Metastatic Eccrine Porocarcinoma

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Practice Points

- Eccrine porocarcinoma (EP) is a rare malignancy that has the potential to metastasize to both visceral and cutaneous sites.
- Rarely, EPs can be aggressive with widespread metastasis at presentation.
- Lesional pain is an infrequent finding, but it may suggest malignant transformation or metastasis.

Eccrine porocarcinoma (EP) is a rare malignant tumor arising from the intraepidermal sweat duct. The lesions have a varied clinical appearance and usually are asymptomatic. In reported cases, metastasis is rare but can be devastating when it occurs. We describe an unusual case of EP in a patient who presented with metastatic disease. New-onset exquisite pain prompted his evaluation. This case and review of metastatic disease is presented to raise awareness of this tumor, its varied clinical presentations, differential diagnosis, and management.

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Eccrine porocarcinoma (EP) is a rare malignant tumor arising from the intraepidermal sweat duct. Clinically, EP has a variable appearance and usually is asymptomatic, which can make diagnosis difficult. Because the clinical appearance of the tumors may be misleading, histopathologic features have been found to be useful to confirm the diagnosis of EP. Metastasis is rare and can be either visceral or cutaneous. We describe the case of a patient who presented with metastatic disease and new-onset pain. We review the literature on pain and its association with malignant transformation of a previously benign lesion. This case and review of metastatic disease is presented to raise awareness of this rare malignancy and its varied clinical presentations, differential diagnosis, and management.

Case Report

An 87-year-old man presented with an enlarging tender nodule on the left temple that had previously been stable for many years. A review of systems was otherwise negative. On physical examination, a 1-cm violaceous, dome-shaped papule on the left temple and a 3-mm tender, pink, dome-shaped papule on the right upper back were noted. The differential diagnosis for both lesions included angioma; malignancy was not suspected. However, shave excisions were performed due to the history of recent growth of the lesion on the temple and the palpable tenderness of both lesions. Similar histopathologic features were seen in both specimens, which included aggregates of poroid and cuticular cells within the dermis with remarkable cytologic atypia and numerous mitotic figures (more than 14 mitoses per 10 high-power fields [HPFs])(Figure 1). Strong and diffuse staining for carcinoembryonic antigen, CAM 5.2, and AE1/AE3 was noted (Figure 2). CD31, CK20, S-100, HMB-45 (human melanoma black), and CD68 staining was negative against angioma, merkel cell carcinoma, melanoma, metastatic gastrointestinal adenocarcinoma, and histiocytic neoplasm, respectively. The

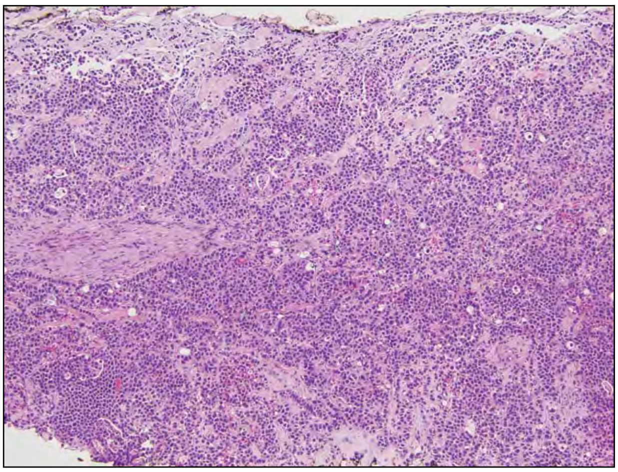

Figure 1. Nests and interanastomosing cords of cytologically atypical epithelial poroid cells with varying degrees of necrosis (H&E, original magnification ×10).

