

Cutaneous Myoepithelial Carcinoma With Disseminated Metastases

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PRACTICE POINTS

- Cutaneous myoepithelial carcinoma is a rare malignant adnexal neoplasm with metastatic potential that can present in the skin.
- Cutaneous myoepithelial carcinoma is a tumor that can occasionally show *EWSR1* gene rearrangement.
- Excision with negative margins and close follow-up is recommended for cutaneous myoepithelial carcinoma.

Primary cutaneous neoplasms of myoepithelial differentiation are uncommon. Cutaneous myoepithelial carcinomas are rare. We report a case of cutaneous myoepithelial carcinoma in a 47-year-old man with a history of end-stage renal disease and renal transplant 19 years prior who presented to the hospital with a 3-month history of diffuse bone pain and an ulcerated scalp mass with multiple satellite lesions. This case illustrates a rare instance of metastatic disease from primary cutaneous myoepithelial carcinoma.

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Cutaneous myoepithelial tumors are rare neoplasms but are being increasingly recognized and reported in the literature.¹⁻⁷ Myoepithelial tumors are related to benign mixed tumors of the skin but lack the epithelial ductules that are present in mixed tumors. Cutaneous myoepithelial tumors may show a variety of architectural, cytological, and stromal features. Their immunophenotype usually is characterized by coexpression of an epithelial marker (eg, keratin, epithelial membrane antigen [EMA]) and S-100 protein; they also may express a variety of other myoepithelial markers, including keratins, smooth muscle actin, calponin, glial fibrillary acidic protein, p63, and desmin.⁷ EWS RNA binding protein 1 (*EWSR1*) and pleomorphic adenoma gene 1 (*PLAG1*) gene rearrangement has been detected in subsets of these tumors on in situ hybridization.⁸⁻¹⁰

Malignant myoepithelial tumors of the skin, also referred to as cutaneous myoepithelial carcinomas, are exceedingly rare. Including the current case, a search of PubMed articles indexed for MEDLINE and Google Scholar using the terms *myoepithelial carcinoma* and *cutaneous* revealed 12 cases that have been reported in the literature (Table).^{1-7,11-13} These tumors often occur in the head and neck areas and the lower extremities and display a bimodal age distribution, generally occurring in patients younger than 21 years and older than 50 years of age; they also show a slight female predominance. Available

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follow-up data from the literature have shown local recurrence or metastasis in 3 cases^{3,4,6}; however, in one case the metastatic focus was not histologically identified.⁴ Cutaneous myoepithelial carcinoma presenting with metastatic disease further limits treatment options. Here, we describe a case of metastatic cutaneous myoepithelial carcinoma in a 47-year-old man, a rare example of cutaneous myoepithelial carcinoma with histologically documented metastatic disease at the initial presentation.

Case Report

A 47-year-old man who underwent a renal transplant 19 years prior presented with a weeping, ulcerated, mildly tender lesion on the scalp of 4 months' duration with neck and back pain of 3 months' duration. Physical examination demonstrated a 6-cm area of ulceration on the anterior crown of the scalp with adjacent enlarged keratoacanthomalike craters and satellite nodules (Figure 1). He was previously diagnosed with basal cell carcinoma (BCC) of the scalp at an outside institution 4 years prior and was treated with radiation therapy. The prior scalp biopsy for BCC diagnosis was unavailable for review. The patient had a history of chronic eczematous dermatitis in the waistband area that had been present for 19 years and another BCC with nodular and infiltrative patterns on the left helix. Of note, he also had been taking long-term immunosuppressant medications (ie, cyclosporine, azathioprine) for maintenance following the renal transplant.

Because of the extensive ulceration of the primary lesion, a shave biopsy of the scalp was performed on an adjacent satellite nodule. Histopathologic findings showed an intradermal neoplasm characterized



Figure 1. Myoepithelial carcinoma. Weeping, ulcerated, mildly tender lesion on the scalp showing central ulceration with multiple satellite nodules.

by poorly cohesive cells exhibiting epithelioid to plasmacytoid morphologic features surrounded by abundant chondromyxoid stroma. Ductular differentiation was not identified (Figure 2A). The neoplastic cells displayed hyperchromatic nuclei with marked nuclear pleomorphism and atypical mitotic figures (Figure 2B). On immunohistochemistry the tumor cells stained positive for cytokeratin AE1/AE3 (Figure 3), S-100 protein (Figure 4), and p63, and were negative for calponin, desmin, melan-A, cytokeratin 7, and brachyury (Figure 5).

