

Primary Spinal Nonsacral Ewing's Sarcoma with an Unusual Presentation- a Case Report

Bakr Abo-Jarad**, Hazem Kuheil**, Hani Alanqar***

*Medical Student, Islamic University of Gaza Medical School, Gaza, Palestine

**Neurosurgical Department, Shifaa Hospital, Gaza, Palestine

***Pathological Department, Shifaa Hospital, Gaza, Palestine

Abstract

Primary malignant sarcomas of the spine are not among common types of primary bone sarcomas. There are only few cases reported on the literature about this tumor. Ewing sarcoma occurring in the spine is divided into two types: 1) sacral spine Ewing sarcoma, which is very aggressive with poor prognosis and 2) non-sacral spine Ewing sarcoma, which is an extremely rare occurrence. The patient may present neurological deficit when the tumor extends into the spinal canal causing spinal cord compression; however, a sudden progressive paraplegia also is very rare. Here, we report a case of 8 years girl with sudden weakness and tingling in both lower limbs, inability to walk, progressive neck and upper dorsal pain, and urinary retention problem. Spinal MRI shows extradural mass from T2 to C7 with severe spinal cord compression. Urgent decompressive laminectomy and GTR of the lesion was done with excellent postoperative outcome

Keywords: Ewing sarcoma, Spine, Primitive neuroectodermal tumor

Introduction

Primary Ewing sarcoma is a rare tumor that arises from the undifferentiated matrix.¹ The annual incidence of this tumor is 0.2-0.4 per 1000,000.² This highly malignant tumor, which has a poor prognosis, usually occurs in children and young adults with an age range of 4-15 years.^{1, 3} The primary malignant sarcomas of the spine are rare and they account only 3.5%-14.9% of all primary bone sarcoma.⁴ The most

common presentation of this health problem is back pain. Although spinal compression can occur, the latest finding shows that it is rare to be accompanied by acute paraplegia. We presented here an 8-year-old girl that has no significant medical history and presents rapid deterioration in lower limb motor and sensory function, which end as paraplegia.

Corresponding Author:

Bakr Abo Jarad, Medical Student
860, Yafa Street, Gaza, Gaza
strip, Palestine
Tel: +970598786546.
Email: www.bakerkhader@hotmail.com

Case report

Here, we examined cases in neurosurgery department diagnosed with non-sacral Ewing sarcoma, which has a rare presentation from 2016 and such that we only found two cases that they almost have the same picture. Written informed consent was taken from the patients' families to use their data for the scientific purpose.

Case 1

An 8-year-old girl with no significant medical history referred to our hospital due to progressive lower cervical and interscapular pain for 10 days, developed abnormal gait and rapidly progressive lower limbs weakness, urinary retention, no history of trauma, and no constitutional symptoms. Examination revealed bilateral spastic paraplegia and hypoesthesia in both lower limb to all sensations below D2 dermatome. The reflexes in

the lower limb were exaggerated and Babinski was positive bilaterally. There was no evidence of Paraspinal tenderness or swelling. The patient underwent magnetic resonance imaging (MRI) (Figure 1) of the cervicodorsal spine, which revealed extradural hypointense mass extending from C7 to T2, which was much higher in the left side in the spinal canal. Also, it was oval-shaped and extended to the paraspinal region and to the left side of the chest cavity through intervertebral foramina with severe spinal cord compression and severe central canal stenosis. The patient underwent operation including C7-D2 laminectomy with total resection of the tumor from the spinal canal and from the chest cavity with preserved facet joint in all levels (Figure2). The neurological outcome of surgery was very well, the patient was improved rapidly with motor power in Rt 5/5 and Lt 4/5, sensation intact after

