

Unilateral Thigh Pain in a 32-Year-Old Male

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INTRODUCTION AND BACKGROUND

Leg pain that is non-traumatic in origin is an uncommon presentation in a 32-year-old male. In this age group, the differential diagnosis should include tumors of the bone and soft tissue. While roentgenograms are the primary diagnostic imaging tool for bone lesions due to their ability to show calcification, ossification, cortical destruction, and periosteal reactions, they are inadequate for imaging soft tissue. Computerized tomography and magnetic resonance imaging can improve visualization of soft tissues, bone marrow, and sites with complex skeletal anatomy.¹

CASE PRESENTATION

A 32-year-old male presented to the emergency department with a history of left upper anterior thigh pain and weakness. The pain was intermittent for two years, but lately had progressively worsened. He had sustained a fifty-five pound weight loss over two years. He denied any fever, chills, night sweats, or problems urinating. He did have abdominal pain without nausea, vomiting, or diarrhea.

He had previously been diagnosed with muscular pain. Prior to the current presentation he was evaluated by Orthopedic Surgery, including an MRI that showed a solitary central intramedullary bone lesion in the distal diaphysis of the left femur, measuring 2 x 5.5 x 1.5 cm and consistent with a slow-growing benign lesion such as an enchondroma. A CT scan with contrast and MR imaging of the abdomen demonstrated a retroperitoneal heterogeneous mass with solid and cystic components suggestive of a malignant peripheral nerve sheath tumor (Figures 1a, 1b, & 1c).

The patient's vital signs were stable with mild hypertension, and his physical exam was positive for abdominal pain on the left side. There was a large, palpable, non-pulsatile mass in his left abdomen. His only other positive physical exam finding was decreased strength in hip flexion bilaterally, more pronounced on the left. His bowel sounds were normal, his abdomen was not distended, and he did not exhibit guarding, rebound tenderness or flank tenderness. His labs on admission included a CBC, a CMP, coagulation studies, and blood and urine cultures. All were within normal limits except an elevated alkaline phosphatase of 132.

The patient was admitted to the hospital on the Urology service for pain control and further work-up. A three-phase CT scan of

the kidneys with 3-D reconstruction confirmed the suspicion that the mass did not arise from the left kidney. A MRI of the abdomen showed that the 13 x 15 cm retroperitoneal mass appeared to have replaced the entire left psoas muscle and extended into the L2-L3 neural foramen, and demonstrated no liver metastases or retroperitoneal lymphadenopathy (Figures 1b, 1c, 2a & 2b). An orthopedic surgeon was consulted to assist with an exploratory laparotomy. The retroperitoneal mass was excised along with the left psoas muscle and a portion of the paraspinal muscles. Frozen sections continued to show tumor at the margin of resection despite aggressive dissection into the foramen.

The tumor weighed 1250g and measured 18 x 14 x 10 cm. Approximately 10 percent of the tumor was necrotic, and no lymphatic or vascular invasion was identified. Immunohistochemical staining showed positive staining for S-100, Vimentin, and CD34. The tumor was a poorly differentiated stage III malignant peripheral nerve sheath tumor that was T2bNOM0.

The patient's post-operative course was uncomplicated and he was discharged on the fifth day after surgery. Repeat MR imaging of his lumbar spine showed residual solid enhancing tumor in the left paravertebral space, lateral extra-foraminal compartment and left neural foramina in the L1-L2 and L2-L3 segments without involvement of the spinal canal. The patient is being treated with adjuvant radiotherapy to the L1-L3 paravertebral area and has undergone evaluation for adjuvant chemotherapy.

DISCUSSION

Most benign primary bone tumors involve younger people in their second and third decades of life and are painless tumors. These tumors are generally present for a while prior to incidental discovery, such as a pathologic fracture. Leg pain is often the direct result of a tumor; however, it may also be referred. Enchondromas are benign cartilaginous neoplasms, usually solitary and located in intramedullary bone. In the United States, enchondromas account for 12-14% of benign bone neoplasms, and most patients are asymptomatic.² Malignant transformation is generally the cause if the presentation involves pain.

Sarcomas are malignant tumors arising from connective tissue at any body site and account for 1% of adult solid tumors; approximately 13% arise in the retroperitoneum.³ Approximately 9530 cases are diagnosed annually in the United States, representing only 0.70% of new cancers.⁴ Immunohistochemistry is used to

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identify the sarcoma subtype. Vimentin is found in almost all sarcomas, and the presence of S-100 antigen suggests neural sheath cells.

Malignant peripheral nerve sheath tumors (MPNSTs) make up 5 to 10% of sarcomas and originate from peripheral nerves or from cells associated with the nerve sheath, such as Schwann cells, perineural cells, or fibroblasts.⁵ MPNSTs generally occur in adulthood, typically between the ages of 20 and 50 years of age, and usually present as an enlarging palpable mass. Most MPNSTs occur in conjunction with large peripheral nerves and may result in a variety of clinical patterns, including radicular pain, paresthesias and motor weakness.

