makes laminectomy an effective approach for cord decompression.⁹ In either of the cases, postoperative chemotherapy for control of micrometastases and local control by radiotherapy is warranted. In cases where the diagnosis is being anticipated prior to neurological compromise, it is advisable to confirm it by needle biopsy, and once made, the patient should be subjected to a three- or four-drug neoadjuvant chemotherapy regimen.¹⁰ This would not only help shrink the primary tumor, thereby increasing chances of total excision, but also take care of micrometastasis and give an idea about responsiveness of the tumor to adjuvant therapy.¹¹ This is followed by surgery or radiotherapy or both. Primary radiotherapy is not advocated in these cases because the posttreatment edema will lead to development or progression of neural compression.

In our patient, the initial presentation of mild low back pain was misinterpreted as due to benign spinal pathology

and managed medically. But the onset of rapid lower limb weakness pointed toward a more aggressive pathology. Acute paraplegia as a presenting symptom is extremely rare in Ewing's sarcoma of the nonsacral spine and requires a high index of suspicion in children for early diagnosis. A detailed patient history and a careful physical examination are essential to lower down the delay in diagnosis. An atypical clinical course in a musculoskeletal or neurological condition should alert us to a possible underlying malignant disease.

CONCLUSION

Ewing's sarcoma of dorsal spine can present with acute onset and rapidly progressive neurological deficit and needs high index of suspicious and urgent intervention for good outcome.

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REFERENCES

1. Bacci G, Picci P, Gherlinzoni F, Capanna R, Calderoni P, Putti C, et al. Localized Ewing's sarcoma of bone: Ten years' experience at the Istituto Ortopedico Rizzoli in 124 cases treated with multimodal therapy. *Eur J Cancer Clin Oncol.* 1985;21:163-73.
2. Bemporad JA, Sze G, Chaloupka JC, Duncan C. Pseudohemangioma of the vertebra: An unusual radiographic manifestation of primary Ewing's sarcoma. *AJNR Am J Neuroradiol.* 1999;20:1809-13.
3. Bruckner JD, Conrad EU., Spine. In: Simon MA, Springfield D, editors. *Surgery for bone and soft-tissue tumours.* Philadelphia: Lippincott-Raven; 1998:435-50.
4. Dini LI, Mendonça R, Gallo P. Primary Ewing's sarcoma of the spine: Case report. *Arq Neuropsiquiatr.* 2006;64:654-9.
5. Ebb DH, Green DM, Shamberger RC, Tarbell NJ. Solid tumours of childhood. In: DeVita VT Jr, Hellman S, Rosenberg SA, editors. *Cancer Principles and Practice of Oncology.* Philadelphia: Lippincott; 2005:1919-23.
6. Estes DN, Magill HL, Thompson EI, Hayes FA. Primary Ewing's sarcoma: Follow-up with Ga-67 scintigraphy. *Radiology.* 1990;177:449-53.
7. Grubb MR, Currier BL, Pritchard DJ, Ebersold MJ. Primary Ewing's sarcoma of the spine. *Spine (Phila Pa 1976)* 1994;19:309-13.
8. Pilepich MV, Vietti TJ, Nesbit ME, Tefft M, Kissane J, Burgert O, et al. Ewing's sarcoma of the vertebral column. *Int J Radiat Oncol Biol Phys.* 1981;7:27-31.
9. Gopalakrishnan CV, Shrivastava A, Easwer HV, Nair S. Primary Ewing's sarcoma of the spine

- presenting as acute paraplegia. *J Pediatric Neurosci.* 2012 Jan;7(1):64.
10. Sharafuddin MJ, Haddad FS, Hitchon PW, Haddad SF, el-Khoury GY. Treatment options in primary Ewing's sarcoma of the spine: Report of seven cases and review of the literature. *Neurosurgery.* 1992;30:610-8.
 11. Widhe B, Widhe T. Initial symptoms and clinical features in osteosarcoma and Ewing sarcoma. *J Bone Joint Surg Am.* 2000;82:667-74.

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