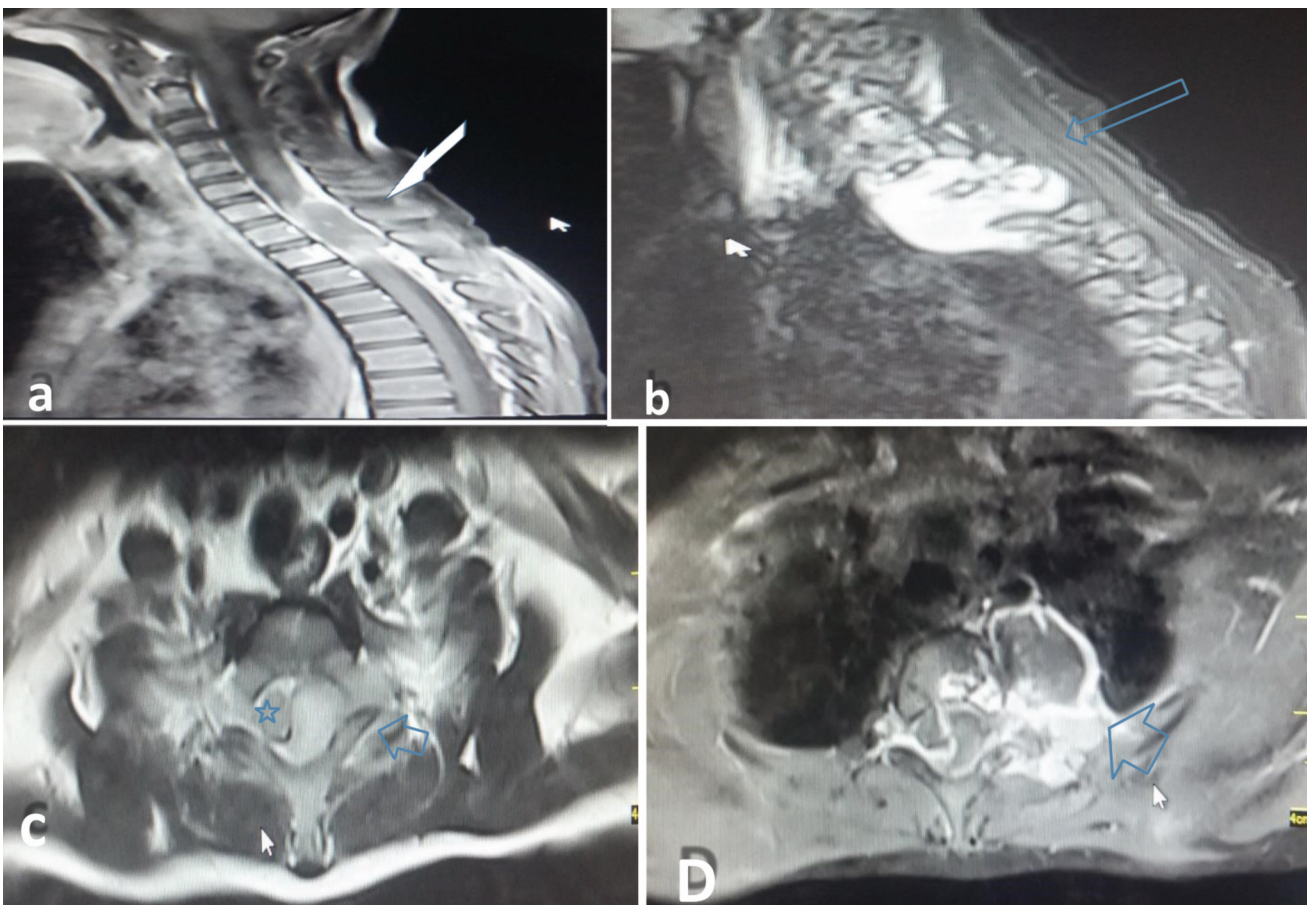


Figure 1. Magnetic resonance imaging of cervicodorsal spine (a) T1 sagittal showing hypointense lesion compressing the spinal cord, (b) T2 sagittal showing hyperintense lesion compressing the spinal cord, (c) T2 axial without contrast showing hypointense tumor (arrowhead) compressing the spinal cord to the right, and (d) T1 axial with contrast showing tumor heterogeneity enhancing to contrast (arrowhead) compressing the spinal cord to the right.

Table 1. Complaint and outcomes of extraskelatal Ewing's sarcoma cases.

Author/year	Age/sex	Complaint	Level of tumor	Outcome
Virani et al. 2002 ¹⁴	5 Yr M	Neck and left leg pain	C7-D1	Asymptomatic patient
Harimaya et al. 2003 ¹⁵	10 yr M	Progressive paresis	C6-D3	Death of patient
Kogawa et al. 2004 ¹⁶	7Yr M	Neck pain with weakness in left arm	C2-C4	Asymptomatic with cervical deformity
Erkutlu et al. 2007 ¹⁷	7Yr M	Progressive weakness and numbness in LL and nuchal rigidity	C5-T1	Asymptomatic patient
Kumar et al. 2007 ¹⁸	8Yr M	Painful neck swelling with progressive quadriplegia	C2-C4	Recurrence of tumor
Duan et al. 2011 ¹⁹	7yr F	Persistent pain with the weakness of limbs	C6-T2	Asymptomatic patient
Khmou et al. 2016 ²⁰	5yr M	Torticollis	C1-C7	Asymptomatic patient
Kutty et al. 2017 ²¹	12yr F	Neck pain with tetraplegia	C2-C4	Asymptomatic patient
Our case	8yr F	Neck pain with paraplegia	C7-D2	Asymptomatic patient

1 month, after one year patient has totally normal motor power in both limbs with no evidence of recurrent and need to undergo spinal fixation, which was required to be done after 1 year.

