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Case Report

Adamantinoma filling the medullary space of the tibia: A case report

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ABSTRACT

Adamantinoma of the bone is a rare low-grade bony tumor that accounts for less than 1% of all primary bone tumors. On imaging, adamantinoma may be similar to other tumors such as osteofibrous dysplasia, for which the treatment protocol is completely different. Therefore, correct diagnosis and staging of adamantinoma ensures that the patient will undergo appropriate surgery. We present a case of atypical adamantinoma to highlight the fact that adamantinoma should be considered in the differential diagnosis of tibial tumors.

Introduction

Adamantinoma of long bone is a miscellaneous tumor, according to WHO Classification of Bone Tumors in 2013. It is a rare low-grade primary neoplasm of the bone that originates from epithelial cells arranged in islands of fibers and proliferating fibroblastic medullary tissue. Adamantinoma often manifests as local recurrences and metastasis to the lung [1]. This tumor has not previously been reported as an isolated lesion filling the medullary space of the tibia. Therefore, we present a patient with adamantinoma, emphasizing its imaging characteristics.

Case presentation

A 67-year old female patient presented with pain in the left leg caused by a fall. She developed difficulty in standing, accompanied by limitation of activity for 5 hours. The patient was generally in good health, without fever, chills, swelling or pain in her left leg. During history-taking, she mentioned that she had a several-month history of painless swelling in the upper segment of her lower left leg. This did not alter her activities of daily living. On physical examination, the upper end of the left tibia was markedly swollen, with tenderness and pain provoked by tapping. A subcutaneous bone rubbing

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sensation was palpable; the left knee joint movement was limited, the peripheral blood supply was normal, and no other significant abnormalities were found. Laboratory tests showed that calcium and phosphorus were in normal ranges and serum potassium was low (3.24 mmol/L).

A radiograph of the left tibia and fibula revealed an expansile osteolytic lesion with incomplete sclerotic rim and thinning of adjacent cortex, as well as a pathological fracture in the upper segment of left tibia (Fig. 1A). On computed tomography (CT), expansive bone destruction accompanied with a pathological fracture was detected in the upper segment of the left tibia, approximately 72 mm × 34 mm × 31 mm. The CT attenuation was about 52 HU. There was an incomplete sclerotic rim, and several cystic and honeycombing-like changes were found on multiplanar reformation images (Fig. 1B-E). Axial (Fig. 1F-G) and coronal (Fig. 1H) fat-saturated T2-weighted magnetic resonance images (MRI) of the left tibia revealed that the upper tibial shaft was infiltrated with a marrow neoplasm accompanied by edema of surrounding soft tissue. However, there was no soft tissue mass. Chest CT showed infiltration of both lungs, and upper abdominal CT showed multiple cysts of the liver. The basis for diagnosis of metastatic lesions was insufficient. Color Doppler ultrasound showed no obvious abnormalities in the common femoral vein, superficial femoral vein or deep femoral vein of the left lower extremity.

Surgical biopsy was performed. Histological findings (Fig. 1I and J) revealed many glandular tubular structures among fibrous tissues. Immunohistochemistry revealed positive staining for cytokeratin and vimentin, and negative staining for CK7, CK20, S-100, CD34, CD31, and FVIII. The pathological diagnosis was primary bone adamantinoma. The patient was given symptomatic treatment including subsidence of swelling and pain relief after hospitalization. After learning the pathological results, the patient's family members asked us to transfer her to another hospital for further treatment. Later, the patient underwent en bloc resection of the proximal segment of the left tibia and reconstruction in the other hospital, including excision of the tumor, femoral osteotomy and implantation of femoral and tibial prosthesis. The postoperative follow-up was good. A postoperative radiograph is shown in Fig. 1K.

Discussion

Adamantinoma is a rare low-grade primary neoplasm of the bone that accounts for less than 1% of all primary bone tumors. The lesion was first described by Fischer in 1913. Its histogenesis has been controversial since its first description; nevertheless, it is broadly accepted that adamantinoma is an intermediate malignant tumor of fibroblast origin.

Electron microscopy and immunohistochemical studies have confirmed the epithelial origin of the neoplasm [2]. Histologically, the tumor is composed of various types and proportions of epithelial cells in a background of fibrous or osteofibrous stroma. Four histological patterns have been described for classic adamantinoma: basaloid, spindle, tubular, and squamous, of which the basaloid and tubular patterns are the most common [2,3]. Because of its epithelial nature and histological variability, adamantinoma can resemble lesions ranging from benign to malignant, including epithelial metastasis. Dorfman and Czerniak divided adamantinoma into 2 forms. The first is the classical form, characterized by intraperiosteal radiographic destruction. On histology, this form is dominated by basophil and tubular tumor cells. The other is the differentiated form, characterized by the presence of small isolated epithelial cells and multifocal fibrous dysplasia of bone in the cortex [1]. Our case fits the description of the classical form. Key points for identification are described below.

