Endocrine mucin-producing sweat gland carcinoma of the scalp treated with Mohs micrographic surgery

Kristina Navrazhina, BA,[a] Tatyana Petukhova, MD, MS,[a] Horatio F. Wildman, MD,[a] Cynthia M. Magro, MD,[b] and Kira Minkis, MD, PhD[a]
New York, New York

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Endocrine mucin-producing sweat gland carcinoma (EMPSGC) is a rare, low-grade neoplasm most frequently found on the eyelids of elderly women.[1] We present a rare variant of extrafacial EMPSGC on the scalp and discuss successful treatment with Mohs micrographic surgery (MMS).

Abbreviations used:

EMPSGC:	endocrine mucin-producing sweat gland carcinoma
MMS:	Mohs micrographic surgery

REPORT OF A CASE

An 81-year-old white woman with a medical history of Alzheimer's disease, hypertension, and osteoporosis presented for evaluation of a growth on the scalp that was slowly enlarging over 2 to 3 years. She denied trauma or prior treatment to the area. Her physical examination was remarkable for a 1.8-cm poorly demarcated nontender bluish, translucent, lobulated nodule on the vertex scalp with few telangiectasias (Fig 1). Punch biopsy found monomorphic epithelial cells surrounded by abundant mucin and separated by fibrous septa, with extensive immunoreactivity for synaptophysin and focal reactivity for CK7, favoring the diagnosis of EMPSGC (Fig 2). The patient was treated with MMS, and tumor extirpation required 5 Mohs stages to obtain a tumor-free plane with negative margins. The final defect was reconstructed with a rotation flap (see Fig 1).

DISCUSSION

EMPSGC presents as slow-growing, flesh-colored, solid or partially cystic nodules, which typically arise from the periorbital region, with a predilection for the lower eyelid.[1,2] Histologically, EMPSGCs grow as well-circumscribed, multinodular dermal tumors with solid, cystic or papillary differentiation containing intracytoplasmic and extracellular mucin deposition.[1-3] Cytologic atypia is limited within the tumors, and nuclear pleomorphism is not frequently present.[1] The differential diagnoses for EMPSGCs include adenoidal basal cell carcinoma, primary mucinous carcinoma of the skin, and metastatic extracutaneous mucinous carcinoma of breast, lung, gastrointestinal tract, ovarian, prostatic and pancreatic origins.[2] Expression of at least 1 neuroendocrine marker (ie, synaptophysin, CD57, neuron-specific enolase, chromogranin) and low-molecular cytokeratin (CK7 and Cam5.2) is required for diagnosis.[1] The tumor rarely encompasses preexisting benign structures, suggesting it is of primary cutaneous origin.[1] The presence of calponin, p63 and CD10-positive peripheral layer of myoepithelial cells further guides diagnosis toward carcinoma in situ.[2] However, a subset of EMPSGCs shows small tumor islands within mucin lakes, suggestive of a transition to invasive mucinous carcinoma.[1]

From the Department of Dermatology, Weill Cornell Medical College[a] and Department of Pathology and Laboratory Medicine, New York Presbyterian Hospital/Weill Cornell Medicine.[b]
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Correspondence to: Kira Minkis, MD, PhD, Director of Mohs and Dermatologic Surgery, Assistant Professor of Dermatology, Weill Cornell Medical College, 1305 York Avenue, 9th Floor, New York, NY 10021. E-mail: kim9036@med.cornell.edu.

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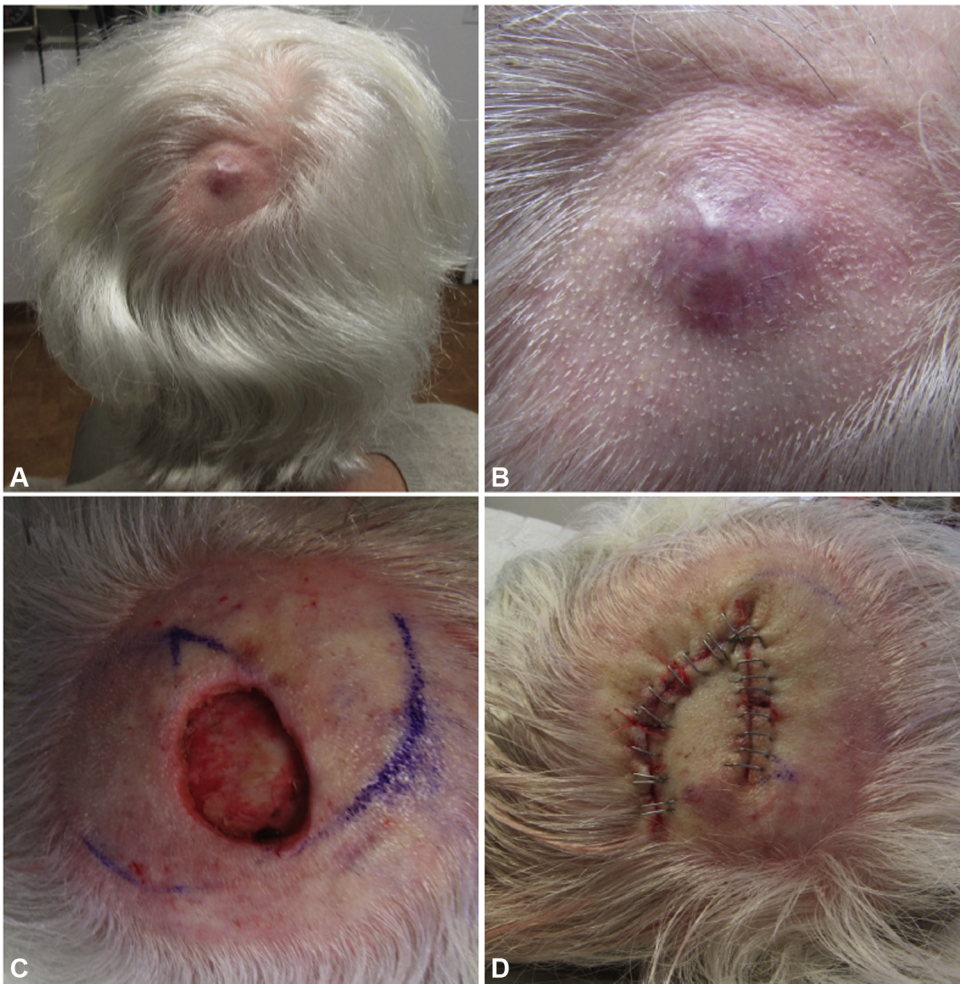

