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The Spine Journal 8 (2008) 1024–1029

THE SPINE JOURNAL

Sacral intraspinal extradural primitive neuroectodermal tumor

Volker Musahl, MD, Jeffrey A. Rihn, MD, Frank E. Fumich, MD, James D. Kang, MD*

Department of Orthopaedic Surgery, Division of Spinal Surgery, University of Pittsburgh Medical Center, Liliane Kaufmann Building, 3471 Fifth Avenue, Suite 1010, Pittsburgh, Pennsylvania 15213, USA

Received 21 October 2006; accepted 8 April 2007

Abstract

BACKGROUND CONTEXT: Intraspinal primitive neuroectodermal tumors (PNETs) are an exceedingly rare entity. A recent literature research revealed 28 cases reported. Only a few tumors in the literature were extradural in location, in the cervical and thoracic spine. The average survival after combination treatment including chemotherapy, radiation, and surgical resection is 20 months for the cases reported in the literature.

PURPOSE: We report a case of a patient with sciatica and cauda equine–like symptoms.

STUDY DESIGN: Case report.

METHODS: Urgent sacral decompression and resection of the tumor was performed with rapid pain relief for the patient.

RESULTS: Histology revealed a sacral extradural small blue-cell tumor, consistent with ES/PNET family tumors. An oncological workup revealed that the tumor presentation was metastatic with pulmonary and abdominal nodules. The patient underwent combination chemotherapy with vincristine, doxorubicin, and cyclophosphamide with mesna for 4 months.

CONCLUSIONS: The patient was without disease after excision, two courses of 4-month chemotherapy, and one course of 5-week radiation to the sacrum at 2 years.

Keywords: Primitive neuroectodermal tumors; Extradural; Sacral; Tumor

Introduction

Intraspinal primitive neuroectodermal tumors (PNETs) are an exceedingly rare entity. A recent literature research revealed less than 30 cases reported. Predilection of tumor location seemed to be the thoracic and lumbar spine [1]. Almost all reported cases of PNETs were found in an intradural location. Only three tumors were extradural in location, in the cervical and thoracic spine [2–4]. Unfortunately, many PNETs were found to be metastatic at initial presentation and therefore bear a devastating prognosis. Average survival after combination treatment including chemotherapy, radiation, and surgical resection is 20 months for the cases reported in the literature. We report a case of a sacral intraspinal, extradural PNET in a young patient that was metastatic at presentation.

Case report

JW is a 27-year-old gentleman who was referred from his primary care physician. He complained of low back pain that suddenly occurred upon awakening 1 month earlier. He described the pain as sharp, severe (9/10), continuous, and only minimally relived by pain medications. He also complained of pain radiating into the right buttock, the posterior aspect of the right thigh, and the posterior leg. In addition, he experienced weakness in his right calf and a numb sensation over his right heel. Upon further asking, the patient admitted to new onset of urinary incontinence and sexual dysfunction. He denied recent or past trauma to the low back or lower extremities. A review of systems revealed no history of headaches, dizziness, nausea, vomiting, fever, chills, night sweats, and no change in his weight. A review of the cardiovascular, respiratory, gastrointestinal, endocrine, integument, and psychiatric systems was also negative. Oral steroids and physical therapy prescribed by his family physician did not provide sufficient relief of his symptoms. The patient used 60 mg of oxycontin per day, which was prescribed by his primary care physician for pain control. The patient's past medical

FDA device/drug status: not applicable.

Nothing of value received from a commercial entity related to this manuscript.

* Corresponding author. Department of Orthopaedic Surgery, Liliane Kaufmann Building, 3471 Fifth Avenue, Suite 1010, Pittsburgh, PA 15213. Tel.: (412) 605-3241; fax: (412) 687-3724.

E-mail address: kangjd@upmc.edu (J.D. Kang)

1529-9430/08/$ – see front matter
doi:10.1016/j.spinee.2007.04.001

history was not significant for any medical problems. His social history was not significant for alcohol or tobacco use. The patient was unable to work as a telecommunications worker for the past 2 weeks because of his leg pain and weakness.

On physical examination, the patient was in no acute distress and was alert and oriented. The patient had a normal-based and well-balanced gait, which was coordinated but slow. He had positive heel walk, but no toe walking was possible on the right side. Strength testing of the lower-extremity muscle groups revealed 5/5 strength for bilateral iliopsoas, quadriceps, tibialis anterior, extensor hallucis longus, hamstrings, peroneals, and the left gastrocnemius-soleus complex. The right gastrocnemius-soleus complex was graded slightly weaker, 4/5. The straight leg raise was positive at 30° of flexion on the right and negative on the left. The Achilles' tendon reflex was absent on the right side and normal on the left. There was a well-described area of numbness to light touch on the right heel. A rectal examination was performed in the office and showed a normal sphincter tone and intact perineal sensation. The patient had a positive bulbocavernosus reflex and absent clonus, Hoffmann's, or Babinski reflex.