Histopathological examination (Figure 3)

revealed that the tumor consisted of small, round, malignant cells with hyperchromatic nuclei, scant cytoplasm, and brisk mitotic figures. Immunohistochemically, the tumor cells showed intense membrane reactivity for CD99.

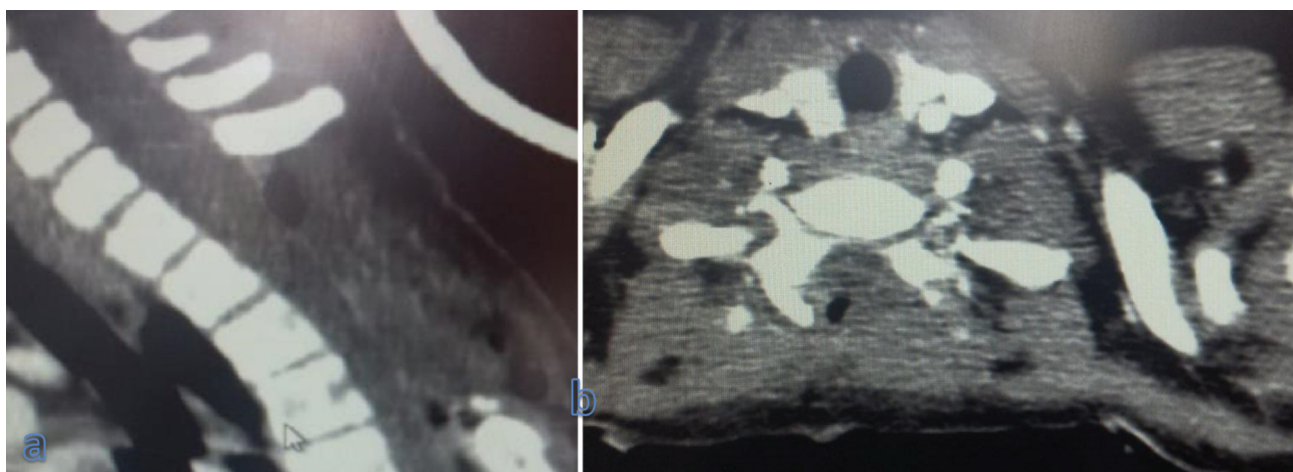


Figure 2. Computed tomography scan of cervicodorsal spine (a) sagittal view showing C7-T1 postoperative and removal of all the tumor and (b) axial view showing spinal cord that became free after removing the tumor.

Discussion

Ewing sarcoma, which is a primary bone tumor, was defined in details by James Ewing in 1921.⁵ Internationally, the annual incidence rate is approximately three cases per million children.⁶ Ewing sarcoma frequently affects patients in the second decade with an increased incidence in males above the age of 13 years.⁷

Most of the Ewing sarcoma occurs in the long bones, pelvis, or ribs.⁸ An extraskeletal origin that named extraskeletal Ewing sarcoma has similar histology to skeletal Ewing sarcoma, which commonly affects the epidural spaces and paravertebral regions and most common in the sacral spine. Primary involvement of the nonsacral spine represents approximately 0.9% of all cases.⁹ The dorsal vertebrae are involved in 1% of cases.¹⁰ Non-sacral Ewing sarcoma usually shows signs of spinal cord compression, which is often late in the course of the disease. Unlike other malignant spinal lesions, which cause progressive and continuous pain and increase with recumbency, in the majority of non-sacral spinal Ewing sarcoma, pain is often intermittent and without nocturnal exacerbation.¹¹

In our review, we found eight cases below 12 years that had extradural Ewing sarcoma in the cervical region from 2002. Among these cases, five from eight cases (5/8) have no symptom now, only one case died, and one case had a

recurrence of symptoms (Table 1). The most common symptom was neck pain that happens in five from eight cases (5/8).

