

Cutaneous myoepithelioma | case study



**ASSIGN
BUSTER**

TITLE: 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 of 25-year-old male who had solitary, painless mass over right middle finger, measuring 3cm in greatest dimensions and light microscopy revealed a tumor 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 initial diagnosis of benign mixed stromal tumor was considered.

Immunohistochemistry(IHC) was performed and the tumor showed strong positivity for Calponin and SMA, Ki-index showed low index, weak and focal positivity for S-100 and negative for Pan-CK. The final diagnosis of benign myoepithelioma was entertained.

Keywords: Myoepithelioma, Cutaneous, Soft tissue

INTRODUCTION:

Myoepithelioma of the skin and soft tissue is a newly recognized entity only ten years ago with fewer than 50 case reports. It has characteristic histopathologic and immunohistochemical features, which need to be differentiated from a variety of tumors

CASE REPORT:

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

Gross:

Multiple tissue bits aggregate measuring 3×2×1cm and skin flap measuring 2×1cm.

Light microscopy:

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.

Immunohistochemistry(IHC):

IHC was performed and showed strongly positive Calponin and SMA. S-100 was weak and focally positive. Pan CK was negative. Ki 67 revealed low index (2%).

The final diagnosis of Benign myoepithelioma was entertained.

DIFFERENTIAL DIAGNOSIS

The primary differential diagnoses considered were extraskelatal myxoid chondrosarcoma (EMC) and ossifying fibromyxoid tumor (OFMT). EMC

typically shows a multinodular growth pattern with cords of cells in a myxoid matrix. The tumor cells in EMC are more spindled than those of myoepitheliomas. 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.

OFMT is

a lobulated tumor surrounded by a rim of metaplastic bone. The tumor cells are mostly

pale-staining ovoid to round cells. 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 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 that usually shows a superficial dermal tumor with a well-developed epidermal collarette. Spitz nevus is characterized by a junctional

component, nesting and maturation of tumor cells. In epithelioid sarcoma, multiple tumor nodules around central necrosis or even myxoid degeneration are often seen. More morphologic uniformity is observed in epithelioid sarcoma over myoepithelioma. Moreover, approximately 90% of epithelioid sarcoma are positive for vimentin, cytokeratin, and EMA, and around 60% are positive for CD34, but are generally negative for other markers typical myoepithelial differentiation (S-100 protein, GFAP, myogenic markers). Cellular neurothekeoma consists of nesting of tumor cells, and are consistently S-100 negative. [2]

COMMENT

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 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]

Therefore, the possibility of an underlying primary salivary gland neoplasm should be considered in myoepitheliomas presenting in the head and neck.

Myoepithelial tumors were described only recently in soft tissue, and, to date, fewer than 50 cases have been reported. Kilpatrick et al [3] reported a study of 19 patients with mixed tumors and myoepitheliomas of soft tissue in 1997. Michalet et al [7] 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 (7 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 areas, at least focally prominent stroma, and occasional foci of cartilaginous or osseous differentiation. [8] A small subset

of tumors approximately 10% are predominantly solid proliferations of spindled or plasmacytoid myoepithelial cells. Occasional tumors display features of so-called parachordoma, namely, large epithelioid cells with eosinophilic epitheliomas. Initially, myoepitheliomas were only recognized to contain spindled or plasmacytoid cells growing in solid sheets. ^[9] 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. ^[10, 11] 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. ^[11] 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. ^[12] 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. ^[13] As has been reported in the salivary gland, we found calponin to be the most sensitive myogenic marker, staining 86% of tumors, whereas SMA stained 36% and desmin only a small subset (14%). Interestingly, the basal cell/myoepithelial marker p63, which has shown utility in the differential diagnosis of carcinoma of breast and prostate due to the staining of myoepithelial or basal cells in in situ lesions, appears to be detectable in only one fourth of soft tissue myoepithelial tumors. Immunostaining for p63 is not specific for myoepithelial tumors, however, as this antigen has also been reported in other neoplasms, especially squamous cell and urothelial carcinomas. ^[13, 14] Nonetheless, detection of p63 expression may provide helpful supportive evidence of myoepithelial differentiation in the proper morphologic context.

