Endocrine mucin-producing sweat gland carcinoma of the cheek

Yuri Katsura[1], Azusa Sasaki[1], Daisuke Minagawa[1], Ikuma Kato[2], Takashi Ueda[1]

Affiliations: [1]Department of Dermatology, National Hospital Organization Yokohama Medical Center, Yokohama, Kanagawa, Japan, [2]Department of Molecular Pathology, Yokohama City University School of Medicine, Yokohama, Kanagawa, Japan

Corresponding Author: T Ueda, 3-60-2 Totsuka-ku Harajuku, Yokohama, Kanagawa 245-8575, Japan, Tel: 81-45-851-2621, Email: ueder@med.kitasato-u.ac.jp

Abstract

Endocrine mucin-producing sweat gland carcinoma (EMPSGC) is a low-grade adnexal malignant neoplasm. We report a 90-year-old man who had a hard, dome-shaped tumor approximately 9 mm in diameter on the left cheek. Dermoscopy showed an overall, non-uniformly light-pink tumor with crust. The diagnosis of EMPSGC is made histologically from excisional biopsy. No signs of recurrent disease were evident at 42 months postoperatively.

Keywords: carcinoma, dermoscopy, endocrine, mucin-producing, non-pigmented basal cell, sweat gland, tumor

Introduction

Endocrine mucin-producing sweat gland carcinoma (EMPSGC) is a low-grade adnexal malignant neoplasm of the skin with neuroendocrine differentiation. The tumor typically presents as slow-growing, flesh-pink, nonspecific nodules occurring on the eyelids or periorbital skin in elderly patients. In this case report, we describe EMPSGC of the cheek in a 90-year-old man.

Case Synopsis

A 90-year-old man presented with a 3-year history of a subcutaneous tumor on the left cheek, which had been gradually increasing in size. He had a medical history of stroke and spinal canal stenosis. He had no oncologic history but did have a history of smoking 10 cigarettes a day for 70 years. On clinical examination, a hard, dome-shaped nodule, approximately 9mm, in diameter with small crust was evident on the left cheek (**Figure 1A**). No lymphadenopathy or metastases were seen on computed tomography. Dermoscopy showed an overall, non-uniformly light-pink tumor with crust and a white partial band (**Figure 1B**). No arborizing vessels were apparent. The differential diagnosis included Merkel cell carcinoma, sebaceous gland carcinoma, non-pigmented basal cell carcinoma (BCC), and amelanotic melanoma.

Excisional biopsy of skin from the tumor showed a well-circumscribed, multinodular, dermal tumor consisting of round-to-oval cells (**Figure 2**). In addition, intracellular mucin was occasionally noted. Tumor cells were positive for synaptophysin, estrogen receptor, and progesterone receptor. In addition, cells were focally positive for CD56, but negative for chromogranin A, CK5/6, CK20, and HER-2. The concentration of carcinoembryonic antigen (CEA) was 4.5ng/mL. Endocrine mucin-producing sweat gland carcinoma was diagnosed, based on the clinical presentation and histopathology. Complete excision was achieved, with negative margins and the patient did not wish to undergo a wider excision. No signs of recurrent disease were evident at 42 months postoperatively.

Case Discussion

The term EMPSGC was first used by Flieder et al. in 1997 [1] who noted that this entity was histologically analogous to solid papillary carcinoma of the breast. Recent case reports have demonstrated that some