Radiographic imaging was performed due to the patient's history of neck and back pain. Magnetic resonance imaging showed innumerable slightly expansile, T1-hypointense, T2-hyperintense, and robustly enhancing lesions involving the cervical, thoracic, lumbar, and sacral spine, as well as the

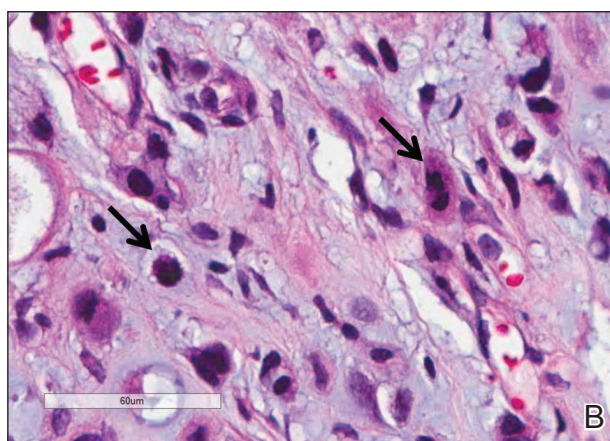
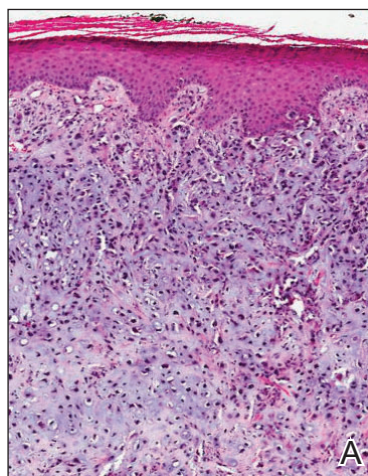


Figure 2. Histology revealed a dermal-based neoplasm with chondromyxoid stroma closely approximating the epidermis (A)(H&E, original magnification $\times 40$). High magnification showed neoplastic cells with hyperchromatic nuclei, marked nuclear pleomorphism, and atypical mitotic figures (arrows)(B)(H&E, original magnification $\times 400$).

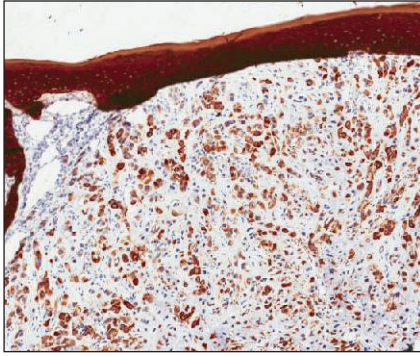


Figure 3. Immunohistochemistry showed positive staining for cytokeratin AE1/AE3 (original magnification $\times 40$).

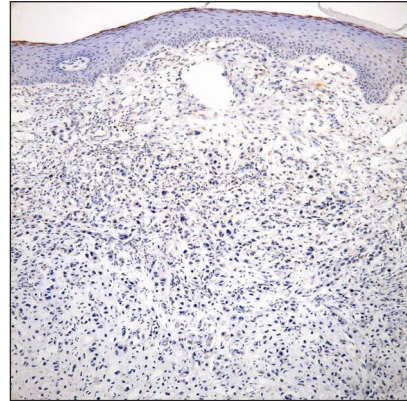


Figure 5. Immunohistochemistry showed negative nuclear staining for brachyury (original magnification $\times 40$).

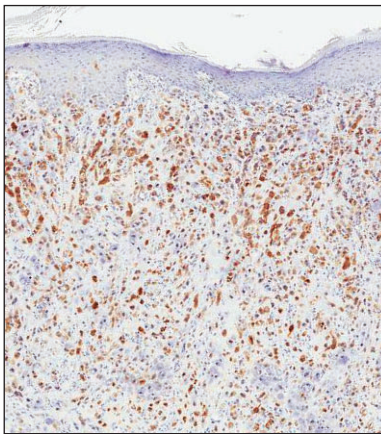


Figure 4. Immunohistochemistry showed positive staining for S-100 protein (original magnification $\times 40$).

