A Painful Tibial Mass in a 37-year-old Man

Seena C. Aisner MD, Marcia Blacksin MD,
Francis Patterson MD, Meera R. Hameed MD

© The Association of Bone and Joint Surgeons 2008

History and Physical Examination

A 37-year-old man presented with right leg pain of 4 months' duration. He was athletic and jogged several miles daily. He initially was evaluated and diagnosed with a stress fracture and was told to avoid weightbearing and exercise, but his symptoms did not improve. He had little symptomatic relief with the use of nonsteroidal antiinflammatory medications. Except for a history of ulcerative colitis in remission, his medical history was unremarkable.

Physical examination revealed a healthy and athletic appearing man with a 2-cm hard and immobile mass on the anterior aspect of his right upper tibial shaft. The lesion was extremely tender to palpation particularly in the center.

Each author certifies that he or she has no commercial associations (eg, consultancies, stock ownership, equity interest, patent/licensing arrangements, etc) that might pose a conflict of interest in connection with the submitted article.

Each author certifies that his or her institution has approved the reporting of this case report, that all investigations were conducted in conformity with ethical principles of research, and that informed consent for participation in the study was obtained.

S. C. Aisner (✉), M. R. Hameed
Department of Pathology, University of Medicine and Dentistry
of New Jersey, New Jersey Medical School, University Hospital,
150 Bergen Street, E155, Newark, NJ 07103, USA
e-mail: aisnersc@umdnj.edu

M. Blacksin
Department of Radiology, University of Medicine and Dentistry
of New Jersey, New Jersey Medical School, Newark, NJ, USA

M. Blacksin, F. Patterson
Department of Orthopedic Surgery, University of Medicine and
Dentistry of New Jersey, New Jersey Medical School, Newark,
NJ, USA

The patient, however, showed a full range of motion of his right leg. There were no skin changes. The remainder of the physical examination was unremarkable. He had no fever and his laboratory studies, including white blood count and differential, were normal.

Radiographs, computed axial tomography (CT) images, and magnetic resonance imaging (MRI) scans of the right tibia were obtained (Fig. 1).

Based on the history, physical examination, and imaging studies, what is the differential diagnosis?

Imaging Interpretation

Radiographs revealed a subperiosteal or intracortical lytic lesion in the anterior cortex of the tibia at midshaft (Fig. 1A). Axial CT images showed the lesion was located on the surface of the anterior tibial cortex (Fig. 1B). A faintly calcified central nidus and surrounding periosteal new bone were noted. MRI revealed a well-circumscribed, oval mass on the surface of the tibia with no adjacent bone marrow edema (Fig. 1C–D). The lesion was slightly higher in signal intensity than muscle on T1-weighted images and centrally isointense in signal to suppressed fat on T2-weighted images. A bone scan also was performed and revealed discrete increased tracer uptake in the lesion.

Differential Diagnosis

Osteoid osteoma
Intracortical hemangioma
Leiomyoma
Fibrous dysplasia
Adamantinoma
Intracortical osteosarcoma

Fig. 1A–D (A) A sagittal radiograph shows a lytic lesion (arrowhead) in or on the anterior cortex of the tibia. (B) An axial CT image shows a well-circumscribed lytic lesion (arrowhead) on the surface of the tibia and a faintly calcified nidus. Periosteal reaction on either side of lesion can be seen (arrow). (C) A sagittal T1-weighted MR image shows an oval neoplasm slightly higher in signal intensity than muscle with a thin low signal capsule (arrowhead). Adjacent periostitis can be seen (arrow). (D) An axial fat-suppressed T2-weighted MR image shows a mild increase in signal intensity at the periphery of the lesion with signal intensity in the center of the lesion isointense to suppressed fat (arrowhead).

The lesion was exposed and observed completely at surgery. Examination of the cortex revealed an expansile mass lesion. An intraoperative frozen section was performed, which revealed a spindle cell neoplasm. The specimen consisted of a nodular well-circumscribed firm,

tan, rubbery mass and histologic studies were performed (Fig. 2).

Based on the history, physical findings, radiographic studies, and the histologic picture, what is the diagnosis and how should this lesion be treated?

Fig. 2A–B (A) This photomicrograph shows neoplastic spindle cells interspersed with moderate amounts of stroma (Stain, hematoxylin and eosin; original magnification, $\times 400$). (B) Immunohistochemical staining using smooth muscle actin shows strong cytoplasmic positivity in the tumor cells (Original magnification $\times 400$).

Histology Interpretation

Gross evaluation of the specimen revealed a 3.5-cm segment of resected cortical bone. After decalcification, the bone was bisected and revealed a well-defined, rubbery, tan-pink nodule measuring $0.7 \times 0.6 \times 0.4$ cm located on the surface of the cortex. There was a second smaller nodule adjacent to and attached to the main mass, measuring 0.4 cm.

Microscopically, the tumor was well defined with bony sclerosis seen rimming the entire neoplasm. The tumor was composed of moderately cellular spindle cells in a whorled pattern with abundant pink cytoplasm (Fig. 2A). Cellular atypia and mitoses were absent and necrosis was not seen. The neoplastic cells were strongly positive for smooth muscle actin (Fig. 2B), focally positive for desmin, and negative for the cytokeratin marker AE1/3, the neural marker S-100, and the vascular marker CD34 (Dako North America, Inc, Carpinteria, CA). A trichrome stain revealed pink-stained tumor cells, suggesting muscle differentiation.

Diagnosis

Leiomyoma

Discussion and Treatment

Primary leiomyomas of bone are composed of smooth muscle stroma histologically similar to leiomyomas seen in other locations and can be identified with the use of routine and immunohistochemical stains.

The histologic differential diagnosis of benign cortical spindle cell lesions is limited to several neoplasms including leiomyoma, fibrous histiocytoma, neural neoplasms, and nonossifying fibroma. While neural neoplasms, fibrous histiocytoma, and nonossifying fibroma may share some histologic similarities, they will not stain strongly for muscle markers (smooth muscle actin, desmin). We considered adamantinoma far less likely in the differential diagnosis, as histologically this neoplasm is noted for its heterogeneity and presence of epithelial islands. Osteosarcoma was not considered in the histologic differential diagnosis, as the tumor histologically displayed no osteoid or suggestions of malignancy. The use of immunohistochemical staining was helpful in this case in that the tumor cells stained strongly for muscle markers, including smooth muscle actin and desmin, thus confirming this tumor's smooth muscle origin. The neoplastic cells did not stain for neural, fibrous, or epithelial markers (S-100, CD34, and AE1/3, respectively). A malignant spindle cell tumor was not considered because of the tumor's lack of

pleomorphism, mitoses, and necrosis. The sclerotic margins seen on the conventional radiograph and in the histologic sections also supported the benign nature of this tumor.

Primary leiomyomas involving bone are exceedingly rare neoplasms, with fewer than 20 cases reported in the literature. The majority of the reported cases involved gnathic sites, predominantly the mandible [4]. Two cases occurred in the maxillary tooth socket and one in the temporal bone [2]. Appendicular involvement is extremely rare. There have been four reported cases occurring in the ulna [12], tibia [9, 10], and femoral neck [1]. In one case report involving the ulna [12], the radiographic features simulated an osteoid osteoma and the lesion was located in the cortex, similar to our case. That patient also presented with pain. The periosteum was the site of origin in the first two reported cases of leiomyoma of the tibia by Taxy et al. [9] and Mirra [5]. The lesion of the femoral neck was cystic and presented as a manifestation of disseminated leiomyomatosis [1]. An angioleiomyoma of the tibia has been reported by Tomoda and Iyama [10], which presented as an expansile growing mass.

The radiographic differential diagnosis includes a variety of benign primary bone tumors and processes. We believed the most likely differential diagnosis was a subperiosteal osteoid osteoma. This tumor is less common than the cortical variety and may produce less bony sclerosis. Faintly calcified nidus and periosteal new bone are seen with this lesion. This lesion can cause reactive bone marrow edema, producing a focus of increased uptake on bone scan and edema on MRI. Thin-section CT is the study of choice to identify a nidus, as this can sometimes be missed on MRI. Surface hemangiomas can be associated with cortical thickening, cortical erosion, and a lytic focus resembling a nidus on CT. A soft tissue mass extending from the cortex may be seen but is an infrequent finding [5, 7]. Radiographically, adamantinoma was also considered because of the tibial location of the lesion but was believed unlikely for several reasons. MRI clearly demonstrated this was a surface lesion and its epicenter was not intracortical, as seen with adamantinoma. The cortex can be eroded with adamantinoma, but adamantinoma is usually a larger neoplasm than the one visualized here. Adamantinoma begins in and expands the cortex. Intracortical osteosarcoma is the rarest of osteosarcomas, with approximately 18 reported cases in the literature. These neoplasms are seen in patients 9 to 43 years of age and are frequently found in the tibia or femur. These neoplasms can rarely demonstrate matrix calcifications on CT. On MRI, the central cortical components of these lesions are isointense to muscle on T1-weighted images and heterogeneously high in signal on T2-weighted images. Peritumoral bone marrow edema is occasionally seen as well. The lesion in our case did not share these MR characteristics [11].

