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CASE REPORT

Myoepithelial carcinoma of tibia mimic giant cell tumor: a case report with emphasis on MR features

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Abstract

Intraosseous myoepithelial carcinoma is an extremely rare type of bone tumor that most often presents in the long tubular bones, but also occurs in small tubular bones and the axial skeleton. We report the radiographic images and complete magnetic resonance (MR) features of a 44-year-old male with right knee pain of 7 months' duration. The radiographic findings and convention MR images indicated a giant cell tumor of the bone. The dynamic contrast-enhanced images showed a patent with the early wash-in and early wash-out usually noted in a giant cell tumor of the bone. Only water restriction on diffusion-weighted imaging (DWI) showed the malignant impression. Care should be taken when conventional images indicate giant cell tumor of the bone, as intraosseous myoepithelial carcinoma, although rare, can mimic this more common diagnosis. Further studies with DWI are warranted.

Keywords Intraosseous myoepithelial carcinoma · MRI · Diffusion-weighted imaging

Introduction

Intraosseous myoepithelial carcinoma is an extremely rare type of bone tumor that most often presents in long tubular bones, but also occurs in small tubular bones and the axial skeleton [1]. Only a few case reports have presented the imaging findings of intraosseous myoepithelial carcinoma [2–8]. The tumor is usually presented as a well-demarcated, osteolytic tumor with or without cortical destruction and invasion of the surrounding soft tissue [1]. However, the magnetic resonance imaging (MRI) findings are not well discussed.

We report a case of an intraosseous myoepithelial carcinoma in the proximal tibia, interpreted as an aggressive bone tumor with high SI on T2-weighted (T2W) fat-suppressed (FS) images and intermediate SI on T1-weighted (T1W) images, mimicking a giant cell tumor of the bone. The dynamic contrast-enhanced images presented the patent as early wash-in and early wash-out, a condition usually noted in a giant cell tumor of the bone. Only water restriction on diffusion-weighted imaging (DWI) revealed the malignant impression. To our knowledge, it is the first case of an intraosseous myoepithelial carcinoma with complete MRI and the functional MR images of dynamic contrast-enhanced imaging and DWI may provide the information about the possible malignant characteristics of the mass.

Case report

A 44-year-old male presented with right knee pain of 7 months' duration. There was no trauma history. The patient's medical and family histories were unremarkable. The physical examinations were unremarkable and there was no neurovascular disturbance. Plain radiographs showed a well-defined, expansive, geographic lytic lesion in the proximal tibia and cortical destruction and periosteal reaction at the adjacent posterior cortex of the tibia (Fig. 1).

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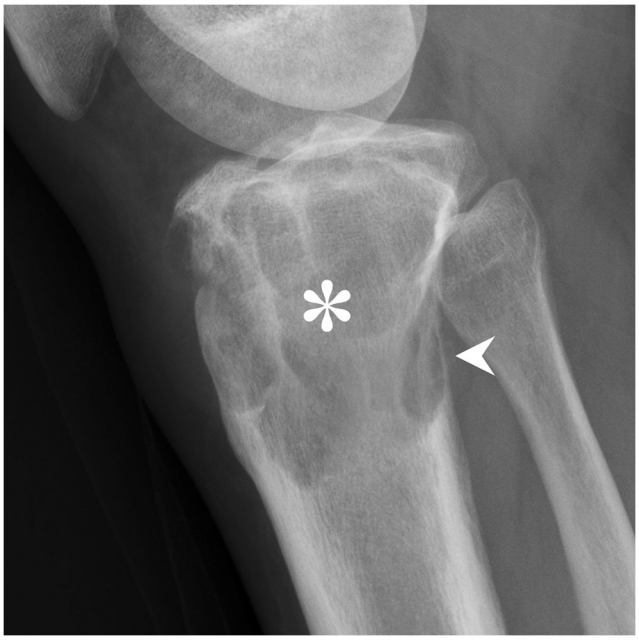