Magnetic resonance imaging (MRI) obtained by his primary care physician was available for review (Fig. 1). On both the sagittal and axial images, a mass within the spinal canal at the S1–S2 level was detected. The mass had low-signal intensity on T1-weighted MRI and moderate signal intensity on T2-weighted MRI. There was a small area of heterogenicity seen on both the T1- and T2-weighted MRI. The mass was seen in the S1–S2 area and emanated from the S1 nerve root on the right side and crossing the midline halfway to the left. This mass appeared to be causing extradural compression of the thecal sac in the sacrum pushing the remaining elements to the contralateral left side. However, there was no surrounding edema or soft-tissue destruction, and the mass appeared to be well marginated on MRI.

Hospital course

The patient was admitted to the hospital for further workup, preoperative planning, and intravenous corticosteroid therapy. He was started on 8 mg of decadron every 6 hours. After 48 hours in the hospital, the patient explained that were was significant improvement of his pain. To further delineate the intradural extraspinal mass and evaluate possible destruction of surrounding bony structures, a computed tomography (CT) myelogram was obtained. As previously seen on MRI, the mass was right sided and extradural and did not show bony destruction. The obtained laboratory studies returned within normal limits and included an erythrocyte sedimentation rate, C-reactive protein, and complete blood count with differential.

The preliminarily impression was that the lesion in question was a benign peripheral nerve sheath tumor such as a schwannoma that compressed the sacral nerve roots and caused the right leg pain, weakness, urinary, and sexual dysfunction. The differential diagnosis also included massive disc herniation, neurofibroma, infection, and hemangioma. Rare tumors in the young adult population such as lymphoma, Ewing's sarcoma, plasmocytoma, and even metastasis were included in the differential as well. The most likely diagnosis for this common presentation of sciatica in a young patient like JW with a documented intraspinal extradural mass was schwannoma. However, alerted by the

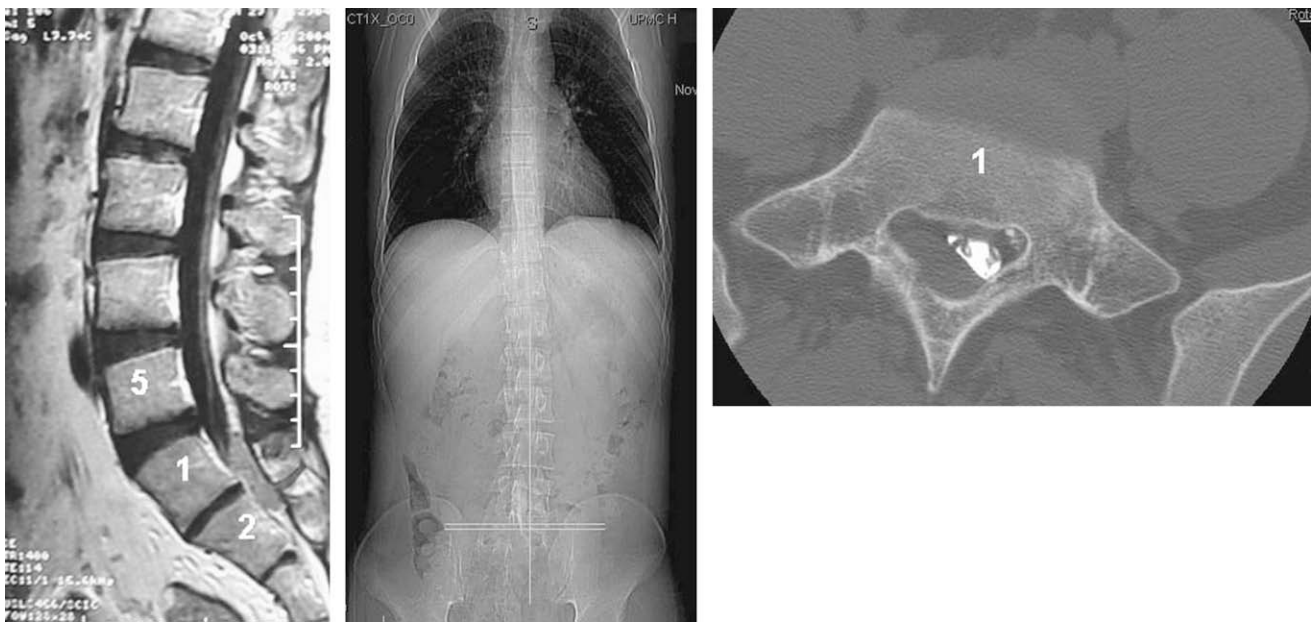

Fig. 1. (Left) T2 sagittal MRI of the lumbosacral spine. Low-signal intensity intraspinal extradural lesion posterior to S1/2 disc. (Middle) Scout and (Right) axial CT myelogram. Intraspinal extradural lesion at S1 (low-signal intensity) and spinal cord (high-signal intensity).