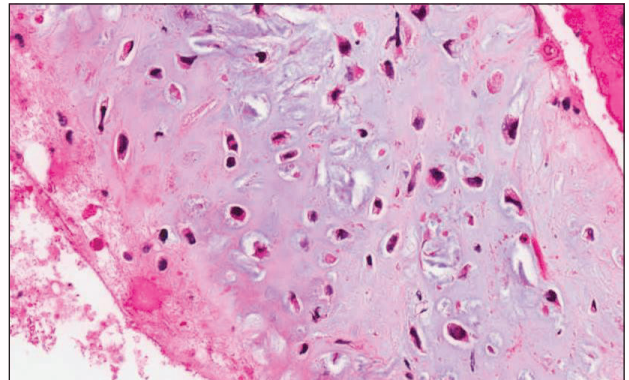


Figure 6. Fine needle aspiration cytopathology of a metastatic osseous lesion showed hyperchromatic nuclei in abundant chondromyxoid stroma (identical morphologic features as the cutaneous lesion)(H&E, original magnification $\times 200$).

thoracic ribs and bilateral iliac bones. There was no evidence of soft tissue tumor around the bone lesions. Ventral cervical spinal cord compression was detected at the C4 vertebra, causing a symptomatic radiculopathy; however, due to widely metastatic disease, the patient was not considered appropriate for neurosurgical intervention of the compression. Computerized tomography of the chest, abdomen, and pelvis did not identify any visceral source of malignancy, though multiple bilaterally enlarged cervical lymph nodes were identified on magnetic resonance imaging.

Fine needle aspiration of a left iliac bone lesion demonstrated neoplastic cells and chondromyxoid stroma essentially identical to the features shown in the skin biopsy (Figure 6). Given the morphologic features of the tumor and coexpression of cytokeratin and S-100 protein, the findings were interpreted as primary cutaneous myoepithelial carcinoma with disseminated metastatic lesions. The patient began treatment with carboplatin and paclitaxel

chemotherapy. To combat the symptomatic bone pain and upper extremity radiculopathy, palliative radiation was administered to the cervical spine, lumbar spine, and right sacrum (30 Gy to each site in 10 fractions at 3 Gy per fraction). Despite the attempted chemotherapy and radiation, the patient continued to decline, and after 2 months, he elected to pursue palliative care. The patient died after 3 months in palliative care (5 months after the initial presentation).

Comment

Myoepithelial cells normally surround ducts in secretory organs, such as the breasts, salivary glands, and cutaneous sweat glands. Myoepithelial neoplasms are well recognized in the salivary glands^{14,15}; however, myoepithelial neoplasms also can arise in other sites, including the soft tissue^{4,5,16-18} and skin.^{1-3,7,11,19,20} Myoepithelioma of soft tissue was

first described by Burke et al²¹ in 1995 and later described in the skin by Fernandez-Figueras et al²² in 1998. Since then, diagnostic criteria for cutaneous myoepithelial neoplasms have evolved, suggesting a spectrum of disease rather than a single distinct entity.¹¹ Most often, cutaneous myoepithelial carcinomas arise as soft nodular lesions in the head and neck areas or extremities of adults. The nodules typically are nontender and range in size from 0.5 to 18.0 cm. Our review of the literature revealed 11 additional cases of cutaneous myoepithelial carcinomas have been reported, ranging in size from 0.7 to 7.0 cm (Table). In our case, the main lesion was 6 cm, mildly tender, ulcerated, and accompanied by satellite nodules.

Histologically, cutaneous myoepithelial tumors typically are well-defined, dermal-based nodules with no connection to the overlying epidermis. Similar to myoepithelial tumors of other sites, they can be diagnostically challenging due to the heterogeneity of both their architectural and cytological features. The presence of a chondromyxoid or hyalinized stroma is common but not always present. Neoplastic myoepithelial cells can exhibit spindled, epithelioid, plasmacytoid, or clear cell morphologic features and show growth patterns in clusters, cords, glands, or sheets. Focal epithelial cells can be present. Although benign myoepithelial neoplasms with overt ductal differentiation are consistent with cutaneous mixed tumors (chondroid syringomas), those without ducts are characterized as myoepitheliomas. It is uncertain if cases with only focal ductal differentiation should be classified as mixed tumors or as myoepitheliomas. Malignant myoepithelial tumors show infiltrative borders, nuclear pleomorphism, coarse nuclear chromatin, prominent nucleoli, and increased mitotic activity. A 2003 study by Hornick and Fletcher¹⁶ found that cytologic atypia was the primary predictor of malignant behavior for myoepithelial neoplasms of the soft tissue.

