Cutaneous myoepithelioma: A case report of an unusual and recently recognized entity

Abstract

Myoepitheliomas and mixed tumors were only recently recognized to occur primarily in soft tissue, and only small case numbers have been described. The present case is a 25-year-old male who had solitary, painless mass over the right middle finger, measuring 3 cm in greatest dimensions, and light microscopy revealed a tumor composed of a mixed population of the spindle, epithelioid, and plasmacytoid cells arranged around a central chondromyxoid stroma. No definite diagnosis could be reached on this morphology, and initial diagnosis of benign mixed stromal tumor was considered. Immunohistochemistry was performed and the tumor showed strong positivity for calponin and smooth muscle actin, Ki-index showed low index, weak, and focal positivity for S-100 and negative for pan-cytokeratin. The final diagnosis of benign myoepithelioma was entertained.

Key words: Cutaneous, Myoepithelioma, Soft tissue.

INTRODUCTION

Myoepithelioma of the skin and soft tissue is a newly recognized entity only 10 years ago with fewer than 50 case reports. It has characteristic histopathologic and immunohistochemical features, should be considered as differential diagnosis for a variety of tumors. The primary differential diagnoses considered were extraskeletal myxoid chondrosarcoma (EMC) and ossifying fibromyxoid tumor (OFMT). S-100 protein and epithelial markers are expressed in a minority of EMC and usually only focally while both the markers are often extensively expressed in myoepitheliomas. Approximately 70% of OFMT show positivity for S-100 protein and vimentin and 50% of tumor cells are positive for desmin. The tumor cells in OFMT are rarely positive for epithelial markers and glial fibrillary acidic protein (GFAP). Myoepitheliomas are generally negative for desmin, nearly half positive for GFAP, and nearly always show positivity for keratin and S-100 protein.[1] Other tumors that should be differentiated are epithelioid benign fibrous histiocytoma spitz nevus and epithelioid sarcoma.[2]

CASE REPORT

Case history

A 25-year-old male presented with a solitary, painless mass over the right middle finger, measuring 3 cm in greatest dimension for 6 months. There is no increase in size and overlying skin is smooth and unremarkable. The mass is firm, nontender, and mobile. X-ray showed a soft tissue mass with underlying bone unremarkable.

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Light microscopy

It revealed a tumor in the superficial dermis composed of a mixed population of spindle, epithelioid, and plasmacytoid cells arranged around a central chondromyxoid stroma. No definite diagnosis could be reached on this morphology and diagnosis of benign mixed stromal tumor was considered (Figure 1).

Immunohistochemistry

Immunohistochemistry was performed and showed strongly positive calponin and smooth muscle actin (SMA). S-100 was weak and focally positive. Pan-cytokeratin was negative (Figure 2). Ki 67 (Figure 3) revealed low index (2%) as mentioned in Table 1.

The final diagnosis of benign myoepithelioma was entertained.

DISCUSSION

Cutaneous myoepitheliomas are relatively rare accounting for 1% of all cutaneous soft tissue tumors. Although 80% cutaneous and soft tissue myoepitheliomas behave in a benign fashion, 3.4% have a risk for local recurrence and a low metastatic potential and 1% has a risk of malignant transformation. Tumors comprised mostly of myoepithelial cells without obvious epithelial differentiation are designated myoepitheliomas.[1] Neoplasms of myoepithelial cells can occur in a pure form as myoepitheliomas or in association with glandular structures as mixed tumors.[2] Myoepitheliomas of the skin and soft tissue were recognized only 10 years ago.[3] Myoepithelial cells can exhibit dual epithelial and myoid differentiation. They may also show divergent metaplasia, including squamous, adipocytic, bone, and cartilaginous differentiation.[4,5] As a consequence, proliferating myoepithelial cells in neoplasms display a variety of histologic and immunohistochemical expression patterns. It has been postulated that cutaneous myoepitheliomas are related to mixed tumors of the skin and that soft tissue myoepitheliomas are derived from deeply located adnexal structures. Cutaneous myoepitheliomas of the head and neck may be derived from salivary gland tissue as has been reported in two parotid gland myoepitheliomas presenting as infra-auricular subcutaneous masses.[6] Myoepithelial tumors were described only recently in soft tissue, and to date, fewer than 50 cases have been reported. Kilpatrick et al.[7] reported a study of 19 patients with mixed tumors and myoepitheliomas of soft tissue in 1997. Michal and Miettinen[8] reported 12 additional cases of myoepitheliomas of the skin and soft tissues in 1999, Hornick and Fletcher conducted a study of 14 cutaneous myoepitheliomas. There were 11 males and 3 females. The study indicated that cutaneous myoepitheliomas occur with peaks in childhood (seven patients were between 10 and 20 years of age) and middle age and are most common on the extremities, in contrast to mixed tumors of the skin, which typically occur on the head and neck in middle-aged or elderly adults.[1,3,4]