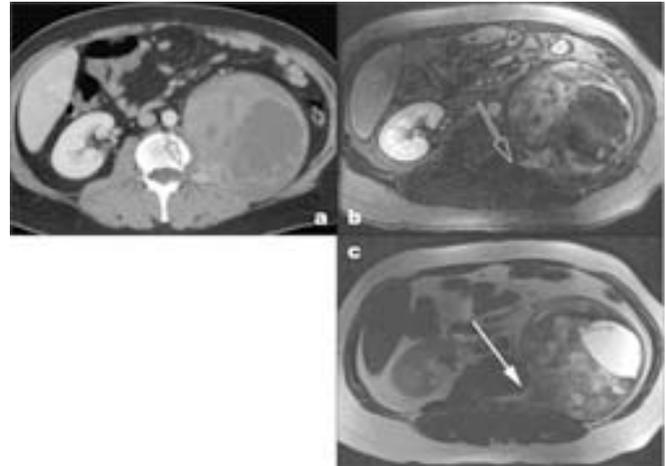
Staging is dependent upon histologic grade, tumor size, tumor depth, and the presence or absence of metastases. Histologic grade, tumor size, and tumor depth are the strongest predictors of eventual metastases in the absence of detectable metastases. The local and distant recurrence rates for MPNSTs are reported to range from 40-70%. Five-year survival has been reported to range from 16-52%.⁶ Complete surgical excision, small tumor size (<5 cm), and low-grade disease correlate with longer survival.⁵

This patient's prognosis is poor due to the delay in seeking medical attention, the tumor's stage and the tumor's retroperitoneal location. Using this patient's criteria and a post-operative nomogram for 12-year sarcoma-specific death based on data from 2163 patients, his probability from dying within twelve years from his sarcoma is around 88%.⁷ His prognosis might have been improved if clinical suspicion was initially higher and/or more effective radiological imaging was utilized earlier with initial presentation.

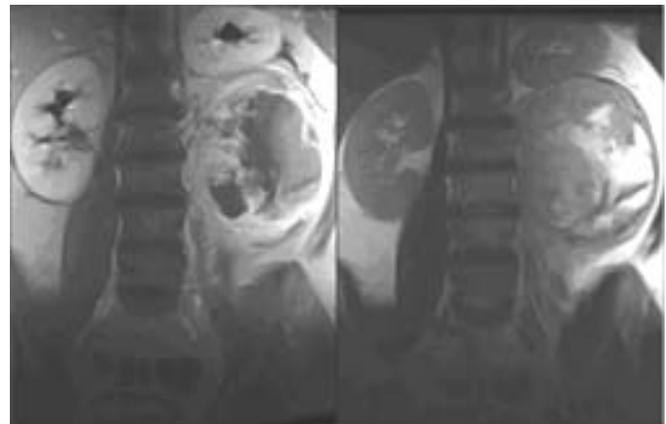
REFERENCES

1. Novelline RA: *Squire's Fundamentals of Radiology*. Harvard University Press, Cambridge, MA, 2006, p.365.
2. Chew FS, et al. *Enchondroma and Enchondromatosis*. *Emedicine*. <http://www.emedicine.com/RADIO/topic247.htm>
3. Lawrence, W Jr, Donegan, WL, Natarajan, N, et al. *Adult Soft Tissue Sarcomas: A Pattern of Care Survey of the American College of Surgeons*. *Ann Surg* 1987; 205:349.
4. Jemal, A, Siegel, R, Ward, E, et al. *Cancer Statistics, 2006*. *CA Cancer J Clin* 2006; 56:106.
5. Geller, DS, Gebhardt, MC. *Malignant Peripheral Nerve Sheath Tumors (MPNSTs)*. <http://www.liddyshiversarcomainitiative.org/Newsletters/V03N03/mpnst/mpnst.htm>
6. Coindre, JM, Terrier, P, Guillou, L, et al: *Predictive Value of Grade for Metastasis Development in the Main Histological types of Adult Soft Tissue Sarcomas*. *Cancer* 2001; 91:1914.

7. Kattan, MW, Leung, DHY, Brennan, MF. *Postoperative Nomogram for 12-year Sarcoma-specific Death*. *J Clin Onc* 2002; 20:791.



Figures 1a, 1b, 1c: Axial contrast-enhanced CT, axial T1 GRE with gadolinium, and axial T2 FSE images of the abdomen showing a retroperitoneal heterogeneous mass with solid and cystic components. All images show the involvement of the L2-L3 left neural foramen (arrows). The heterogeneity and neural foramen involvement are suggestive of a malignant peripheral nerve sheath tumor.



Figures 2a & 2b: Coronal T1 FSE post-gadolinium and T2 FSE images showing the cephalo-caudad view of the mass and demonstrating the cephalad displacement of the left kidney.

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