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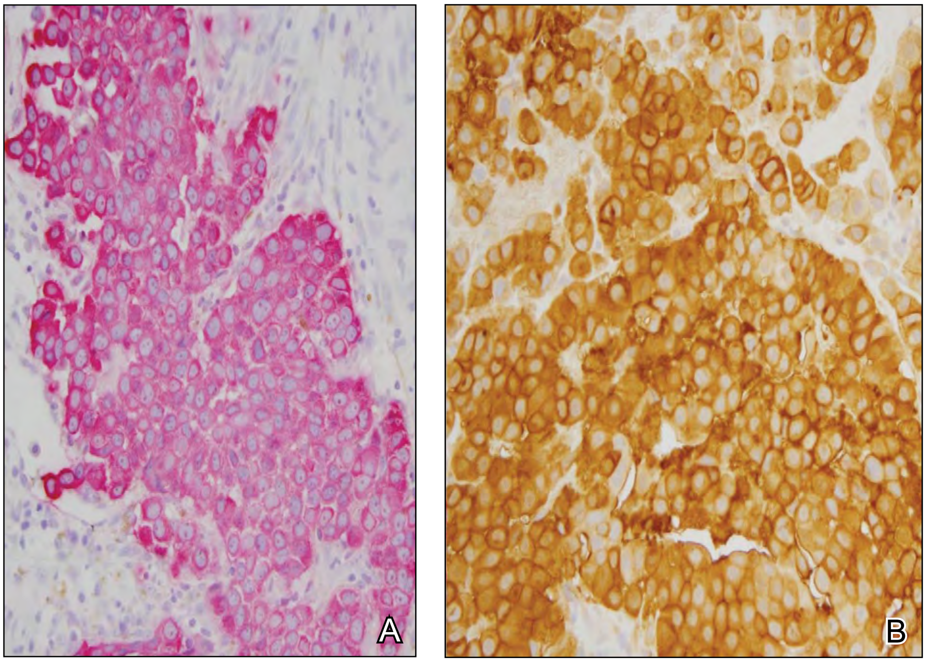

Figure 2. Strong and diffuse staining for carcinoembryonic antigen (A) and CAM 5.2 (B)(both original magnification ×40).

histopathologic features and immunohistochemical profile supported the diagnosis of EP. It was determined that the lesion on the upper back was indicative of metastasis rather than a second primary lesion because of its relative proximity to the primary lesion on the temple and smaller size. Wide local excision along with imaging was recommended despite apparent clear clinical margins, but the patient declined treatment and further evaluation.

Five months after initial diagnosis, the patient returned to report a rapid decline in his quality of life, including fatigue, pain, anorexia, and depression, and requested treatment. He consented to excision of both lesions and imaging. The lesion on the left temple showed positive margins with perineural invasion. Positron emission tomography for staging showed widespread visceral metastatic disease involving the cervical, mediastinal, and hilar lymph nodes; right lung; and multiple bony sites. The patient opted for hospice care.

Comment

Eccrine porocarcinoma is a rare primary adenocarcinoma and represents only 0.01% of all epithelial cutaneous neoplasms.[1] A population-based study using the Surveillance, Epidemiology, and End Results Program data identified 126 cases of EP from 2001 to 2005.[2] The most common location of EPs was the lower extremities, followed by the face. A rapid increase in incidence rates with advanced age also was noted. The median age at diagnosis was 75 years with a male to female incidence ratio of 2.34. The most common ethnicity among patients diagnosed with EP was non-Hispanic white (82%), followed by black (8%) and Hispanic white (6%).[2]

Clinically, EP has a variable appearance, which can make diagnosis difficult. It can appear as a red or flesh-colored papule, plaque, or polypoid or nodular growth.[3] The clinical differential diagnosis includes other cutaneous neoplasms such as squamous cell carcinoma, basal cell carcinoma, amelanotic melanoma, and metastatic adenocarcinoma, as well as benign entities such as seborrheic keratosis, pyogenic granuloma, verruca vulgaris, fibroma, or nevus.[4]

