Spinal Cord (2002) 40, 367–369

www.nature.com/sc

*Case Report*

# Paraplegia: an unusual presentation of Ewing's sarcoma

AS Goktepe*,[1], R Alaca[1], H Mohur[1] and U Coskun[1]

[1]*Turkish Armed Forces Rehabilitation and Care Center, Gulhane Military Medical Academy, Ankara/Turkey*

**Study design:** Single-subject case.
**Objectives:** To describe the atypical presentation of a rare tumor and difficulties in diagnosis.
**Setting:** Spinal cord unit of a rehabilitation and care center in Ankara, Turkey.
**Methods:** A 22-year-old male patient with paraplegia was admitted to our center for rehabilitation. He underwent various diagnostic procedures to explain his clinical situation.
**Results:** Abdominal ultrasound and computed tomography revealed a mass and vertebral destruction. The pathology report confirmed the diagnosis of Ewing's sarcoma.
**Conclusion:** Spinal cord compression is an important complication of primary or metastatic malignant tumors. Although it is a late complication in most cases, some tumors including Ewing's sarcoma may present with paraplegia. Trauma may be found in the patient's history. An atypical clinical course in a musculoskeletal or neurological condition should alert us to a possible underlying malignant disease.
*Spinal Cord* (2002) **40,** 367–369. doi:10.1038/sj.sc.3101326

**Keywords:** Ewing's sarcoma; paraplegia; spinal cord compression; delay in diagnosis

## Introduction

Ewing's sarcoma is a small, round cell tumor which accounts for one-quarter of all primary bone tumors during childhood.[1] It has a peak incidence during the second decade and is very rare after 30 years of age.[2] Typical complaints of patients with Ewing's sarcoma are pain and swelling of the affected bone. Systemic symptoms may also exist. The most commonly affected bones are the femur, pelvis and other long bones of the extremities. Vertebrae are affected in less than 5% of the cases and may present with nerve root or spinal cord compression. The prognosis is usually poor. However, multimodality therapy has increased the 5 year survival rate to about 40%.[3]

## Case report

A 22-year-old male was admitted to our rehabilitation center with paraplegia and severe right leg pain. His medical history had started with a complaint of low back pain following minor trauma (a fall during running) 10 months before his admission. He had no previous pain or systemic symptoms such as fever, weight loss and fatigue. Direct radiographs were taken and no pathologic finding was detected. He was given analgesic medications, but his persisting pain led to further investigation. Magnetic resonance imaging (MRI) revealed a fracture of the third lumbar vertebra. After having a lumbosacral orthosis for 3 months his complaints were partially relieved. Four weeks prior to his admission, he underwent two consecutive operations in a Neurosurgery Department for his increasing pain and rapidly progressing paraplegia. The first operation was done to decompress the spinal roots and included total laminectomy and bilateral foraminotomy of the third lumbar vertebra as well as granulation tissue debridement. Posterior transpedicular fixation was performed in the second operation. Having limited improvement from these operations he was referred to our rehabilitation center.

On admission, the patient was suffering from severe pain which did not respond to opioids. Manual muscle testing showed strength at grade 0 in his lower extremities with the following exceptions: Left knee extension was at grade 1, right toe dorsiflexion grade 2, left toe dorsiflexion grade 3 and ankle plantar flexion grade 4 on both sides. Light touch sensation was decreased below the first lumbar dermatome. Pin prick sensation was decreased in the third lumbar dermatome, while other dermatomes were normal. He had normal proprioceptive sensation. The muscle stretch reflexes of his lower extremities were absent. The Babinski response was indifferent on the right side

*Correspondence: AS Goktepe, TSK Rehabilitasyon ve Bakim Merkezi, 06530 Bilkent-Ankara, Turkey

and flexor on the left side. He had flaccid paralysis, his bladder and bowel functions were found to be normal. Voluntary anal sphincter function and perianal sensation were also preserved.

Laboratory assessments revealed a slightly increased erythrocyte sedimentation rate (32 mm/h). The level of his serum lactate dehydrogenase was 278 (range, 91–232) U/l. He had a normal white blood cell count and a normal alkaline phosphatase level. Other routine laboratory data indicated no abnormalities.