Fig 1. **A** and **B**, A 1.8-cm nontender mass on the scalp vertex, biopsy proven as EMPSGC. **C**, Surgical defect measuring 3.7 × 3.0 cm after 5 stages of MMS. **D**, Surgical reconstruction of the Mohs defect with a rotation flap.

Therefore, it is a current topic of debate as to whether EMPSGC represents a distinct tumor type or is a possible precursor lesion to invasive mucinous carcinoma with neuroendocrine differentiation.[1,3]

EMPSGC carries a favorable prognosis.[1,2] Recommended treatment includes complete surgical removal with at least a 5-mm margin or MMS in cosmetically sensitive areas.[2] Approximately 11% of previously reported cases of EMPSGC had recurred; however, these cases were initially misdiagnosed at presentation and treated without adequate margin control.[4] The median time for EMPSGC recurrence was reported to be 24 months. Given this, a 2-year follow-up is recommended after resection.[4] No reported cases of metastasis or mortality has been attributed to these tumors.[1,2]

Although EMPSGCs have been reported in extraocular areas including the cheek, to our knowledge, this is the first reported case of EMPSGC presenting on the scalp. Because of the rarity of this tumor, minimal data exist comparing outcomes of wide local excision and MMS.[5] Reconstruction of lesions on the scalp can be challenging; the scalp is relatively immobile, and a careful surgical approach must be taken to prevent exposure of the calvarial bone. The significant subclinical extension and deep nature of the tumor, necessitating 5 stages of MMS to achieve negative margins, further lend credence to the use of close margin control and MMS for the treatment of these tumors. Our case shows that MMS is a viable treatment for EMPSGC in cosmetically and surgically difficult regions.

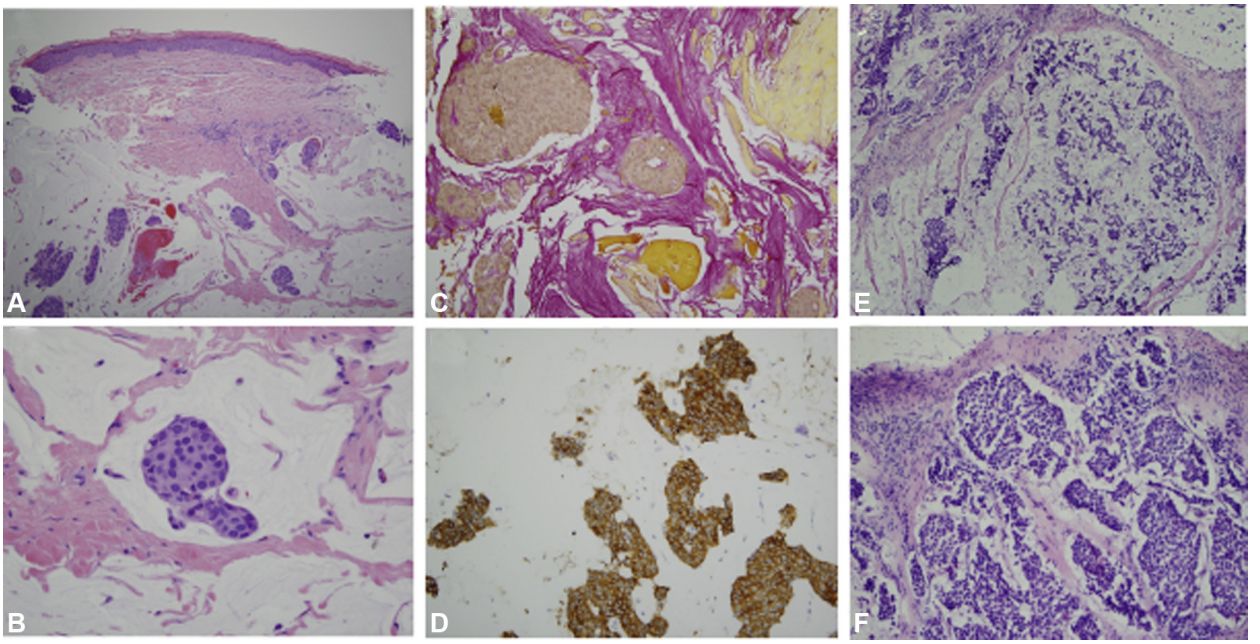

Fig 2. **A** and **B**, Punch biopsy scalp lesion shows monomorphic epithelial cells surrounded by abundant mucin and separated by fibrous septa. **C**, Immunoreactivity for mucicarmine. **D**, Extensive immunoreactivity for synaptophysin, favoring EMPSGC of the scalp. **E** and **F**, Mohs stage I with proliferation of monomorphic small round cells surrounded by mucin.

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