Fig. 1 Radiography. Lateral radiographs showed an expansive, geographic lytic lesion in the proximal tibia (*white star*) and cortical destruction with periosteal reaction at the adjacent posterior cortex of the tibia (*white arrowhead*)

The patient underwent 3.0-T MR imaging, including axial iterative decomposition of water and fat with echo asymmetry and least squares estimation (IDEAL) T1W images, sagittal and coronal fast spin-echo (FSE) T1W images; axial, sagittal, and coronal FSE T2W FS images; axial DWI (b = 0, 1000); axial dynamic contrast media enhanced images; and axial, sagittal and coronal contrast-enhanced T1W FS images. An apparent diffusion coefficient (ADC) map was obtained by using the DWI. The MR images showed a lobulated mass lesion, about 5.8 cm × 6.4 cm × 4.9 cm, in the proximal tibia, mainly in the epiphysis and metaphysis. The lesion showed intermediate intensities on T1W images and hyperintensities on FS T2W images (Fig. 2a–b). The adjacent bone cortex was destroyed, and invasion of the adjacent periosteal area is noted (Fig. 2a–d). After administration of contrast media, most components of the lesion, especially at the peripheral regions, showed intense enhancement (Fig. 2c–d). There were irregular regions without enhancement, mainly in the central part of the lesion, usually indicating tumor necrosis or cystic degeneration (Fig. 2c–d). The location, morphology, and intensity of the lesion fitted the typical findings of a giant cell tumor of the bone. The dynamic pattern of contrast enhancement showed early wash-in and early wash-out (Fig. 2e), suggestive of an aggressive tumor. The DWI (Fig. 2f–g) showed high intensities; low ADC values (mean ADC: about 900) were noted at the regions with enhancement in the lesion. The findings suggested water restriction of the lesion, a condition commonly noted in malignant tumors.

The pathologic report of the open biopsy indicated a myoepithelial neoplasm of uncertain malignant potential. For general survey, the Tc-99 m MDP whole-body scan and chest CT were performed and there was no metastasis. The patient then received extensive tumor curettage with autograft, allograft, and internal fixation of locking plate. This tumor showed growth of predominant sheets or nodules of epithelioid cells with amphophilic to clear cytoplasm. The background revealed a fibromyxoid to sclerotic matrix. The tumor infiltrated into the surrounding fibroadipose tissue. Infarction-type necrosis was noted. Moderate nuclear pleomorphisms and visible nucleoli were identified focally. Mitotic figures are frequently seen focally (up to 14/10 in high-power field). Immunohistochemically, the tumor cells expressed cytokeratin AE1/AE3 and S-100 proteins, epithelial membrane antigen and vimentin focally, but not p63 (Fig. 3). In the molecular study, EWSR1 and FUS gene fusions are demonstrated using a fluorescence in situ hybridization (FISH) assay. The overall features supported the diagnosis of myoepithelial carcinoma.

After the tumor curettage, the patient received regular radiography follow-up per 6 months. In the radiography after 18 months, a local osteolytic lesion was noted in proximal tibia, just below the previous tumor site. The MRI showed a new hypervascular lesion in the proximal tibia and local recurrent tumor is suspected. Then, the patient received tumor curettage with allograft and demineralized bone matrix.

Discussion

Myoepithelial tumors of soft tissue were first reported by Kilpatrick and colleagues in 1997 as skin adnexal or salivary gland tumors resembling myoepithelial components [9]. According to the 2013 World Health Organization (WHO) classification of tumors of soft tissue and bone, myoepithelial tumors with malignant histomorphology and clinical behavior are named myoepithelial carcinomas. Most known cases of myoepithelial tumors are located in soft tissue and only rare cases have been reported in bone [2–8]. In the immunohistochemistry examination, the large majority of myoepithelial tumors demonstrate expression of cytokeratin, EMA, S100, and glial fibrillary acidic protein [1]. Different from the myoepithelial tumors in salivary glands, the molecular pathology of EWSR1 gene fusion is noted in nearly half of myoepithelial tumors of bone [1]. In our case, EWSR1 and FUS gene fusions are demonstrated.