Despite a wide variety of expression patterns, immunohistochemistry is critical in demonstrating myoepithelial differentiation and establishing a diagnosis of a myoepithelial neoplasm. Most cases display coexpression of epithelial markers, including keratins and/or EMA as well as S-100 protein. Myogenic markers also may be variably expressed; however, the absence of myogenic markers does not exclude the diagnosis of a myoepithelial tumor. Commonly expressed epithelial markers are cytokeratin AE1/AE3, cytokeratin 8/18, and EMA, while commonly expressed myogenic markers include muscle specific actin and smooth muscle actin.^{5,7,11,19} Myoepithelial tumors also may express calponin, p63, and glial fibrillary acidic protein.¹⁶

Molecular studies also can aid in the diagnosis of myoepithelial tumors. A study by Antonescu et al⁸ demonstrated *EWSR1* gene rearrangement in 45% (30/66) of extrasalivary myoepithelial tumors and the absence of *EWSR1* gene rearrangement in salivary gland myoepithelial tumors. The authors also showed that *EWSR1*-negative tumors were more likely to be superficially located, display ductal differentiation, and possess a benign clinical course.⁸ In another study, Bahrami et al²³ suggested that a subset of mixed tumors, specifically those with tubuloductal differentiation, are genetically linked to salivary gland pleomorphic adenomas, which was achieved by the coexpression of the *PLAG1* protein and *PLAG1* gene rearrangement on immunohistochemistry and fluorescence in situ hybridization (FISH), respectively. Of the 19 cases evaluated, 11 (58%) expressed nuclear staining for *PLAG1* immunohistochemistry; 8 of those 11 showed positive gene rearrangement for *PLAG1* using FISH. These findings raise the possibility that cutaneous mixed tumors may be more closely related to those of the salivary glands, while deep myoepithelial tumors that lack ductal differentiation may represent a distinct group. Similar to the study by Antonescu et al,⁸ Flucke et al¹⁰ investigated *EWSR1* gene rearrangement but limited their sample to cutaneous tumors, including myoepitheliomas, mixed tumors, and myoepithelial carcinoma. The authors found that 44% of cases (7/16) expressed *EWSR1*; this expression suggests that cutaneous myoepithelial tumors may have a genetic relationship to their soft tissue, bone, and visceral counterparts.¹⁰

Myoepithelial tumors display a broad spectrum of morphologic features; however, one of the most common growth patterns is that of oval to round cells forming cords and chains in a chondromyxoid stroma. As such, the histopathologic differential diagnosis for myoepithelial tumors includes other epithelioid or round-cell neoplasms with similar growth patterns including extraskeletal myxoid chondrosarcoma (EMC), ossifying fibromyxoid tumor of soft parts, and extra-axial soft tissue chordoma. Extraskeletal myxoid chondrosarcoma bears the closest similarity to myoepithelial tumors both histologically and by ancillary studies. It typically possesses cords or chains of small round tumor cells set in a chondromyxoid or myxoid background. In contrast to myoepithelial tumors, which typically have more abundant cytoplasm and can show at least focal areas of spindle cell growth, the cells of EMC are more uniform, small, round cells with relatively scant cytoplasm. Extraskeletal myxoid chondrosarcomas lack the typical myoepithelial coexpression of cytokeratin and S-100 protein, with a minority of