Adamantinoma is typically a slowly-proliferating, locally-invasive tumor. There is a slight male predominance. The ages of patients with adamantinoma were older and less equally distributed over the second to fourth decades; nevertheless, the tumor can occur in patients from 3 to 86 years [4]. Pain is the most frequent symptom, followed by slow local swelling and pathologic fracture [4]. Because the disease is often diagnosed with a history of trauma, it is reasonable to propose that the tumor is caused by chronic injury and repair. Ryrie postulated that because of the tibia's superficial location and sharp anterior edge, injury leads to subperiosteal epithelial cell implantation with hematoma and subsequent ossification; ultimately, the aberrant repair leads to tumor formation [3]. The date of trauma in our case was too close to presentation to reason that trauma was a cause of tumor formation; nevertheless, it led us to incidentally diagnose the disease.

Adamantinoma is characterized by a slow clinical progression with lung metastases and local recurrences over time. Hence, complete and sufficient surgery is usually performed to avoid local recurrence or metastasis after diagnosis of solitary foci [1]. Radiation and chemotherapy have not proven useful for the treatment of adamantinoma. The current treatment of choice is en bloc resection with wide operative margins and reconstruction [4]. Allografts and fibular autografts have been most commonly used [3].

In our patient, there was expansive bone destruction, cortical and trabecular involvement, and replacement of normal bone marrow by tumor on MRI, supporting the diagnosis of pathological fracture rather than simple fracture, even with a history of trauma. It was difficult to determine whether the edema around the lesion was caused by trauma or tumor, a crucial factor for tumor staging. Careful wide en bloc resection of an adamantinoma is pivotal because marginal or intralesional removal significantly increases the risk of local recurrence (≤32%) and of lung metastases (≤25%) [5]. In this respect, the surgeon is often challenged by the multifocal appearance of the adamantinoma, possibly hampering the objective of optimal functional recovery. An optimal preoperative staging procedure is therefore essential. In our case, the staging of the lesion was Stage IB (G1T2M0) according to the Enneking system [6].

Imaging plays an important role in the diagnosis of bone tumors. The imaging findings of adamantinoma are characteristic. Radiography is the most reliable diagnostic imaging method [3]. Typical lesions are located in the anterior medial axis of the tibia. They are eccentric, dilated, cortical or intramedullary soluble lesions with multilocular features and sclerotic margins. Pathological fractures may occur. Anterior arch deformity of tibia is common. According to Desai et al.,

