Delayed treatment of endocrine mucin–producing sweat gland carcinoma initially diagnosed as a chalazion

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Key words: benign lesion; chalazion; chromogranin A; CK7; clinical suspicion; cutaneous malignancy; delayed treatment; endocrine-producing mucin sweat gland carcinoma; eyelid cancer; eyelid lesion; eyelid margin; malignant lesion; metastasis; misdiagnosis; Mohs; oculoplastic reconstruction; recurrence; synaptophysin.

INTRODUCTION

Endocrine mucin–producing sweat gland carcinoma (EMPSGC) is a slow-growing, rare variant of mucinous carcinoma most commonly presenting on the eyelid.[1,2] Its rarity, inflammatory characteristics, and slow-growing nature all increase the risk of misdiagnosis.[2] We present a case of a EMPSGC initially diagnosed as a chalazion. Conservative treatment was attempted on multiple occasions and over an extended period, despite a lack of clinical response. This treatment caused a delay in diagnosis and appropriate treatment for this rare malignant lesion. Our case illustrates EMPSGC as a lesion that can masquerade as a chalazion or hordeolum.

Abbreviation used:

EMPSGC: endocrine mucin–producing sweat gland carcinoma

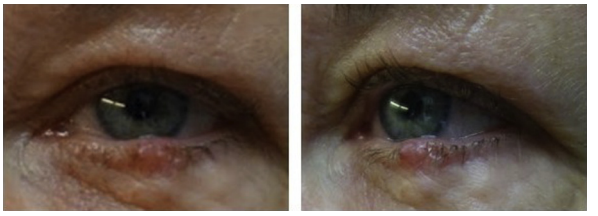

Fig 1. Clinical appearance of the lesion at presentation to principal author.

CASE

We present the case of a 64-year-old woman with a 1.5-year history of a raised inflamed lesion of the left lower eyelid margin. The lesion was initially diagnosed and managed by her dermatologist as a chalazion. The presumed chalazion was treated with multiple triamcinolone acetonide injections. It slightly decreased in size, however, months later continued to progress prompting the treating physician to utilize another series of injections with triamcinolone acetonide. Over a year, the lesion was injected with triamcinolone acetonide 5 times with minimal overall improvement. The patient was then referred to the principal author to treat this nonresolving lesion. Slit lamp examination found several features concerning for malignancy: irregular borders, central ulceration, and destruction of lid margin architecture and lash follicles (Fig 1). Because of these concerning features and the prior poor response to conservative treatment, a biopsy was performed to assess for cutaneous malignancy.

The histopathologic section exhibited a dome-shaped biopsy of skin with pools of mucin with islands of epithelial cells containing cuboidal and glandular spaces within (Fig 2, *A*), resulting in an initial diagnosis of mucinous carcinoma. Further immunohistochemistry stains were positive for chromogranin A (Fig 2, *B*), CK7 (Fig 2, *C*), and synaptophysin (Fig 2, *D*) and negative for CK20, CDX2, and TTF1, further specifying the diagnosis as EMPSGC, a rare tumor variant of mucinous carcinoma.

From the Skin Cancer and Reconstructive Surgery Center.

Funding sources: Skin Cancer and Reconstructive Surgery Foundation.

Conflicts of interest: Dr Joseph receives honoraria from Allergan, Evolus, and Inmode Aesthetics. Ms Meltzer has no conflicts to disclose.

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JAAD Case Reports 2019;5:789-91.
2352-5126

https://doi.org/10.1016/j.jdcr.2019.06.031

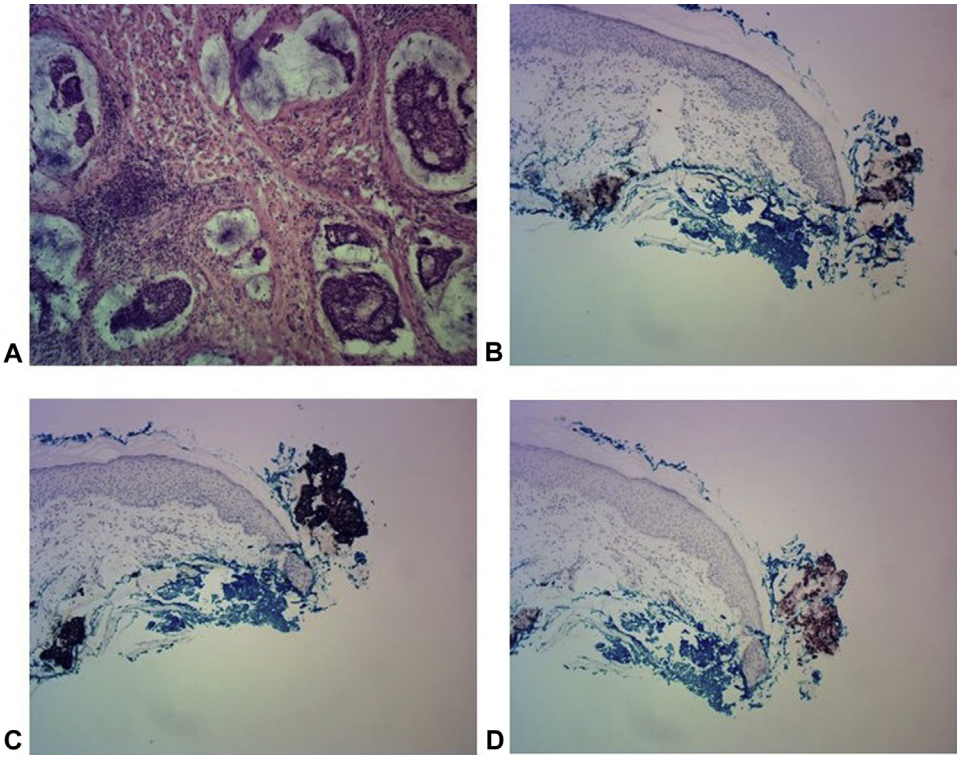

Fig 2. **A**, Dome-shaped cells with pools of mucin. **B**, Positive staining for chromogranin A. **C**, Positive staining for CK7. **D**, Positive staining for synaptophysin. (Original magnifications: **A-D**, ×40.)