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 most cutaneous and soft tissue myoepitheliomas behave in a benign fashion, there is a significant risk for local recurrence and a low metastatic potential. Wide excision with safe surgical margins and

regular follow-up are crucial for the management of cutaneous and soft tissue myoepitheliomas.

References:

1. Hornick JL, Fletche CDM. Myoepithelial tumors of soft tissue a clinicopathologic and Immunohistochemical study of 101 cases with evaluation of prognostic parameters. *Am J Surg Pathol.* 2003; 27: 1183–1196.
2. Hornick JL, Fletcher CD. Cutaneous myoepithelioma: a clinicopathologic and immunohistochemical study of 14 cases. *Hum Pathol.* 2004; 35: 14-24.
3. Kilpatrick SE, Hitchcock MG, Kraus MD, Calonje E, Fletcher CD. Mixed tumors and myoepitheliomas of soft tissue: a clinicopathologic study of 19 cases with a unifying concept. *Am J Surg Pathol.* 1997; 21: 13-22.
4. Mentzel T, Requena L, Kaddu S et al. Cutaneous myoepithelial neoplasms: clinicopathologic and immunohistochemical study of 20 cases suggesting a continuous spectrum ranging from benign mixed tumor of the skin to cutaneous myoepithelioma and myoepithelial carcinoma. *J Cutan Pathol.* 2003; 30: 294-302.
5. Fernández-Figueras MT, Puig L, Trias I, Lorenzo JC, Navas-Palacios JJ. Benign myoepithelioma of the skin. *Am J Dermatopathol.* 1998; 20: 208-12.
6. Lewin MR, Montgomery EA, Barrett TL. New or unusual dermatopathology tumors: a review. *J Cutan Pathol.* 2011; 38: 689-96.

7. Kutzner H, Mentzel T, Kaddu S et al. Cutaneous myoepithelioma: an under-recognized cutaneous neoplasm composed of myoepithelial cells. *Am J Surg Pathol.* 2001; 25: 348-55
8. Dix BT, Hentges MJ, Saltrick KR, Krishnamurti U. Cutaneous myoepithelioma in the foot: case report. *Foot Ankle Spec.* 2013; 6: 239-41.
9. Michal M, Miettinen M. Myoepitheliomas of the skin and soft tissues. Report of 12 cases. *Virchows Arch.* 1999; 434: 393-400.
10. Franklin G, Chen S, Szynter LA, Morgenstern NJ. Cutaneous myoepithelioma with a plexiform pattern of growth: a case report. *J Cutan Pathol.* 2009; 36: 42-5.
11. Jo VY, Antonescu CR, Zhang L et al. Cutaneous Syncytial Myoepithelioma: Clinicopathologic Characterization in a Series of 38 Cases. *Am J Surg Pathol.* 2013 ; 37: 710–718.
12. Jakate K, Wong K, Sirbovan J, Hanna W. Cutaneous myoepithelioma arising within hidradenoma of the scalp. *J Cutan Pathol.* 2012; 39: 279-85.
13. Stojisic Z, Brasanac D, Boricic I, Bacetic D. Clear cell myoepithelial carcinoma of the skin. A case report. *J Cutan Pathol.* 2009; 36: 680-3.
14. Tanahashi J, Kashima K, Daa T, Kondo Y, Kuratomi E, Yokoyama S. A case of cutaneous myoepithelial carcinoma. . *J Cutan Pathol.* 2007; 34: 648-53.

LEGENDS

Figure 1: Myoepithelioma(100X): composed of a mixed population of spindled, epithelioid, and plasmacytoid cells arranged around a central chondromyxoid stroma. Insert shows 400X view and reveals mild nuclear atypia (coarse chromatin and prominent nucleoli) .

Figure 2: Myoepithelioma(400X): (a) shows SMA positivity in the cytoplasm of myoepithelial cells.(b) shows strong positivity for Calponin in the cytoplasm of spindle cells.(c) shows PAN CK negativity(d) shows S100 negativity.

Figure 3 : Myoepithelioma(400X): shows Low Ki 67 index.