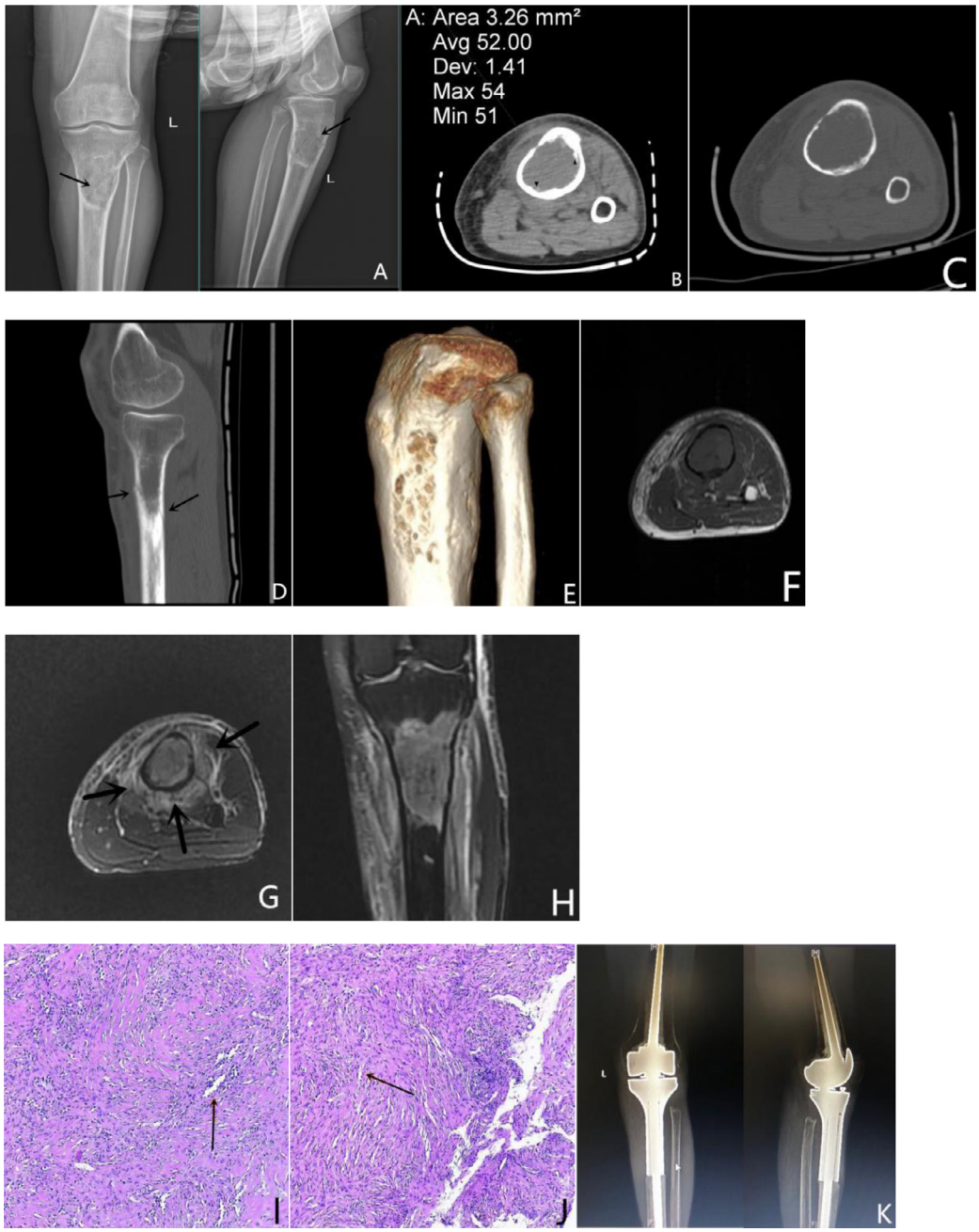

Fig. 1 – (A) Frontal radiograph of the left tibia and fibula: an expansile osteolytic lesion with incomplete sclerotic rim and thinning of adjacent cortex, as well as pathological fracture (arrow) in the upper segment of left tibia. Soft tissue window (B), bone window (C), MPR (D) and 3D reconstruction (E) of the left tibia and fibula: an expansive bone destruction accompanied with pathological fracture (arrow) was detected in the upper segment of the left tibia. The CT attenuation was about 52 HU. Incomplete sclerotic rim, and multiple cystic and honeycomb-like changes were found on multiplanar reformation images. The medial cortex showed ridge-like changes (triangle). Axial (F) T1-weighted, axial (G) and coronal (H) fat-saturated T2-weighted magnetic resonance images of the left tibia and fibula: a low T1, high T2 signal marrow infiltrating neoplasm involving the upper tibial shaft accompanied by edema(arrow) of surrounding soft tissue. (I, J) Histological specimen: there were many glandular tubular structures (arrow) among fibrous tissues under the microscope. (K) Radiograph for postoperative review.

location in the tibia with intracortical development are 2 diagnostic indicators for adamantinoma [1]. However, in a series of 22 patients analyzed by Vander Woude [7], 60% of the tumors had discontinuous or more obvious bone marrow extensions, while only 40% were located in cortical bone. In our case, the lesion filled the medullary cavity without eccentricity and the sclerotic margin was incomplete; this differed from the typical imaging findings.

CT and MRI are not specific for making the diagnosis [1]. They can be used as supplements because findings of radiography alone may underestimate the true tumor extent and components. CT is better for showing the edge and internal structure of the lesion including calcification or ossification. Thoracoabdominal CT is used to rule out metastases [4]. MRI, characterized by high soft tissue resolution and multiplanar imaging, has become the primary technique for staging musculoskeletal tumors. It is pivotal for precise locoregional staging, especially for depiction of distant cortical foci, soft tissue, and intramedullary extension. Therefore, MRI is most useful for determining tumor-free margins and developing strategies for reconstructive surgery. Sagittal images of the entire lower leg are usually optimal to show the exact cranial–caudal tumor extension and to identify small distant foci within the cortical bone or bone marrow compartment that can be missed on radiographs. With respect to reconstructive surgery, axial images, either T1- or T2-weighted, are particularly useful to assess the status of the posterior cortical bone. Vander Woude [7] summarized 2 morphological patterns of tumors on MRI: a solitary lobulated focus versus a pattern of multiple small nodules in one or more foci. MRI will show low T1, high T2 signal-intensity lesions that extend into the soft tissue. The lesion shows either homogenous or peripheral enhancement after gadolinium administration [7]. Unfortunately, our patient's imaging was restricted to native T1- and T2-weighted images without static or dynamic contrast enhancement.

When lesions are confined to the cortex, it is necessary to differentiate them from osteofibrous dysplasia (OFD), because of the common origin shared between both. In fact, a well-differentiated adamantinoma may develop from OFD of bone over time [2]. OFD and OFD-like adamantinoma are confined to the cortex, whereas soft tissue and intramedullary extension is usually observed in classical adamantinoma. Occasionally, adamantinoma displays erosion-like edges and sclerotic margins are not obvious [8]. However, when a lesion is completely intracortical, imaging features do not permit distinction of classic adamantinoma from differentiated adamantinoma or OFD [3]. In this context, age is good differentiator. Classical adamantinomas occur more often in patients more than 20 years of age, and fibrous dysplasia most often occur in patients younger than 20 years [9]. In our case, the patient is an elderly woman. The lesion filled the intramedullary space, and the cortex was only thinned and presented with ridge-like changes. This is not consistent with the imaging appearance of OFD; therefore OFD was not considered.

Conclusions

Adamantinoma is relatively rare tumor that is difficult to differentiate from other bone tumors. Although the diagnosis of primary adamantinoma of the tibia depends on histopathologic and immunohistochemical findings, imaging may also be helpful in making an early diagnosis of primary adamantinoma. Adamantinoma should be considered in the differential diagnosis of any tibial anterior cortical lesion, regardless of size, shape, or benign signs.

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