Adamantinoma

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Case report

A 15-year-old athletic female presented with a several-week history of right mid-tibial pain. The pain was aggravated during a volleyball game. She did not recollect any direct trauma to the area. The anterior mid-tibial crest pain was dull, nonradiating and worsened with activity and weight-bearing. The pain was no worse at night. There was no recent history of illness, fever, or weight loss.

Examination of the lower extremities revealed symmetrical muscular development with moderate point tenderness to direct palpation of the right mid-tibia. No swelling, discoloration, or distortion of the limb was appreciated. There was no adenopathy. She had a normal gait and full, active range of motion of the knee and ankle.

Presenting radiographs (Fig. 1) revealed a focal oval radiolucency in the right anterior mid-tibial cortex measuring 8 mm in the longest dimension. Several linear radiolucencies radiated from this lesion. No periosteal reaction or soft tissue mass was associated with this lesion.

An MR examination revealed a focal area of abnormal signal intensity in the anterior cortex of the mid-tibia. The signal intensity was slightly hypointense to bone marrow on T1-weighted axial and sagittal images (Figs. 2A, 3) and hyperintense to bone marrow on T2-weighted images (Fig. 2B). Following the administration of Gd-DTPA intravenously, there was no significant enhancement of the lesion.

These clinical and radiographic assessments led to the differential diagnosis of a bone (Brodie's) abscess, with less likely possibilities of osteoid osteoma or stress fracture. Surgical biopsy was deferred at this time because of the mildness of the symptoms, the benign appearance of radiographic studies, and the reluctance of the patient to interrupt her demanding athletic schedule.

She was followed at regular intervals for the next 9 months, during which time she was involved in school basketball, track, and volleyball. Plain radiographs were evaluated at each visit and revealed minimal interval change. Low-level pain persisted during these 9 months, and the end of the school year afforded the opportunity to perform an open biopsy.

The biopsy comprised reddish, gritty material. Histologically, a small island of epithelial cells was present within a fibrovascular matrix (Fig. 4). Immunohistochemical stains for cytokeratins (AE1/3) were strongly positive while stains to evaluate possible neuroectodermal origin (S100 and chromogranin) were negative. The histologic features were those of an adamantinoma.

A follow-up CT scan revealed a second small focus of radiolucency 45 mm distal to the original lesion (Fig. 5).

Treatment included a wide resection of the biopsy tract along with the anterior cortex and medullary contents of the tibia to include the second lesion. Reconstruction was accomplished with the introduction of an unreamed intramedullary nail and placement of both autogenous iliac crest and cancellous allograft bone graft into the defect. Pathologic
Adamantinoma is a rare bone lesion. Clinically, the most common presenting symptom is swelling, with or without pain [1]. It is considered to be a low-grade malignancy by most authors. One report [2] has predicted a mortality of 18% in a large, multicenter review of 200 cases, although several patients died without evidence of disease. Another study reported a 31% local recurrence rate [3].

Between 80% and 90% of adamantinomas occur in the tibia, typically in the anterior mid-diaphysis [1–4]. The usual radiographic appearance of the adamantinoma is a large (3–16 cm), bubbly and multiloculated lesion with sharp, well-defined margins. One study noted that 80% of all lesions were more than 5 cm in length [3]. Multifocal lesions within the same bone are histologically contiguous satellite lesions in most instances. In one large study, only 2 of 85 cases demonstrated distinctly separate lesions in the same bone, yet synchronously presenting lesions in the tibia and fibula are common (13%) [3].

Most tumors involve both the cortical and medullary portions of the bone. However, approximately 10% of the lesions involve either the cortex only or are predominantly cortical with minimal medullary involvement [3]. There are at least two reports of adamantinoma arising in the pre-tibial soft tissues [5, 6].

The histogenesis of the tumor has been controversial, but recent immunohistochemical and electron microscopic analysis has confirmed an epithelial ancestry [7, 8]. One commonly held theory espoused by Fischer [9] contends that the tumor arises from an area of epithelium trapped in bone or periosteum in fetal development. Juxtacortical adamantinomas have been described by several authors [4, 10, 11]. Jaffe [12] concluded that adamantinoma starts its development in relation to the outer surface of the cortex and not in the medullary cavity of the bone. The radiographic and histologic findings in our case support a cortical or subperiosteal origin for this tumor.

Controversy developed over the proper treatment of adamantinoma due to the high local recurrence rate with incomplete curettage and an underestimation of the aggressiveness of this tumor. This prompted several authors to advocate ablative surgery with amputation [13–15]. More re-