Rapidly progressing paraplegia is uncommon and a high index of suspicion is essential for diagnosis, especially in a young patient.¹² Our case of cervicodorsal spine Ewing sarcoma presented an acute onset of progressive paraplegia. Ewing sarcoma often tends to invade the spinal canal from the paravertebral soft tissue component through the intervertebral foramen and compressing the cord circumferentially. This makes laminectomy an effective approach for cord decompression.¹³ Due to the aggressive behavior of the neoplasm and its great potential to metastasis, treatment should be multimodal involving radical excision, radiotherapy, and chemotherapy.¹

Conflict of Interest

None declared.

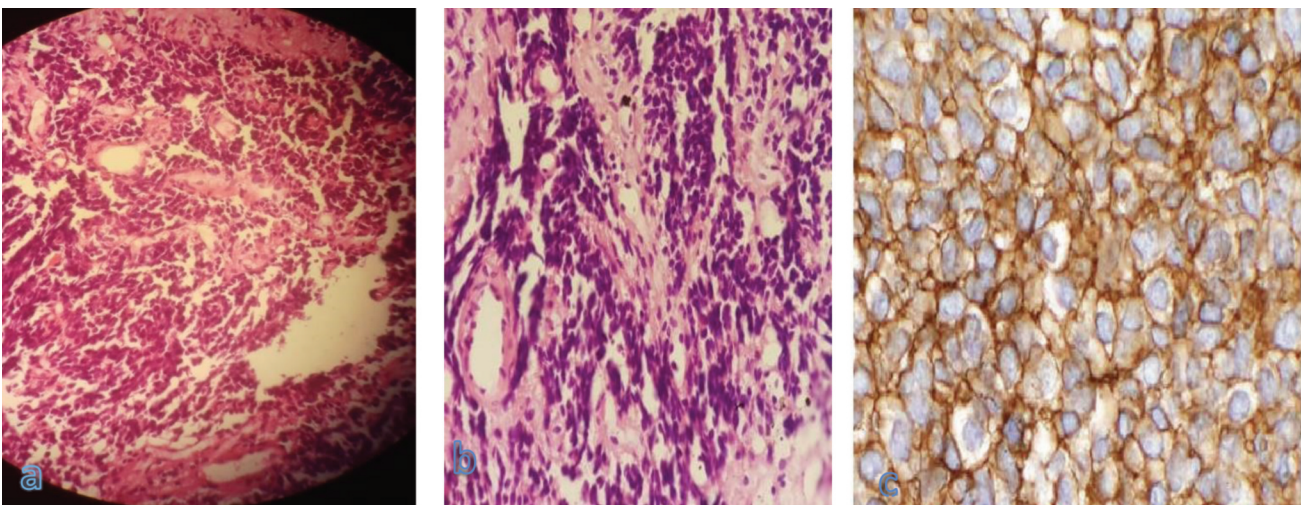


Figure 3. (a) The low power photo of a vaguely lobular appearance with monotonous cells (H&E: 20×), (b) High magnification shows admixed population of degenerating tumor cells with smaller hyperchromatic and angulated nuclei (H&E: 20×), and (c) Immunohistochemical staining for CD99: the tumor cells show membranous staining for CD99 (CD99: 400×).

