Cutaneous myoepithelioma with cartilaginous metaplasia: A new (but not surprising) histopathologic feature

Ashley R. Mason MD†, Shelby Y. Kunishima MD†, Jonathan D. Cuda MD†, Jennifer M. McNiff MD†

Department of Dermatology, Yale School of Medicine, New Haven, CT; †Department of Pathology and Laboratory Medicine, Yale School of Medicine, New Haven, CT; †Bethesda Dermatopathology Laboratory, Silver Spring, MD

History of present illness

A 37-year-old man with Fitzpatrick Type II skin presented with a new papule on the left flank. He had no significant medical history and was on no medications. Family history was unremarkable.

Physical examination

Physical examination showed a pink papule on the left anterolateral flank measuring 0.3 cm in greatest diameter. Skin examination was otherwise unremarkable except for a benign facial angioma and multiple lentigines on the chest.

Histopathology

A biopsy specimen showed a dome-shaped tumor with a broad central zone of cartilaginous differentiation (Figure 1). Outside the cartilaginous zone, epithelioid cells with eosinophilic cytoplasm formed a vague syncytium without a ductal component (Figure 2). Mitoses were less than 1 per 10 high power fields. Immunostaining for S100 protein and epithelial membrane antigen (EMA) was strongly positive in the chondrocyte-like cell nuclei as well as in the surrounding epithelioid tumor cells. Focal staining was seen for cytokeratin antibodies, Cam 5.2 and AE1/AE3 (Figure 3). Staining for Melan-A was negative.

Course

The lesion was excised. To date, no recurrence has occurred at 3 months.

Discussion

Cutaneous myoepitheliomas are rare, benign skin tumors composed of myoepithelial cells without ductal differentiation. Myoepithelial cells occur in normal skin surrounding the secretory portions of apocrine and eccrine glands and represent one component of cutaneous mixed tumors. Most reported cutaneous myoepitheliomas present as dome-shaped, exophytic nodules on the head, neck, and extremities of adults and range in size from 0.5 to 2.5 cm. Histologically, these are well-circumscribed dermal lesions composed of spindled, ovoid, epithelioid, or plasmacytoid cells that can be arranged in clusters, strands, or sheets. Some tumors have sparse stroma, while others show a hyalinized or myxoid stroma. Epidermal hyperplasia with hyperkeratosis is sometimes present.

Cartilaginous metaplasia within cutaneous myoepitheliomas has not yet been reported in the literature. However, this is not a surprising histologic finding considering the ability of myoepithelial cells to differentiate along mesenchymal lines in addition to epithelial lines. Cutaneous mixed tumors/chondroid syringomas are also on the spectrum of myoepithelial neoplasia with distinct chondromyxoid stroma, but these lesions show epithelial differentiation, usually in the form of ductal structures, which myoepitheliomas lack.

Cutaneous myoepitheliomas tend to stain positively for S100 protein, actins, GFAP, calponin, EMA and cytokeratins. Melan-A staining was negative, militating against the possibility of a melanocytic lesion with cartilaginous differentiation. Although cutaneous myoepitheliomas are mainly benign neoplasms treated by complete simple excision, one must always rule out a diagnosis of myoepithelial carcinoma, myoepithelioma’s malignant counterpart. In our patient, the mitotic rate was very low to absent, and cystic features were bland, making the diagnosis of a myoepithelial carcinoma highly unlikely.

Cartilaginous metaplasia could pose a potential pitfall for misclassification of a myoepithelioma as a benign cartilaginous tumor. However, primary cartilaginous tumors would not stain for cytokeratin or EMA. Recognition of this pattern of differentiation in primary cutaneous myoepitheliomas expands our understanding of the histologic spectrum of this rare tumor. To the best of our knowledge, this is the first reported case of a cutaneous myoepithelioma with cartilaginous metaplasia.

References