Cases of Cutaneous Myoepithelial Carcinoma

Reference (Year)	Patient Age, y/Sex	Lesion Location	Lesion Size, cm	Symptoms	Diagnosis	Follow-up, mo	Metastasis/Recurrence	Histology	IHC (+)
Michal and Miettinen ⁴ (1999)	52/F	Left sole	2.5	NR	MC	17	Patient died of multiple pulmonary metastases after 1.5 y	No ductal or syringomatous epithelial structures or cartilaginous areas were identified; no hair follicle, apocrine, sebaceous differentiation, or necrosis	S-100, cytokeratin
Hornick and Fletcher ⁷ (2004)	14/F	Left great toe	2.0	Swelling, redness	MC	NR	NR	Cytologically malignant with severe atypia (coarse chromatin and prominent large nucleoli); there was a small focus of tumor necrosis and 39 mitoses per HPF	Pan-cytokeratin, cytokeratin 8/18, EMA, GFAP, calponin, SMA
Menzel et al ¹¹ (2003)	70/F	Left cheek	1.0	NR	MC	24	NED	Dermal and SC lesions not connected with overlying epidermis or deeper structures; cellular neoplasm composed of atypical cells with enlarged and pleomorphic nuclei containing occasionally prominent nucleoli; necrosis and numerous mitoses were identified; ductal structures were not identified	Vimentin, pan-cytokeratin, EMA, SMA, calponin (focal)
79/M		Left retro-auricular region	0.7	NR	MC	24	NED	Nodular dermal neoplasm with extension into SC tissue with plump, spindled, round, and stellate tumor cells set in a prominent myxoid matrix, with tissue culture-like appearance in some areas; tumor cells contained a well-delineated, pale, eosinophilic cytoplasm and enlarged pleomorphic nuclei with prominent nucleoli; scattered mononucleated and multinucleated tumor giant cells were noted; mitotic rate was increased (mean, 16 per HPF) with atypical mitoses found	Vimentin, pan-cytokeratin, SMA, S-100 (focal), calponin (focal)
Tanahashi et al ⁹ (2007)	62/F	Right side of the back	7.0	Lesion was present for 9 y with no change, then rapidly increased in size over a 10-mo period	MC	NR	15 mo later, local recurrence and metastasis to the right lateral chest wall	Superficial to mid dermis contained small nests with collagenous or myxoid stroma; multiple large nodules with central necrosis located in the deep dermis and subcutis; tumor cells were epithelioid and plasmacytoid intermingled with clear cells in the large deep nodules; rare superficial multinucleated giant cells present and small nucleoli; nuclear pleomorphism was not prominent, and mitotic figures were present in the deep but rare in superficial; lymphatic invasion present deep	Dermal component: pan-cytokeratin, cytokeratin 14, 34βE12, vimentin, p63, S-100 (weak), GFAP (weak)
Gleason and Fletcher ⁵ (2007)	9/M	Great toe	1.3	NR	MC	20	NED	At least moderate cytologic atypia with nuclear enlargement, vesicular or coarse chromatin, and variably prominent nucleoli; no benign precursor identified	Cytokeratin, calponin (weak)

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Table. (continued)

Reference (Year)	Patient Age, y/Sex	Lesion Location	Lesion Size, cm	Symptoms	Diagnosis	Follow-up, mo	Metastasis/Recurrence	Histology	IHC (+)
Law et al ⁶ (2008)	13/F	Right parieto-temporal region	NR	Nodule developed over several months	MC	NR	Metastasis, primarily to the lymph nodes	Spindle cell lesion with focal nesting; no ductal differentiation; cells displayed vacuolated cytoplasm, open chromatin, and focally prominent nucleoli; mitotic figures readily identified	Broad-spectrum cytokeratin, S-100, SMA, calponin
Stojic et al ² (2009)	51/F	Right femoral region	1.0	8 mo	MC	20	3 local recurrences after NED	Dermal and SC lesion with multinodular pattern; clear cells predominated, with rare epithelioid and spindled cells in scant myxoid and collagenous stroma; nuclear atypia and mitotic activity were present; a Ki-67 index of 20% was identified	Cytokeratin AE1/AE3, EMA, vimentin, SMA, muscle-specific actin, S-100
Bajoghli and Limpert ¹³ (2009)	2.5/F	Nose	NR	Developed over several months	MC	48	NED	NR	NR
Garcia-Sanchez et al ¹² (2009)	NR	NR	NR	NR	MC	NR	NR	NR	NR
Frost et al ¹ (2014)	80/M	Scalp	2.0	Developed over 2 mo	MC	18	NED	Located deep in the dermis, extended into the subcutaneous adipose tissue, and infiltrated the underlying facial muscle; the tumor comprised poorly defined sheets and strands of highly pleomorphic cells; the cells had a spindlelike or polygonal shape with pleomorphic nuclei and often prominent nucleoli; the cytoplasm was abundant, eosinophilic, and mostly homogeneous, though some cells showed slight granularity; the tumor stroma was rather vascular with only a few inflammatory cells; atypical mitotic figures were observed, and the Ki-67 proliferation index was as high as 80% in some tumor regions	Pan-cytokeratin (KL-1), cytokeratin 5/6, cytokeratin 7 (focal), vimentin, SMA
Current case	47/M	Scalp	6.0	4-mo history of weeping, ulcerated, and mildly tender scalp lesion	MC	5	Multiple metastases to bone	Intradermal lesion of poorly cohesive cells with epithelioid to plasmacytoid morphology with abundant chondromyxoid stroma; nuclear hyperchromasia and pleomorphism were identified; atypical mitotic figures were present; ductular differentiation was not identified.	Cytokeratin AE1/AE3, S-100, p63

Abbreviations: IHC, immunohistochemistry; F, female; NR, not reported; MC, myoepithelial carcinoma; HPF, high-power field; EMA, epithelial membrane antigen; GFAP, glial fibrillary acidic protein; SMA, smooth muscle actin; NED, no evidence of disease; SC, subcutaneous; M, male.

