Double trouble: Endocrine mucin-producing sweat gland carcinoma of bilateral temples

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Background/Introduction

- Endocrine mucin-producing sweat gland carcinoma (EMPSGC) is a rare, low-grade cutaneous adnexal carcinoma with predilection for the periorbital skin of women in their seventh decade of life.[1,2]
- EMPSGC is a rare but increasingly recognized neoplasm since being first described by Flieder et al. in 1997 with 190 cases described as of the time of the writing of this case report, most of which were identified in the last five years as diagnostic criteria have been standardized.[1,2,3]
- EMPSGC has been identified as a precursor lesion to neuroendocrine-type mucinous sweat gland carcinoma (MSC) but is frequently mistaken for other common lesions of the skin.
- We report an interesting case of EMPSGC affecting symmetric locations of bilateral temples which, to the authors' knowledge, is the first such documented presentation.

Case Report

A 59 year-old Caucasian woman with no prior oncologic history presented with bilateral flat, skin-colored lesions with post-operative scarring to her temples (Figure 1). She noticed the right-sided lesion 2-3 years ago and the left-sided lesion only within the last year. She had associated mild pain and irritation to the left lesion. Four months prior to presentation, she had undergone curettage excision of the left-sided lesion with no known pathologic diagnosis. She underwent excision of the right-sided lesion two months later at an outside facility with pathology confirming an EMPSGC with positive margins. Upon presentation to Houston Methodist Hospital (HMH), she had recurrence of the bilateral lesions. Radiographic imaging, including MRI face/neck, PET/CT scan, bilateral breast mammogram and ultrasounds, were negative. The patient underwent wide local excision of bilateral temple lesions at HMH. Final pathology confirmed the bilateral temple lesions as EMPSGCs. Due to close margins, presence of focal MSC, and history of recurrent lesions, she underwent adjuvant radiation therapy, and has remained cancer-free on subsequent follow-up and tumor surveillance.

Figures

Bilateral EMPSGC Lesions

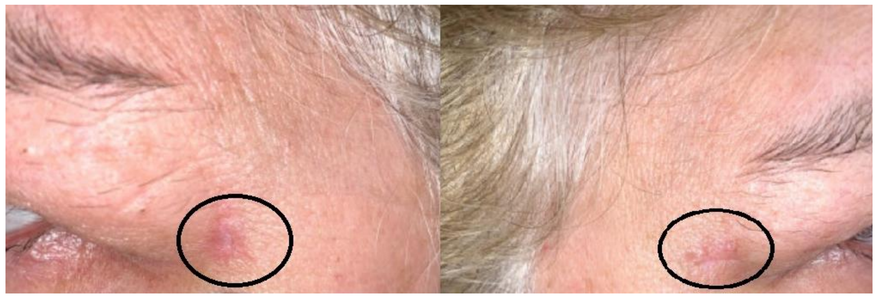

Figure 1 - Images of bilateral temple lesions (circled), status-post outside resections. Left: Left temple lesion. Right: Right temple lesion.