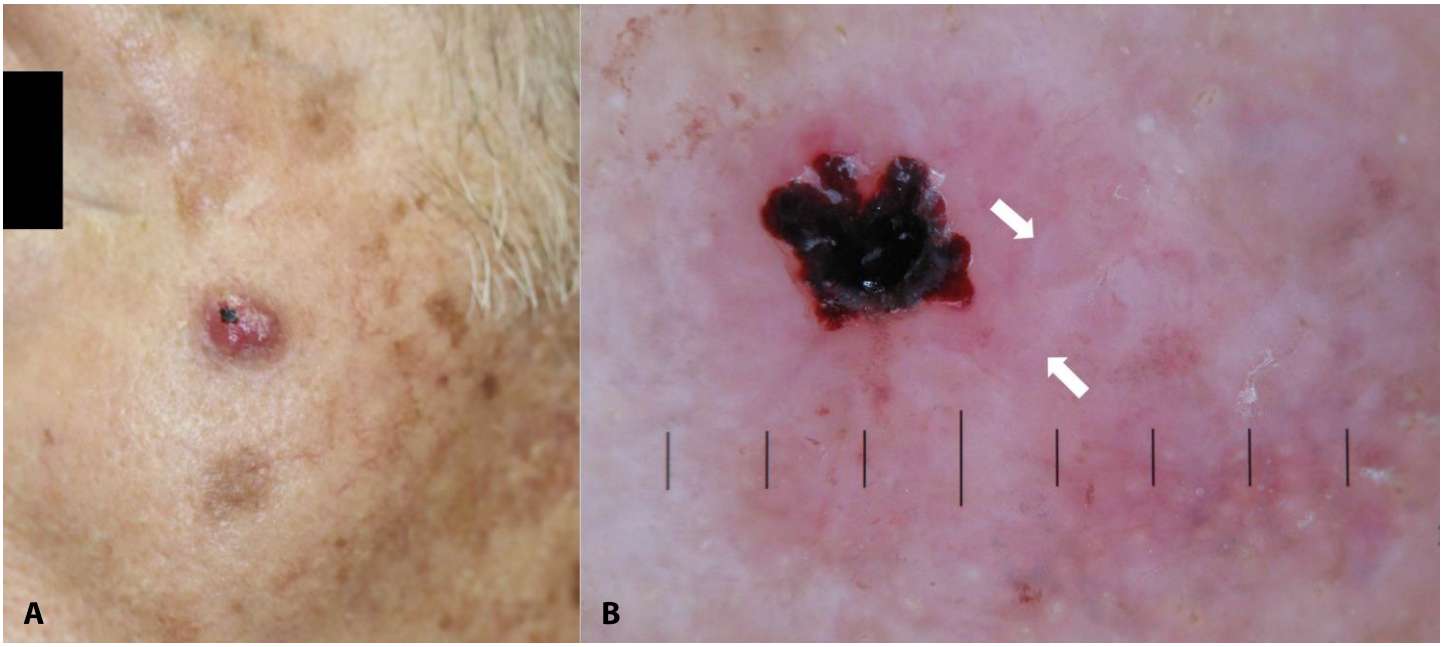

Figure 1. ***A)*** *A reddish, dome-shaped tumor approximately 9 mm in diameter is seen on the left cheek.* ***B)*** *Dermoscopic examinations show light-pink ovoid nests with a white bundle (arrow) and crust.*

cases have been found to co-exist with mucinous carcinoma with which they may be related [2-4].

Most reports have described cases presenting on the eyelid and nearby cheek. The male:female ratio is approximately 1:2. However, Nakamura et al. reported four men with EMPSGC [5] and noted that six of all seven cases in past reports from East-Asian countries were men. Interestingly, gender differences may be seen according to race. Mean age at presentation was 70 years (range, 48–90 years). To the best of our knowledge, this patient with EMPSGC