Venous Doppler ultrasound of his lower extremities showed a slow response to augmentation and Valsalva maneuver which aroused suspicion of an intra-abdominal pathology. Abdominal ultrasound revealed a $10 \times 9 \times 8$ cm mass located in the retroperitoneal area. It was encircling the abdominal aorta and the inferior vena cava. A partial thrombus was formed near the bifurcation of the inferior vena cava presumably due to compression of the mass. CT scan confirmed the mass and showed the destructive lesion in the third lumbar vertebra (Figure 1A,B).

A more detailed patient history revealed the fact that a specimen was taken during the first operation and sent to the pathology laboratory. Following completion of the operations, the patient was discharged from the hospital and was advised to go to a rehabilitation center. He left the city and applied to our center for rehabilitation. After getting that information the pathology report was requested from the prior hospital. The report provided the diagnosis of Ewing's sarcoma. A preoperative X-ray and MRI were also requested from the same hospital. Both preoperative and postoperative radiographs showed an anterior wedging of the third lumbar vertebral body with mottled appearance (Figure 2A,B). The MRI demonstrated pathological vertebral fracture and anterolateral soft tissue mass (Figure 3A–C). Two days after his admission the patient was referred to the Oncology Department. He underwent a combined radiotherapy and chemotherapy protocol. He improved quickly; got rid of his pain and started to walk. Two months later, his gross muscle strength was almost at grade five. All the other neurological abnormalities had also disappeared, although his prognosis was still unclear.

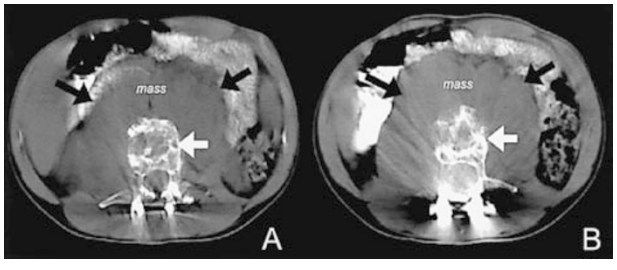


**Figure 1** (**A,B**) Two consecutive slices of CT demonstrating the destruction of the third lumbar vertebral body (white arrows) and the large mass encircling the vertebra (black arrows)

## Discussion

Primary Ewing's sarcoma of the spine is a condition that occurs infrequently.[4] Cases with primary involvement of the nonsacral spine represent approximately 0.9% of all cases.[5] It has been stated that these tumors are so rare that most doctors will see only a few patients with symptoms from an undiagnosed primary bone tumor during their whole working life.[6]

In our case the diagnosis was delayed 2 or 3 weeks because of poor medical follow-up. Total delay in diagnosis was 40 weeks. Grubb *et al* reported 1–124 weeks delay in their retrospective study of primary Ewing's sarcoma of the spine.[4] However, the mean delay was 8 weeks when sacral lesions were excluded. Widhe *et al* reported a mean delay of 34 weeks (ranging from 3 to 150) in Ewing's sarcoma cases with different locations.[6] In their study, 12 out of 47 patients consulted a doctor more than six times before the tumor was diagnosed, and one patient had to see a

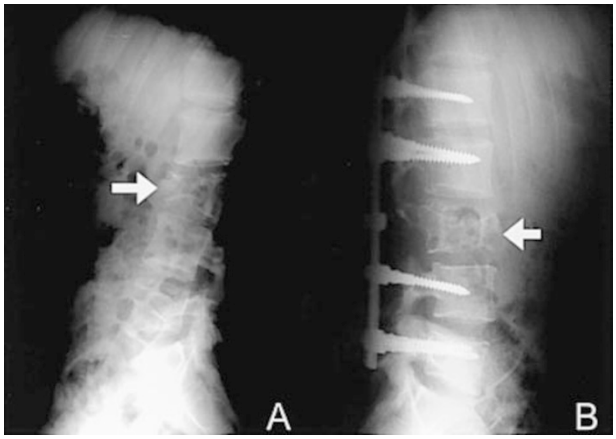


**Figure 2** (**A,B**) Preoperative and postoperative lateral radiographs show anterior wedging of the third lumbar vertebral body with moth-eaten appearance (arrows)