References

1. Saeedinia S, Nouri M, Alimohammadi M, Moradi H, Amirjamshidi A. Primary spinal extradural Ewing's sarcoma (primitive neuroectodermal tumor): Report of a case and meta-analysis of the reported cases in the literature. *Surg Neurol Int.* 2012;3:55. doi: 10.4103/2152-7806.96154.
2. Munjal S, Srivastava A, Tucker S, Bakshi N, Bhalla S, Mehta VS. Primary dorsal spine primitive neuroectodermal tumor in an adult patient: Case report and literature review. *J Craniovertebr Junction Spine.* 2017;8(4):378-81. doi: 10.4103/jcvjs.JCVJS_70_17.
3. Ellis JA, Rothrock RJ, Moise G, McCormick PC 2nd, Tanji K, Canoll P, et al. Primitive neuroectodermal tumors of the spine: a comprehensive review with illustrative clinical cases. *Neurosurg Focus.* 2011;30(1):E1. doi: 10.3171/2010.10.FOCUS10217.
4. Margaix-Muñoz M, Bagán J, Poveda-Roda R. Ewing sarcoma of the oral cavity. A review. *J Clin Exp Dent.* 2017;9(2):e294-e301. doi: 10.4317/jced.53575.
5. Ginsberg JP, Goodman P, Leisenring W, Ness KK, Meyers PA, Wolden SL, et al. Long-term survivors of childhood Ewing sarcoma: report from the childhood cancer survivor study. *J Natl Cancer Inst.* 2010;102(16):1272-83. doi: 10.1093/jnci/djq278.
6. Esiashvili N, Goodman M, Marcus RB Jr. Changes in incidence and survival of Ewing sarcoma patients over the past 3 decades: Surveillance epidemiology and end results data. *J Pediatr Hematol Oncol.* 2008;30(6):425-30. doi:10.1097/MPH.0b013e31816e22f3.
7. Ilaslan H, Sundaram M, Unni KK, Dekutoski MB. Primary Ewing's sarcoma of the vertebral column. *Skeletal Radiol.* 2004;33(9):506-13.
8. Salunke PS, Gupta K, Malik V, Kumar N, Henke LE, Cai C, et al. Primary Ewing's sarcoma of cranial bones: analysis of ten patients. *Acta Neurochir (Wien).* 2011;153(7):1477-85. doi: 10.1007/s00701-011-1028-z.
9. Bordia S, Meena S, Meena BK, Rajak V. Fine-needle aspiration cytology of Ewing's sarcoma of thoracic spine with extension into the intradural space. *Case Rep Oncol Med.* 2014;2014:351386. doi: 10.1155/2014/351386.
10. Bordia S, Meena S, Meena BK, Rajak V. Fine-needle aspiration cytology of Ewing's sarcoma of thoracic spine with extension into the intradural space. *Case Rep Oncol Med.* 2014;2014:351386. doi: 10.1155/2014/351386.
11. Widhe B, Widhe T, Bauer HC. Ewing sarcoma of the rib--initial symptoms and clinical features: tumor missed at the first visit in 21 of 26 patients. *Acta Orthop.* 2007;78(6):840-4. doi: 10.1080/17453670710014644.
12. Gopalakrishnan CV, Shrivastava A, Easwer HV, Nair S. Primary Ewing's sarcoma of the spine presenting as acute paraplegia. *J Pediatr Neurosci.* 2012;7(1):64-6. doi: 10.4103/1817-1745.97630.
13. Dini LI, Mendonça R, Gallo P. Primary Ewing's sarcoma of the spine: case report. *Arq Neuropsiquiatr.* 2006;64(3A):654-9.
14. Virani MJ, Jain S. Primary intraspinal primitive neuroectodermal tumor (PNET): a rare occurrence. *Neurol India.* 2002;50(1):75-80.
15. Harimaya K, Oda Y, Matsuda S, Tanaka K, Chuman H, Iwamoto Y. Primitive neuroectodermal tumor and extraskeletal Ewing sarcoma arising primarily around the spinal column: report of four cases and a review of the literature. *Spine (Phila Pa 1976).* 2003;28(19):E408-12.
16. Kogawa M, Asazuma T, Iso K, Koike Y, Domoto H, Aida S, et al. Primary cervical spinal epidural extraosseous Ewing's sarcoma. *Acta Neurochir (Wien).* 2004;146(9):1051-3; discussion 1053.
17. Erkutlu I, Buyukhatipoglu H, Alptekin M, Ozsarac C, Buyukbese I, Gok A. Primary spinal epidural extraosseous Ewing's sarcoma mimicking a spinal abscess. *Pediatr Hematol Oncol.* 2007;24(7):537-42.
18. Kumar R, Reddy SJ, Wani AA, Pal L. Primary spinal primitive neuroectodermal tumor: case series and review of the literature. *Pediatr Neurosurg.* 2007;43(1):1-6.
19. Duan XH, Ban XH, Liu B, Zhong XM, Guo RM, Zhang F, et al. Intraspinial primitive neuroectodermal tumor: imaging findings in six cases. *Eur J Radiol.* 2011;80(2):426-31. doi: 10.1016/j.ejrad.2010.06.005.
20. Khmou M, Malihi A, Lamalmi N, Rouas L, Alhamany Z. Peripheral primitive neuroectodermal tumors of the spine: a case report and review of the literature. *BMC Res Notes.* 2016;9(1):438. doi: 10.1186/s13104-016-2246-5.
21. Kutty RK, Peethambaran A, Sunilkumar BS, Balachandran Nair KG, Korde P, Jain SK. Ewing sarcoma of the cervical epidural space presenting with tetraplegia: Case report and review of literature. *World Neurosurg.* 2017;107:1046.e9-1046.e15. doi: 10.1016/j.wneu.2017.07.182.