Histopathology

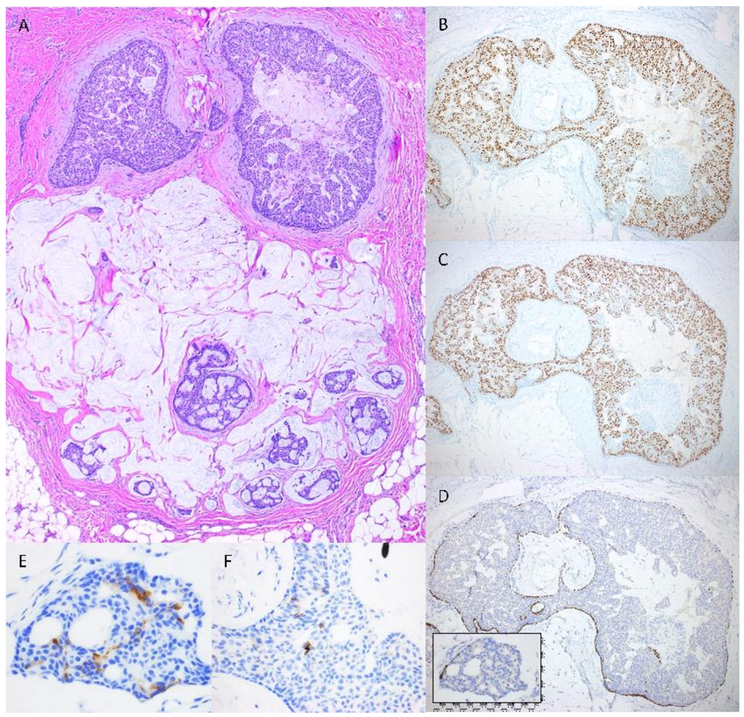

Figure 2 - Morphology and immunoprofile of EMPSGC associated with invasive mucinous adenocarcinoma. A, Low magnification of EMPSGC (top) with associated invasive mucinous carcinoma (bottom), H&E staining. B and C, Strong positive nuclear staining with progesterone and estrogen, respectively. D, P63 staining shows preservation of the myoepithelial rim in the in-situ component and loss of myoepithelial rim around invasive mucinous adenocarcinoma nests (inset). E and F, focal neuroendocrine staining with synaptophysin and CD56, respectively.

Site of EMPSGC	Approximate Proportion of Cases
Eyelids	75-90%
Cheek	7.5-15%
Canthus	3-7%
Scalp	3.5-5%
Periauricular	2-3%
Temple	1%
Occipital	1%

Table 1 – Sites of EMPSGC lesions, adapted from Agni et al.[1] and Au and Bundele[2]; only 2.5% of cases were multicentric

Conclusions

- Classic histopathologic findings are identical to that of papillary carcinoma of the breast, most notably estrogen receptor (ER) and progesterone receptor (PR) positivity.[3]
- Diagnostic workup is necessary to rule out breast cancer as a primary etiology of the lesion prior to definitively diagnosing EMPSGC.
- EMPSCG is an indolent precursor to the more aggressive neuroendocrine type MSC with 33% of EMPSGC containing a component of MSC at the time of diagnosis[1]; early resection of EMPSGC is curative.
- Isolated facial lesions and lesions that metastasize or recur after initial management, such as those described in this case report, are being more frequently identified, necessitating awareness and management by head-and-neck surgeons.[1,4,5]

References

1. Agni M, Raven ML, Bowen RC, Laver NV, Chevez-Barrios P, Milman T, Eberhart CG, Couch S, Bennett DD, Albert DM, Hogan RN, Phelps PO, Stiefel H, Mancera N, Hyrcza M, Wang A, Burris CKH, Steele EA, Campbell AA, Potter HD, Lucarelli MJ. An Update on Endocrine Mucin-producing Sweat Gland Carcinoma: Clinicopathologic Study of 63 Cases and Comparative Analysis. Am J Surg Pathol 2020;44:1005–1016.

2. Au RTM, Bundele MM. Endocrine mucin-producing sweat gland carcinoma and associated primary cutaneous mucinous carcinoma: Review of the literature. J Cutan Pathol. 2021;1–10.

3. Flieder A, Koerner FC, Pilch BZ, Maluf HM. Endocrine Mucin-Producing Sweat Gland Carcinoma: A Cutaneous Neoplasm Analogous to Solid Papillary Carcinoma of Breast. Am J Surg Pathol 1997;21:1501–1506.

4. Nishimoto A, Kuwahara H, Ohashi R, Ansai S. Multicentric endocrine mucin-producing sweat gland carcinoma and mucinous carcinoma of the skin: A case report. J Cutan Pathol. 2020;48:165–170.

5. Froehlich M, Cook J, Bruner E, Stalcup S, Patel K, Day T. Endocrine Mucin-Producing Sweat Gland Carcinoma of the Eyelid With Locoregional Metastasis to the Parotid Gland. Dermatol Surg. 2020;46:1116-1118.