EMCs expressing S-100 protein but rarely cytokeratin. Most cases of EMC possess a balanced t(9;22) translocation involving the *EWSR1* gene,²⁴ a finding that could lead to confusion with soft tissue myoepithelial tumors, which also may show *EWSR1* rearrangement on FISH. Ossifying fibromyxoid tumor of soft parts is also composed of round cells arranged in cords in a myxoid or fibrous stroma; the majority of cases also display a peripheral rim of mature bone, a feature that is not typically seen in myoepithelial tumors. Similar to myoepithelial tumors, ossifying fibromyxoid tumor of soft parts often is positive for S-100 protein; however, it rarely is positive for cytokeratins. Ossifying fibromyxoid tumor of soft parts has been shown to have a rearrangement of the PHD finger protein 1 (*PHF1*) gene in approximately half of cases, a molecular finding that has not been reported for myoepithelial tumors.²⁵ Finally, extra-axial soft tissue chordomas, though quite rare, may possess striking similarities to myoepithelial tumors both histopathologically and immunohistochemically. Chordomas are composed of epithelioid cells arranged in nests, nodules, and chains with a variably myxoid background. A variable amount of cells with bubbly cytoplasm (known as physaliphorous cells) can be seen. High mitotic activity is not a characteristic feature in chordomas. They classically coexpress cytokeratins and S-100 protein, similar to myoepithelial tumors. A subset of myoepitheliomas with similar histologic features to chordoma was historically referred to as parachordoma.^{26,27} The distinction between these 2 entities was challenging until the relatively recent advent of brachyury, a sensitive and specific nuclear marker of chordoma; extra-axial soft tissue chordomas and their central counterparts both express nuclear brachyury, while myoepitheliomas (including those with a parachordoma histologic pattern) do not.²⁸ Our case did not display physaliphorous cells but did demonstrate abundant nuclear pleomorphism and high mitotic activity. In addition, immunohistochemical staining was negative for brachyury.

Because cutaneous myoepithelial tumors are relatively rare, a well-defined standard of care for treatment is lacking. Surgical excision is the primary treatment method in most reported cases in the literature.^{17,19} Miller et al²⁹ reported the successful treatment of recurrent cutaneous myoepitheliomas with Mohs micrographic surgery. Chemotherapy may be useful in the setting of metastatic myoepithelial carcinomas in adults, but reported results are inconsistent.^{30,31} Radiation treatment of recurrent or metastatic disease has not been shown to be effective. A study of children treated with surgical resection and chemotherapy using ifosfamide, cisplatin,

and etoposide followed by radiation therapy showed positive results.³²

Our case highlights several challenges that may arise in establishing a diagnosis of cutaneous myoepithelial carcinoma with disseminated metastases. The diagnostic difficulty in our case was compounded by the advanced nature of the lesion at the time of presentation. Given the rarity of metastatic cutaneous myoepithelial carcinomas and the lack of a prior primary diagnosis of a malignant myoepithelioma, the index of suspicion for this entity was not high. A report of myoepithelial carcinoma of the parotid gland metastatic to the skin has been reported,³³ but in the absence of salivary gland involvement or other visceral lesions, metastasis from any source other than our patient's cutaneous scalp lesion is unlikely. The histopathologic features in combination with the characteristic immunophenotype, unique clinical setting, and radiographic findings were essential to arriving at the correct diagnosis. Unlike previously reported metastatic lesions, our case is unique in that metastatic lesions were identified at the time of initial clinical presentation.

Conclusion

Cutaneous myoepithelial carcinomas are exceedingly rare tumors with a wide range of histopathologic and immunohistochemical findings. In challenging cases, studies for *EWSR1* or *PLAG1* gene rearrangement can be helpful. Furthermore, this case illustrates the potential for widespread dissemination of myoepithelial carcinomas requiring clinical evaluation and imaging studies to exclude metastatic lesions.

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