The lesion was then excised utilizing Mohs micrographic surgery until clear surgical margins were obtained, resulting in a defect size 2.2 × 0.8 cm that extended to the subcutis (Fig 3). The principal author reconstructed the defect with a tarsoconjunctival flap and cutaneous advancement flap. There was no observed recurrence after 6 months of postoperative examinations.

The collected information and analysis of protected patient health information was HIPAA compliant, and informed written consent was obtained. The appropriate treatment was delayed, in that the nonsurgical treatment of the presumed chalazion may have blunted the immune response and thereby delayed the clinical presentation of a papular nodule. If left untreated, it is at risk for metastasis.[2]

DISCUSSION

EMPSGC is a rare, often misdiagnosed, lesion that when occurring on the eyelid, may be mistaken for common inflammatory lesions such as chalazia. Ozdal et al[3] performed a retrospective evaluation of all clinical diagnoses of chalazia from 1993 to 2001 that were submitted to the Henry C Witelson Ophthalmic Pathology Laboratory and Registration. Of the 6.4% of presumed chalazia misdiagnosed, over 1% of these lesions were malignant or premalignant, including sebaceous cell carcinoma, basal cell carcinoma, and chronic inflammation with cellular atypia and mitotic figures.[3] Other studies found malignancy in up to 4.6% of clinically suspected benign eyelid lesions. Although mucinous carcinoma and EMPSGC were not included in Ozdal's analysis, we know EMPSGC has a high risk of misdiagnosis because of its rarity. Qin et al[4] performed a case review of 11 cases of EMPSGC all in the eyelid. Of the 11 cases, 7 were interpreted as either chalazion or nonspecific inflamed lesions of the eyelid. Inflamed lesions most commonly represent chalazia or acute hordeola.[4] At first, this patient presented with a clinical appearance similar to that of a chalazion, prompting its misdiagnosis and anti-inflammatory treatment over an extended period. We hypothesize that blunting of the inflammatory component of this malignancy lesion likely led to delay in definitive treatment. This delay in referral and treatment is particularly important with EMPSGC, as there is a high rate of metastasis when compared with more common eyelid malignancies such as basal cell carcinoma.

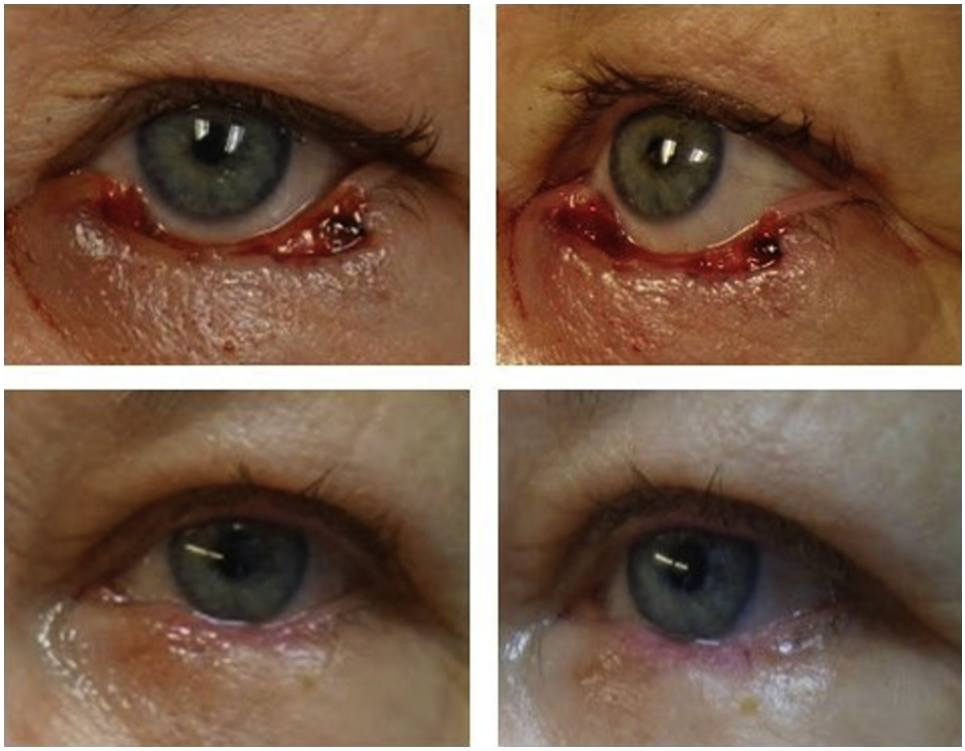

Fig 3. Final Mohs defect size of 2.2 × 0.8 cm and aesthetic result 2 months postoperatively.

Although it is not possible to escape misdiagnosis completely, this case study reaffirms the common teaching that if a presumed inflammatory lesion of the eyelid margin does not respond as expected with standard treatment, then biopsy and histopathologic evaluation are necessary to assess for malignancy. EMPSGC, although rare, must always be considered, as we show with this case that it can share many similar features with chalazia, and the eyelid is the most common location for EMPGSC.[2]

We thank Dr Ronald Barr for assistance with dermatopathologic descriptions and images in this report.

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