Myoepitheliomas of soft tissue are often lobulated, and the most frequent architectural pattern is reticular or trabecular with chondromyxoid or hyalinized stroma. These lesions display the same wide range of histologic features as those of salivary gland origin.

Many tumors are heterogeneous, containing an admixture of epithelioid and spindled cells, reticular areas merging with solid
Table 1: Showing Immunohistochemistry data

<table>
<thead>
<tr>
<th>Immunohistochemistry markers</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>Calponin</td>
<td>Strongly positive</td>
</tr>
<tr>
<td>Smooth muscle actin</td>
<td>Strongly positive</td>
</tr>
<tr>
<td>Ki 67</td>
<td>Low index (2–4%)</td>
</tr>
<tr>
<td>S-100</td>
<td>Weak and focal</td>
</tr>
<tr>
<td>Pan cytokeratin</td>
<td>Negative</td>
</tr>
</tbody>
</table>

areas, at least focally prominent stroma, and occasional foci of cartilaginous or osseous differentiation. A small subset of tumors approximately 10% are predominantly solid proliferations of spindled or plasmacytoid myoepithelial cells. Initially, myoepitheliomas were only recognized to contain spindled or plasmacytoid cells growing in solid sheets. Current classifications, therefore, include all of these patterns within the spectrum of myoepithelioma, simply separating those tumors with ductal differentiation into the mixed tumor category. Whereas some investigators allow up to 5% or 10% ductal differentiation in myoepitheliomas, others classify tumors with any ducts as mixed tumors. In any event, it is now widely thought that myoepitheliomas and mixed tumors fall along a spectrum of tumors with overlapping histologic appearances and similar clinical behavior. Because the immunophenotype of these lesions overlaps with myoepithelioma, and otherwise typical myoepitheliomas can show focal areas with "parachordoma"-like features, it is becoming increasingly clear that parachordoma probably falls within the spectrum of myoepithelioma of soft tissue as is reflected in the new WHO classification. The only apparent difference in immunophenotype is GFAP and SMA negativity in parachordomas because few cases of parachordoma have been studied and only about 50% of otherwise convincing soft tissue myoepitheliomas are GFAP positive and only around 40% are SMA-positive, then this distinction seems very questionable. Awareness of the wide morphologic range of myoepitheliomas is necessary to perform confirmatory immunohistochemical stains and thereby to arrive at the correct diagnosis. In salivary glands, myoepitheliomas are generally positive for cytokeratins and S-100 protein, whereas immunostaining for actin and GFAP is variable. We therefore required immunoreactivity for either keratin or EMA, in conjunction with detection of S-100 protein or myogenic markers, for the diagnosis of myoepithelioma and inclusion in this series. Neoplastic myoepithelial cells of all morphologic types often expressed myogenic markers.

CONCLUSION

Cutaneous myoepitheliomas are relatively rare. Pathologists play an important role in reaching to accurate morphological diagnosis. Myoepitheliomas should be considered in the differential diagnosis of cutaneous and soft tissue tumors. Immunohistochemical study may aid in the diagnosis. Although cutaneous and soft tissue myoepitheliomas behave in a benign fashion, they have a risk for local recurrence and a low metastatic potential and has a risk of malignant transformation. Wide excision with safe surgical margins and regular follow-up are crucial for the management of cutaneous and soft tissue myoepitheliomas.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES