Case Report
Clinicopathological features of primary myoepithelial carcinoma inside oblique eminence of cuboid bone: a case report

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Abstract: Primary intraosseous myoepithelial carcinomas are extremely rare tumors. It is important to make an accurate pathological diagnosis for timely and proper treatment. We evaluated imaging, histological, and immunohistochemical studies of a new case and performed a comprehensive review of the related literature. Plain and enhanced scan CT showed that the left proximal femur had an osteolytic lesion and speckled calcification, inhomogeneous enhancement was also observed after reinforcement. Microscopically, multinodular tumors were comprised mostly of polygonal cells within a stroma containing a variable amount of myxoid, chondroid, hyalinized, and osteoid-like material. Through immunohistochemistry, the tumor cells had diffuse strong staining for CK, P63, and P16. The Ki-67 index was less than 10%. We conclude that correct diagnosis must be achieved by imaging, histological, and immunohistochemical stains. Primary intraosseous myoepithelial carcinoma also should be distinguished from metastatic myoepithelial carcinomas from the salivary gland, conventional osteosarcoma, and epithelial/cartilaginous differentiation in clear cell chondrosarcoma.

Keywords: Myoepithelial carcinomas, rare bone tumors, immunohistochemistry, imaging, histomorphology

Introduction

Myoepithelial tumors are composed of neoplastic cells that have a myoepithelial phenotype. Primary myoepithelial tumor has also been described in the bones as a distinct tumor entity. Diagnosis of a malignant myoepithelial tumor (MET) or a myoepithelial carcinoma is based upon the presence of at least moderate nuclear atypia [1]. There are very few reported cases of intraosseous myoepithelial carcinomas so far [2, 3].

Due to their rarity, unusual morphology, and intraosseous origin, myoepithelial tumors have frequently caused diagnostic difficulties. To better understand the biological features of these tumors, we report here one case and go through comprehensive review of the literature.

Case report

Patients and methods

This study consisted of a consultation case of a 32-year-old male with 6 month discomfort from his left hip, with excruciating pain. We also performed a comprehensive review of the literature.

Clinical data

Plain radiography showed expansive cystic low density shadow on left femur, with uneven and patchy high density shadow of the intracapsular. Plain and enhanced scan CT indicated that the left proximal femur had a osteolytic lesion, with speckled calcification (Figure 1A), and inhomogeneous enhancement after reinforcement (Figure 1B). Plain and enhanced scan nuclear magnetic resonance imaging (MRI) found
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A large abnormal signal in the left femur that extended to the inferior horizontal medullary cavity. Local cortical destruction, soft tissue mass formation, and an obviously inhomogeneous enhancement were also observed. The bone scan displayed increased bone metabolism in the proximal femur. Color Doppler ultrasound suggested that bilateral salivary glands and neck lymph nodes had no obvious abnormality.

**Follow-up results**

In our case, the patient received hip replacement. There was no adjuvant radiochemotherapy needed and there was no recurrence and metastasis for nine month post operation.

**Discussion**

Primary intraosseous myoepithelial tumors include myoepitheliomas/mixed tumours and ra-
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The diagnosis in this case was reinforced with positive immunohistochemical coexpression of epithelial markers along with P63. The tumor cells showed focal staining for S100 and SMA. The studies in a series of 14 METs, including myoepithelial carcinomas of soft tissues [7], observed EMA positivity in 10/12 tumors (83%), CK positivity in 3/12 tumors (25%), along with S100 protein and glial fibrillary acidic protein (GFAP) (11/13, 85%) and, (6/12, 50%) tumors, respectively. In equivocal cases, they recommended P63, CD10, calponin, and SMA as additional, useful, surrogate markers. In Table 1, among 10 intraosseous malignant myoepithelial carcinoma cases, EMA was positive in 7/10 tumors, CK was positive in 4/10, pan CK and SMA were positive in 4/10, P63 and GFAP were positive in 5/10. Additionally, S100 was positive in 6/10.

Several differential diagnoses need to be considered before making diagnosis of an intraosseous malignant MET. These diseases are as follows: (1) Metastatic tumors, especially malignant mixed epithelial tumors and myoepithelial carcinomas from the salivary gland need to be excluded by clinical-radiologic examination. (2) Osteosarcoma, such as in our case where there were many metaplastic bones. STAB2 is positive in osteoblast but negative in tumor cells. The atypia of tumor cell is not obvious. Osteosarcoma is neoplastic osteogenesis. STAB2 is positive in tumor cells of osteosarcoma. In radiographic imaging, osteosarcoma may be completely lytic or sclerotic, but usually a combination of these features enables a preoperative radiographic diagnosis of osteosarcoma in majority cases. Osteosarcoma growing on the surface of bone can elevate the periosteum and induce a periosteal reaction in the form of...
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Table 1. Clinicopathological Features of intraosseous myoepithelial carcinomas

<table>
<thead>
<tr>
<th>Case</th>
<th>Age/Sex</th>
<th>Site</th>
<th>Histopathological features</th>
<th>Matrix</th>
<th>Immunohistochemical results</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 (2)</td>
<td>55/M</td>
<td>Femur</td>
<td>Cords</td>
<td>Hyalinized-to-chondromyxoid</td>
<td>Vimentin+, pan CK+, EMA+, muscle actin+, SMA+, calponin+, caldesmon+, P63+, desmin+, CK, MNF116+, CK7, CK20, CK14, GFAP, S-100</td>
<td>Surgical Resection (Rx)</td>
<td>Metastasis (lung) (13 months)</td>
</tr>
<tr>
<td>2 (3)</td>
<td>21/M</td>
<td>Humerus</td>
<td>Cords</td>
<td>Myxochondroid</td>
<td>CK+, GFAP+, Vimentin+, desmin+, P63+, S100</td>
<td>Surgical Resection (Rx)</td>
<td>No</td>
</tr>
<tr>
<td>3 (4)</td>
<td>34/M</td>
<td>Left Femur</td>
<td>Cords</td>
<td>Myxochondroid</td>
<td>CK+, GFAP+, Vimentin+, desmin+, P63+, S100</td>
<td>Left Extra-articular Hip resection (RO)+ metastatectomy</td>
<td>FOD (10 months)</td>
</tr>
<tr>
<td>4</td>
<td>23/M</td>
<td>Right Tibula</td>
<td>Cords</td>
<td>Myxochondroid</td>
<td>CK+, GFAP+, Vimentin+, desmin+, P63+, S100</td>
<td>Wide excision</td>
<td>FOD (12 months)</td>
</tr>
<tr>
<td>5</td>
<td>8/M</td>
<td>Left Tibula</td>
<td>Cords</td>
<td>Chondromyxoid with squamous metaplasia</td>
<td>CK+, GFAP+, Vimentin+, desmin+, P63+, S100</td>
<td>Surgical Resection (R1)</td>
<td>On FU</td>
</tr>
<tr>
<td>6</td>
<td>40/M</td>
<td>Phalanx</td>
<td>Nests, Cords</td>
<td>Hyaline/sclerotic</td>
<td>CK+, GFAP+, S100, SMA+, S100, P63, desmin, SMA, CD10, CD34, Ki67, 10%</td>
<td>Surgical Resection (Rx)</td>
<td>On FU</td>
</tr>
<tr>
<td>7</td>
<td>26/M</td>
<td>Femur</td>
<td>Cords, Diffuse</td>
<td>Chondromyxoid</td>
<td>CK+, GFAP+, S100, SMA+, S100, P63, desmin, SMA, CD10, CD34, Ki67, 10%</td>
<td>Surgical Resection (Rx)</td>
<td>Metastatectomy (paraspinous region and lung), On FU</td>
</tr>
<tr>
<td>8 (5)</td>
<td>41/F</td>
<td>Maxilla bone</td>
<td>Nests, Spindle cells+small epithelial+plasmacytoid cells</td>
<td>Myxoid</td>
<td>CK+, GFAP++, CD10, S100, Ki67, 10%</td>
<td>Local excision+ chemoradiation</td>
<td>Metastasis (lung) (16 months, die)</td>
</tr>
<tr>
<td>9 (6)</td>
<td>48/F</td>
<td>Rib</td>
<td>Cords</td>
<td>Myxoid</td>
<td>P63+, calponin+, SMA+, EMA+, GFAP, S-100, CK, CK7, CK20, CK14</td>
<td>Surgical Resection (Rx)</td>
<td>On FU</td>
</tr>
<tr>
<td>10</td>
<td>32/M</td>
<td>Oblique</td>
<td>Nests</td>
<td>Myxoid</td>
<td>CK+, P63+, S100, SMA+, Calponin+, CD34, Ki67, 10%</td>
<td>Surgical Resection (Rx)</td>
<td>On FU</td>
</tr>
</tbody>
</table>

M, Male; F, Female; CK, Cytokeratin; +, Positive; -, Negative; EMA, Epithelial membrane antigen; GFAP, Glial fibrillary acidic protein; SMA, Smooth muscle actin; BU, Brachyury (T); Rx, Surgical resection with unknown marginal status; RO, Surgical resection with clear margins; FOD, Free of disease; FU, Follow-up.

an open triangle overlying the diaphyseal side of the lesion. In this case, plain and enhanced scan CT suggested that left proximal femur had an osteolytic lesion, with speckled calcification, as well as inhomogeneous enhancement detected after reinforcement. (3) Epithelial and cartilaginous differentiation was seen in clear cell chondrosarcoma. It is a rare, low-grade variant of chondrosarcoma with a tendency to occur in the humeral or femoral head of elderly patients. Clear cell chondrosarcoma mainly consists of large, rounded clear cell component and woven bone, associated with low-grade chondrosarcoma components. In this case, there are many cartilage components. But chondrocytes are not atypia. There are few clear cells.

Additional benefit of adjuvant chemotherapy has not been reported in malignant cases [8].

In our case, the patient has never received adjuvant radiochemotherapy.

Conclusion

In summary, we analyzed a rare intraosseous myoepithelial carcinoma, which mimics primary osseous and cartilagenous tumors. The correct diagnosis for such a case only can be achieved by imaging, histological, and immunohistochemical stains, which has treatment implications. So far, surgical resection with clear margins remains the better choice of therapy.

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Disclosure of conflict of interest

None.

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References