In these myoepithelial tumors of the bone, most cases were benign tumors and reports of intraosseous myoepithelial carcinomas are extremely rare. In these reported intraosseous myoepithelial carcinomas, most tumors presented as expansile, osteolytic lesions [1–3, 5, 7]. Some cases showed sclerotic margin or sclerotic matrix [1–3]. Cortical destruction

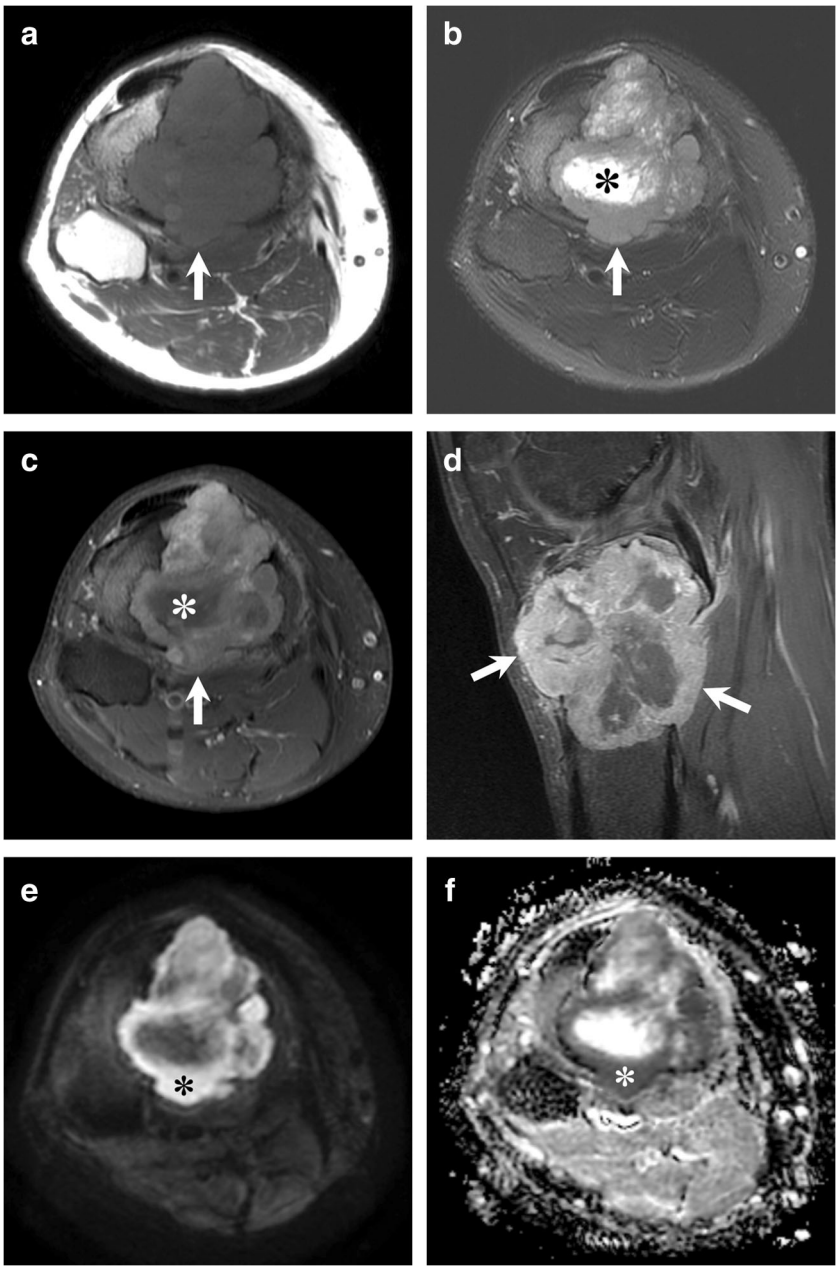

Fig. 2 Conventional magnetic resonance (MR) examination combined with dynamic enhanced images and diffusion-weighted imaging (DWI). The MR images showed a lobulated mass lesion, about 5.8 cm × 6.4 cm × 4.9 cm, in the proximal tibia, mainly in the epiphysis and metaphysis. The lesion showed intermediate intensities on T1-weighted (T1W) images (**a**) and hyperintensities on T2-weighted (T2W) fat-suppressed (FS) images (**b**). Bony destructions with periosteal invasion (a–d, *white arrow*) were noted at the anterior and posterior cortex of the proximal tibia, relatively well identified in the post-enhanced sagittal images (**d**). The post-enhanced T1W FS axial (**c**) and sagittal (**d**) images showed intense enhancement at the most component of the lesion, especially at the peripheral regions. The irregular regions without enhancement (**c**, *white star*) occurred mainly in the central part of the lesion, with hyperintensities on T2W images (**b**, *black star*) indicating tumor necrosis or cystic degeneration. The dynamic contrast-enhanced images (**e**) showed a patent with early wash-in and early wash-out at the lesion (curve 1). The diffusion-weighted images (**f**) showed high intensities and low apparent diffusion coefficient (ADC) values (**g**) were noted at the regions with enhancement in the lesion

was noted in all cases [1–3, 5, 7]. However, MR images were presented only sporadically, with reports typically presenting the common findings of tumors, hyperintensities on T2W FS images, and intermediate to hypointensities on T1W images [2, 3]. No reports completely demonstrated the MR features.

In our case, the radiologic presentation of an osteolytic, expansile mass with cortical destruction was similar to that of previously reported cases. The extraosseous tissue invasion could be well identified in the MR images. In terms of intensity, the case presented hyperintensities on T2W FS images and intermediate intensities on T1W images. The post-enhanced images showed intense enhancement of the lesion and well-identified central necrosis. The intensity features of our case are consistent with previous reports, but such findings are common and non-specific in osteolytic tumors of the bone. According to the radiologic presentation, the intensity of the MR images and the location of the tumor give a first impression of a giant cell tumor of the bone [10, 11].

The vascular features of myoepithelial carcinoma of the bone are not reported. Dynamic contrast-enhanced MRI after a bolus injection of contrast media is usually used to evaluate the vascularity of lesions according to the resulting enhancement curve [12, 13]. The dynamic enhancement curve of our case showed early wash-in and early wash-out, common findings in both malignant tumors and aggressive lesions such as giant cell tumors of the bone. Giant cell tumors of the bone