Eccrine porocarcinoma lesions usually are asymptomatic; however, pruritus, sudden growth, and spontaneous bleeding have been reported as presenting symptoms and may suggest malignant transformation.[1,5-8] Metastasis can be either visceral or cutaneous and occurs in approximately 20% of cases.[9] Sites of visceral metastasis include the lungs,[8,10-14] pleura,[11] brain,[15] bones,[11,12,16] muscles,[11] spinal cord,[8,11] liver,[8,10,17] kidneys,[8] retroperitoneum,[13] breasts,[18] mediastinum,[17] urinary bladder,[17] peritoneum,[17] and ovaries.[18] Cutaneous metastasis is rare and may present as multiple flesh-colored or purple papules or nodules.[19] Multinodularity, ulceration, and rapid growth of the primary lesion may be associated with metastasis.[3,4] To our knowledge, pain has not been cited in the literature as a sign of metastasis, though it has been noted in association with malignant transformation of a previously benign lesion. A PubMed search of articles indexed for MEDLINE using the term *eccrine porocarcinoma* generated 4 case reports describing tender lesions in association with EP (Table).[20-23] Two cases described painful lesions with a subsequent negative metastatic workup.[20,23] Another case involved a woman with recurrent painful lesions on her leg that were diagnosed as EP. She later developed a nonpainful lump in her groin that was found to be metastases in her femoral lymph nodes.[22] Another case report described a patient with a painful lesion at presentation that proved to be a metastatic lesion on evaluation. The patient had an EP lesion that previously had been excised from his left palm and presented with a painful buccal lesion that proved to be metastasis.[21] Our patient initially presented with painful lesions that were to be not only malignant but also metastatic.

As the clinical appearance of the tumors may be misleading, histopathologic features have been found to be useful, not only to confirm the diagnosis of EP but also as a guide to prognostic parameters.[24] Robson et al[24] described 3 indicators for poor prognosis in EP: (1) more than 14 mitoses per 10 HPFs; (2) lymphovascular invasion by tumor; and (3) depth greater than 7 mm. Our patient had 1 of 3 indicators for poor

Painful EP Lesions

Reference	Presentation	Site of Primary Tumor	Pain Associaed With Primary Tumor?	Metastasis?	Site of Metastasis	Pain Associated With Metastatic Lesions?
Moussallem et al[20]	77-year-old man presented with a painful recurrent lesion on the right first toe	Right first toe	Yes	No	N/A	N/A
Toshiko et al[21]	80-year-old man presented with a lesion on the left palm and a painful lesion on the left buccal mucosa	Left palm	No	Yes	Left buccal mucosa	Yes
Landa and Winkelmann[22]	24-year-old woman presented with painful recurrent lesions on the left leg; she later presented with a nonpainful lump in the left groin	Left leg	Yes	Yes	Left femoral lymph nodes	No
Johnson et al[23]	45-year-old man presented with a tender pink nodule on the instep of the left foot	Instep of left foot	Yes	No	N/A	N/A
Current case	87-year-old man presented with enlarging tender lesions on the left temple and right upper back	Left temple	Yes	Yes	Right upper back	Yes

Abbreviations: EP, eccrine porocarcinoma; N/A, not available.

prognosis at presentation with biopsies showing more than 14 mitoses per 10 HPFs. Our patient also showed evidence of perineural invasion but no lymphovascular invasion. The depth of the lesions was not initially recorded but on repeat evaluation showed a depth of 6 mm on the temple and 10 mm on the back.

Treatment of primary lesions includes wide local excision, with a cure rate of 70% to 80%.[8,25] Once metastasis occurs, the prognosis is poor. Eccrine porocarcinoma is relatively refractory to chemotherapy and the role of radiation therapy is unclear.[26,27] Because of the rapid decline in our patient's quality of life, he opted for hospice care instead of further treatment.

Conclusion

Eccrine porocarcinoma is a rare malignancy that has the potential to metastasize to both visceral and cutaneous sites. Although metastasis is uncommon, EPs can be aggressive with widespread metastasis at presentation. Lesional pain is an infrequent finding, but it may suggest malignant transformation or metastasis. Our patient presented with tender lesions and his health rapidly declined from metastatic disease. The incidence rates of apocrine-eccrine tumors are increasing, and awareness of the varied clinical presentation of EPs, including their potential to present as benign in appearance, may facilitate diagnosis or prompt referral to a dermatologist for further evaluation and management.[2] Additionally, the sudden onset of pain may be a clue for clinicians on the character of the lesion and should also prompt further evaluation.

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