Surface and intracortical fibrous cortical defects and fibrous dysplasia of the cortex have been reported, and signal characteristics do not necessarily follow that of fibrous tissue. The radiographic findings in our case also resembled those of subperiosteal ganglion and juxtacortical aneurysmal bone cyst [8]. However, the lesion was solid in nature on the CT and MRI, and these diagnoses were not strongly considered.

This patient had no known primary neoplasm, and metastasis is not a strong consideration in this age group. However, surface metastasis can occur with a wide variety of primary carcinomas, including lung, breast, kidney, and pancreas. The thin capsule seen around the lesion, as well as its oval shape and sharply marginated edge, made metastasis less likely [8].

There are no reports documenting the radiographic or imaging appearance of periosteal leiomyoma. The case report by Taxy et al. [9] showed normal plain radiographs and a positive bone scan. However, a case report on subperiosteal leiomyosarcoma demonstrated a well-defined lytic lesion with reactive sclerosis and solid periosteal reaction in the cortex of a tibia [6]. This lesion had no matrix calcification. Radiographically, these benign and malignant lesions may appear similar. CT of the malignant variant demonstrated a larger and more irregular area of osteolysis than the benign counterpart [6].

Following an en bloc tumor resection and partial tibial corticectomy, the patient noted a substantial decrease in pain, which was completely gone by 3 months. Twenty-four months after the surgery, the patient remains well and has no evidence of recurrence. Subperiosteal leiomyoma is a rare benign spindle cell

neoplasm, in which marginal excision appears adequate to prevent recurrences [3].

References

1. Braun W, Kotter A, Wiedemann M, Wagner T. Intraosseous leiomyoma of the neck of the femur: a case report. *Int Orthop*. 1994;18:47–49.
2. Jeung CU, Song SH, Kim SH, Song KS. Primary leiomyoma of the temporal bone. *J Korean Neurosurg Soc*. 2004;35:321–323.
3. Liang H, Frederiksen NL, Binnie WH, Cheng YS. Intraosseous oral leiomyoma: systematic review and report of one case. *Dentomaxillofac Radiol*. 2003;32:285–290.
4. Loyola AM, Araujo NS, Zanetta-Barbosa D, Mendes VC, Jordao-Silva C, Bittar TO. Intraosseous leiomyoma of the mandible. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod*. 1999;87:78–82.
5. Mirra JM. *Bone Tumors: Clinical, Radiologic and Pathologic Correlations*. Philadelphia, PA: Lea and Febiger; 1989:871–875.
6. Narváez JA, De Lama E, Portabella F, Ortega R, Condom E. Subperiosteal leiomyosarcoma of the tibia. *Skeletal Radiol*. 2005;34:42–46.
7. Seeff J, Blacksin MF, Lyons M, Benevenia J. A case report of intracortical hemangioma. *Clin Orthop Relat Res*. 1994;302:235–238.
8. Seeger LL, Yao L, Eckardt JJ. Surface lesions of bone. *Radiology*. 1998;206:17–33.
9. Taxy JD, Conklin J, Mann JJ, Brooker A. Case report 147: leiomyoma of the periosteum of the tibia. *Skeletal Radiol*. 1981;6:153–154.
10. Tomoda K, Iyama K. A case of intraosseous angioleiomyoma. *Acta Orthop Scand*. 1992;63:568–570.
11. Vanhoenacker F, De Beuckeleer LH, De Schepper AM. Intracortical osteosarcoma: is MRI useful? *Rofa*. 2001;173:959–960.
12. Zikria B, Radevic MR, Jormark SC, Huvos AG, Yang SS. Intraosseous leiomyoma of the ulna: a case report. *J Bone Joint Surg Am*. 2004;86:2522–2525.