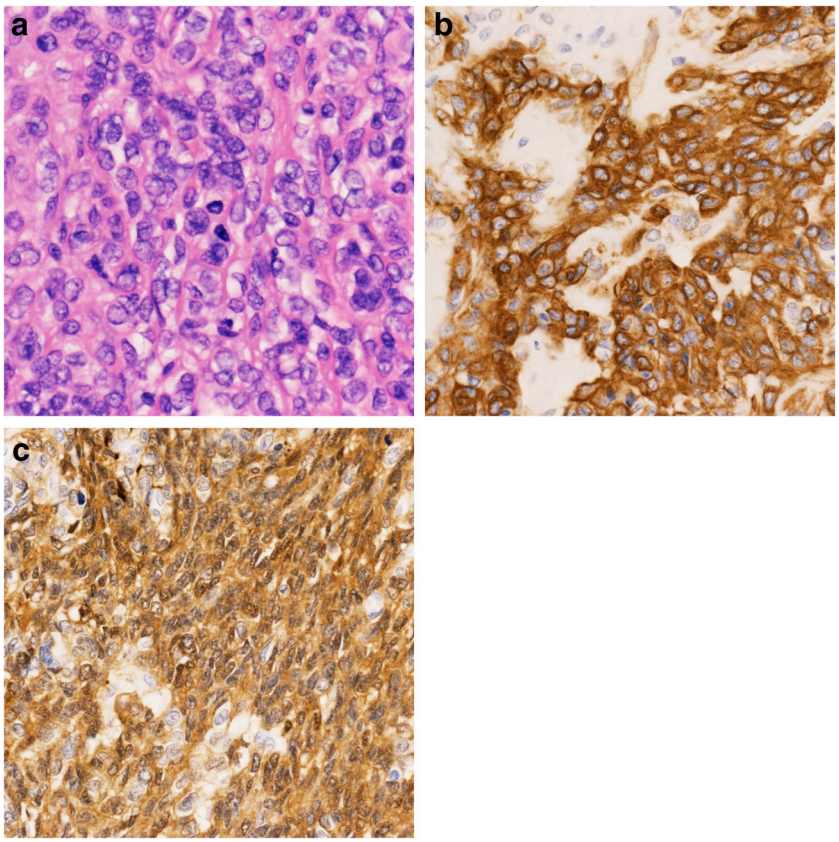

Fig. 3 Immunohistochemical results of tumor tissue analysis. This tumor showed growth of predominant sheets or nodules of epithelioid cells with amphophilic to clear cytoplasm in the hematoxylin and eosin (H&E) stains, and the background revealed a fibromyxoid to sclerotic matrix. Moderate nuclear pleomorphisms and visible nucleoli were identified focally, and mitotic figures were seen up to 14/10 in the high-power field (**a**, HE, 400×). Immunohistochemically, the tumor cells expressed cytokeratin AE1/AE3 (**b**) and S-100 protein (**c**) focally

usually present with early wash-out, although malignant giant cell tumors of the bone may present with a relatively rapid wash-out [13]. According to the vascularity pattern of our case provided by the dynamic contrast-enhanced MRI, a giant cell tumor of the bone was still the first impression, although malignant giant cell tumors of the bone could not be ruled out.

The functional evaluation of MR images using DWI is not reported for intraosseous myoepithelial carcinoma. DWI is an unenhanced evaluation based on the Brownian motion of water that is affected by the tissue microenvironment [14]. The ADC values are derived from DWI and the increase of tumor cellularity could result in the decrease of ADC values [14]. Although the primary bone malignancies are relatively rare, the previous studies suggested that most malignant bone tumors may present lower ADC values and the use of DWI with ADC mapping is an effective method for distinguishing malignant from benign bone tumors [14–16]. In our case, the diffusion-weighted images showed hyperintensities and the ADC map showed hypointensities. The findings suggested that the tumor had the feature of water restriction, a common finding in a malignant tumor [14–16]. Previous reports found that giant cell tumors of the bone usually show high intensities on diffusion-weighted images and the ADC map [16–18]. In our case, the low ADC value was the only key point to rule out the possibility of a giant cell tumor of the bone. A low ADC value reflects a highly cellular microenvironment that suggests a malignant tumor, not a benign giant cell tumor of the bone. Although the finding could not differentiate a myoepithelial carcinoma from a malignant giant cell tumor, the impression of a malignant tumor may influence the treatment algorithm.

In conclusion, intraosseous myoepithelial carcinomas are very rare, but an accurate diagnosis can be made with the support of a wide panel of immunohistochemical markers. However, the radiographic and conventional MR findings may mimic other benign bone tumors, such as a giant cell tumor of the bone in our case. The functional MR images of dynamic contrast-enhanced imaging and DWI may provide more information about the possible malignant characteristics of the mass than the conventional MR images and help guide extensive tumor excision with a possible safe margin to prevent recurrences.

Compliance with ethical standards

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. The study was approved by our Institutional Review Board, and the requirement of informed consent was waived.

Conflict of interest The authors declare that they have no conflicts of interest.

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