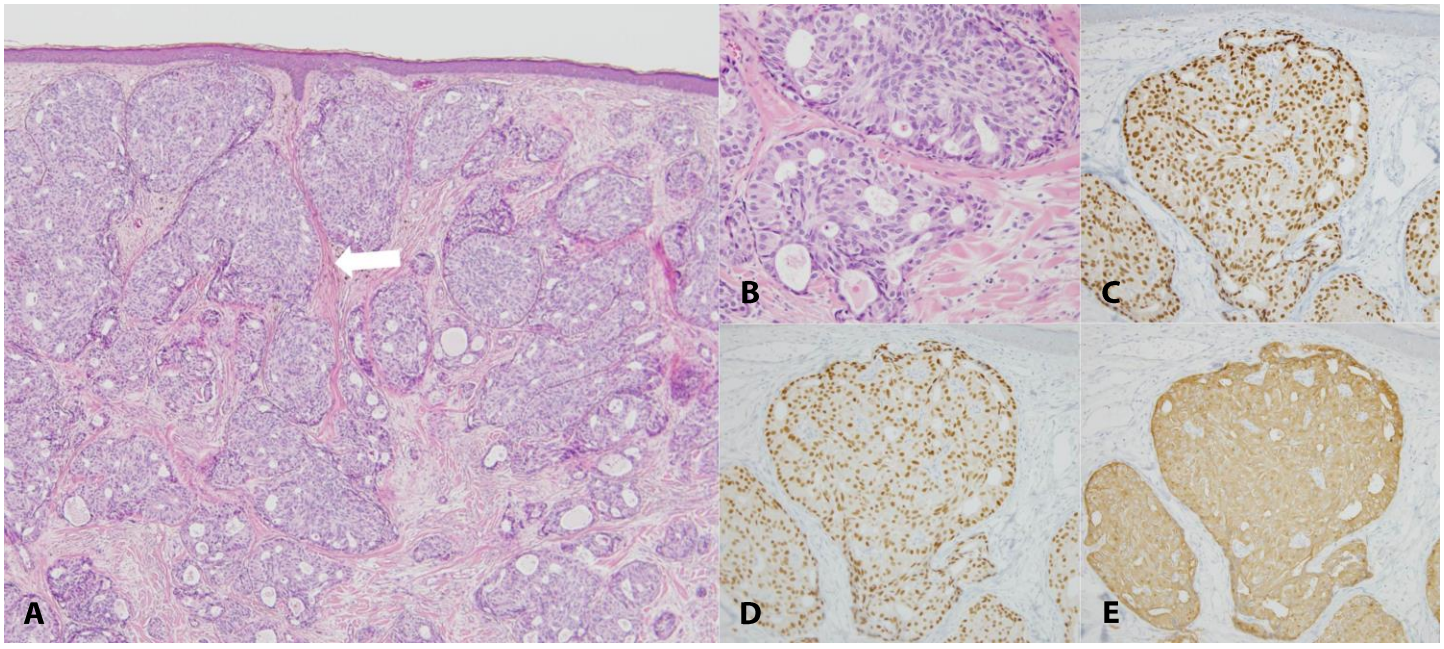

Figure 2. ***A)*** *A multinodular solid dermal tumor is shown. Small, round tumor cells display eosinophilic cytoplasm. Intracellular mucin is occasionally noted. Interstitial fibers appear to correspond to the partial white band in clinical future (arrow). H&E, 100×.* ***B)*** *A palisaded arrangement of tumor cells is seen at the periphery of solid lobules and luminal mucin is shown in nests. H&E, 200×. Almost all tumor cells are positive for* ***C)*** *estrogen receptor,* ***D)*** *progesterone receptor, and* ***E)*** *synaptophysin; all 200×.*

is the oldest yet reported. Complete surgical removal with a margin of at least 5mm is recommended. Sometimes, EMPSGC is treated with Mohs micrographic surgery [6].

Definitive diagnosis of EMPSG requires immunohistochemical staining. Most published cases of EMPSGC have reported positivity with at least one neuroendocrine marker, including synaptophysin (85/92+), chromogranin (55/70+), neuron-specific enolase (41/44+), and CD56 (15/27+). Estrogen (74/74+) and progesterone (51/52+) nuclear stains showed a high rate of positivity. Other staining techniques, GCDFP-15 (41/44+), CK5/6 (1/6+), CK7 (60/60+), CK20 (1/47+), and CEA (17/23+) were described in a few reports [7-9].

Dermoscopic findings of EMPSGC have been rarely discussed. Murakami et al. reported dermoscopic features of EMPSGC in one case [10]. They noted large red/blue globules in pink ovoid nests, with each nest separated by white-to-pink meshes of band (whitish-pink network), which may be a characteristic dermoscopic finding specific to EMPSGC. Those findings reflect lacunae containing secretory fluid with red blood cells in tumors with interstitial fibers. On the other hand, an overall non-uniformly light-pink tumor with crust and a white partial band was seen on dermoscopy in our case. Unlike theirs', our case did not show red/blue globules of fluid with red blood cells in the tumor. When the nests of tumor are small and interstitial fibers are abundant, globules separated by white meshes may be more clearly seen on dermoscopy. Conversely, when the nests are large and interstitial fibers are sparse, the tumor looks like a blurred mass with a partial white band.

Conclusion

We report the EMPSGC of the cheek of an elderly patient. At the wishes of the patient the tumor was excised completely with negative margins, and he remained symptom free postoperatively for 42 months.

Potential conflicts of interest

The authors declare no conflicts of interest.

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