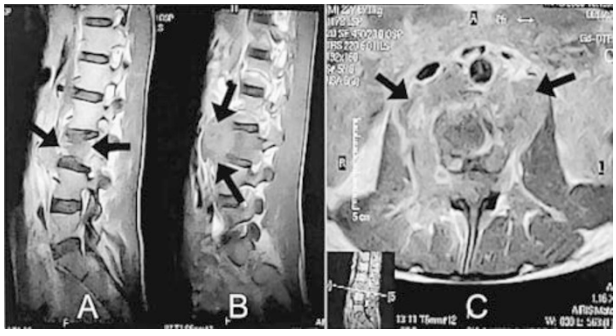


**Figure 3** (**A**) T1 weighted sagittal MRI demonstrates low signal intensity replacement of normally bright marrow of the third lumbar vertebra (arrows) which is indicative for a pathological fracture. (**B,C**) T1 weighted sagittal and axial images show anterolateral soft tissue mass around the same vertebra (arrows)

doctor 23 times about symptoms related to the bone tumor.

Early diagnosis is of great importance in Ewing's sarcoma, because favorable results are reported with early treatment.[5] Nevertheless, several factors may delay the diagnosis. To start with, the musculoskeletal symptoms due to tumor may lead the patient to doctors from different medical specialties that are not always experienced in malignancies. Therefore, most patients are diagnosed with benign musculoskeletal disorders. Grubb *et al* reported that three out of six patients with lumbar involvement were misdiagnosed with lumbar disc disease.[4] Second, the pain in malignancies is expected to be continuous and typically worse at night. Conversely, in more than 80% of cases, the pain was found to be intermittent, strain related, and not aggravated at night, as in benign musculoskeletal conditions.[6] In our case the pain was intermittent at the beginning, continuous and severe later in the course. Patients with severe pain may need opioids.[7] The intermittent course of Ewing's sarcoma *versus* the expectation of a steady progression is a major factor for late diagnosis.

In the retrospective study of Widhe *et al* at the first medical visit, a bone tumor was suspected in only 19% of the patients with Ewing's sarcoma.[6] Tendinitis and sciatica were found to be the most frequent misdiagnoses. Although a radiograph at the first medical visit is believed to reduce the delay in diagnosis, misinterpretation of that radiograph as normal may lead to further delay. In the same study 43% of the first radiographs were misinterpreted as normal by radiologists.

The primary medical errors, which caused late diagnosis in our case, were misinterpretation of the first MRI, and poor medical follow-up after biopsy was taken. It is obvious that Ewing's sarcoma is not easily diagnosed in most cases. A detailed patient history and a careful physical examination are essential to minimize the delay in diagnosis. Also, an atypical clinical course in a benign musculoskeletal condition should warn us for a possible underlying malignant disease. Signs of spinal cord compression may be initial indicators for Ewing's sarcoma[8–10] and trauma may be found in the patient's history in one quarter of the cases.[6]

## Acknowledgements

We thank Dr H Coskun and Ms Mary C Ellis for their support for review of the manuscript and for insightful comments.

## References

1 Ferguson WS. Chronic leg pain in an adolescent male. *Med Health R I* 1999; **82:** 407–409.
2 Pizzo PA *et al*. Solid tumors of childhood. In: DeVita Jr VT, Hellman S, Rosenberg SA (eds). *Cancer Principles and Practice of Oncology*. Philadelphia: Lippincott, 1993, pp 1738–1791.
3 Mori Y *et al*. Disappearance of Ewing's sarcoma following bacterial infection: a case report. *Anticancer Res* 1997; **17:** 1391–1397.
4 Grubb MR, Currier BL, Pritchard DJ, Ebersold MJ. Primary Ewing's sarcoma of the spine. *Spine* 1994; **19:** 309–313.
5 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:** 1809–1813.
6 Widhe B, Widhe T. Initial symptoms and clinical features in osteosarcoma and Ewing sarcoma. *J Bone Joint Surg Am* 2000; **82:** 667–674.
7 Bouffet E *et al*. Spinal cord compression by secondary epi- and intradural metastases in childhood. *Child's Nerv Syst* 1997; **13:** 383–387.
8 Paul FA. Ewing's sarcoma as an etiology for persistent back pain in a 17-year-old girl after trauma to the back. *J Am Osteopath Assoc* 1995; **95:** 58–61.
9 Chi'en LT *et al*. Metastatic epidural tumors in children. *Med Pediatr Oncol* 1982; **10:** 455–462.
10 Sharma BS, Khosla VK, Banerjee AK. Primary spinal epidural Ewing's sarcoma. *Clin Neurol Neurosurg* 1986; **88:** 299–302.