JAMA Ophthalmology Clinical Challenge

# Cystic-Appearing Eyelid Lesion in a 62-Year-Old Man

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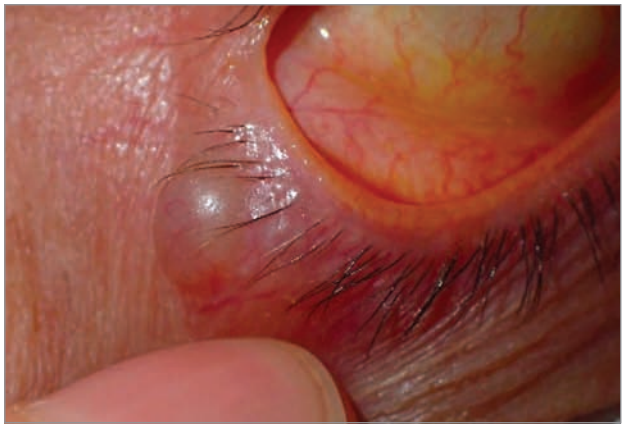
A Cystic-appearing subcutaneous nodule



B Micropapillae of tumor cells

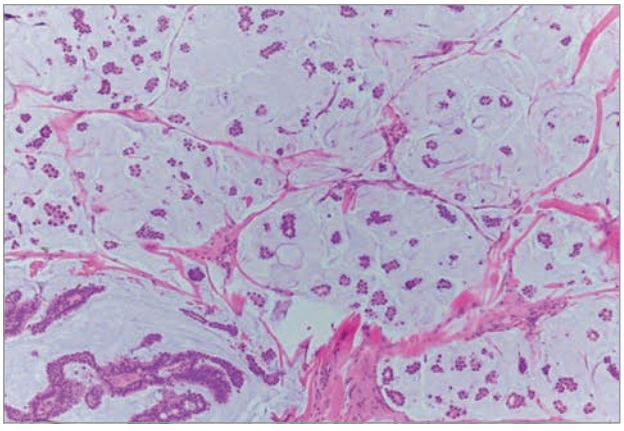


**Figure 1.** Cystic-appearing subcutaneous nodule with a few overlying telangiectasias measuring approximately 1 cm identified just below the eyelash line (A). Histopathology revealed micropapillae of tumor cells floating within pools of mucin (hematoxylin-eosin, original magnification × 10) (B).

**A 62-year-old male** was referred for evaluation of a painless right lower-eyelid lesion noted during routine glaucoma follow-up. The lesion had been present for 3 years with slow, gradual enlargement. The patient denied any associated symptoms. He was a former smoker and had no history of skin cancer or immunosuppression. The remainder of his medical history was noncontributory.

Inspection of the right lower eyelid revealed a cystic-appearing nodule that transilluminated and had a few overlying telangiectasias. The cystic portion was continuous with a deeper, smooth subcutaneous component that was well defined, firm, and nonmobile, with a total lesion length of 1 cm (**Figure 1**A). There was no madarosis, eyelid margin effacement or preauricular lymphadenopathy. Although no overt features of malignancy were present, the deeper component was atypical for a hidrocystoma and an excisional biopsy was performed. Histopathologic evaluation showed a neoplasm composed of low-grade round nuclei growing in a micropapillary pattern within pools of extracellular mucin (Figure 1B). There was tumor present at the margin. Immunostains were positive for estrogen receptor and progesterone receptor. Variable staining for INSM1, a sensitive nuclear stain for neuroendocrine differentiation, and focal staining for synaptophysin, another neuroendocrine marker, were also seen. This immunophenotype is consistent with a primary tumor derived from endocrine mucin-producing sweat gland carcinoma or a metastatic mucinous carcinoma of breast origin.

**WHAT WOULD YOU DO NEXT?**

**A.** Wide local excision

**B.** Excision with frozen sections

**C.** Mohs surgery

**D.** Metastatic workup

CME Quiz at jamacmelookup.com

## Diagnosis

**Mucinous carcinoma of the eyelid**

## What to Do Next

**D.** Metastatic workup

## Discussion

Cutaneous mucinous carcinoma is a rare, low-grade malignant neoplasm of the sweat glands. It often presents as a slow-growing asymptomatic nodule, most commonly on the eyelid and the face.[1] The histogenesis is disputed; while previously thought to be of eccrine origin, recent evidence suggests it demonstrates apocrine-type differentiation.[2] Particularly on the eyelid, it may arise from endocrine mucin-producing sweat gland carcinoma. Due to its low incidence, literature on this disease is limited.

Most commonly, cutaneous mucinous carcinoma presents as a primary lesion but 5% to 6% are metastatic in origin.[1,3] In metastatic disease, common sites of tumor origin include the breast and gastrointestinal tract. Thus, once a diagnosis of mucinous carcinoma is made, it is critical to evaluate for metastatic disease with full-body imaging (choice D).[4,5] After metastatic workup is completed, a treatment plan can be determined. In this case, wide local resection (choice A) is not the preferred next step as systemic disease should first be ruled out. Similarly, excision with frozen sections

(choice B) would not be recommended before evaluating for metastatic disease. Mohs surgery (choice C) would also not be preferred until a systemic workup is complete. After ruling out systemic disease, treatment of mucinous carcinoma is complete excision. Evidence suggests a lower rate of recurrence in patients undergoing excision with frozen section margins or Mohs surgery than wide local excision, as the extent of the tumor can be difficult to gauge clinically due to its infiltrative nature and the grossly normal appearance and texture of involved tissues.[6]

The incidence of distant metastasis in cutaneous mucinous carcinoma is significantly higher in eyelid tumors than other primary anatomic sites.[1,3] In addition, eyelid involvement has been shown to have higher rates of recurrence of up to 30% to 40%.[6-8] In a large population-based study of 411 cases, there were 10 deaths reported due to primary cutaneous mucinous carcinoma; however, only 1 eyelid tumor was associated with death.[3] Because of the high recurrence rate and possibly devastating outcomes, patients with primary cutaneous mucinous carcinoma should be monitored closely. Given the benign appearance of the tumor, ophthalmologists should take extra care when evaluating eyelid lesions with cystic features to not miss disease with metastatic potential. Fortunately, despite the higher incidence of distant metastasis and recurrence, eyelid tumors have not been found to be associated with worse survival.[3]

## Patient Outcome

After the initial biopsy confirmed mucinous carcinoma with positive margins, the patient underwent metastatic workup with a computed tomography of the neck, chest, abdomen, and pelvis, which were unrevealing. He then underwent a full-thickness wedge resection of the entire visibly involved area with additional 2-mm margins of normal-appearing tissue medially, laterally, and inferiorly. These specimens were sent for rush permanent pathology. Surprisingly, all the margins returned positive. The patient returned for excision with frozen sections. In total, 22 frozen section specimens were needed to achieve negative margins, leaving a large lower-eyelid defect (**Figure 2**). The patient ultimately underwent reconstruction and has been disease free since.

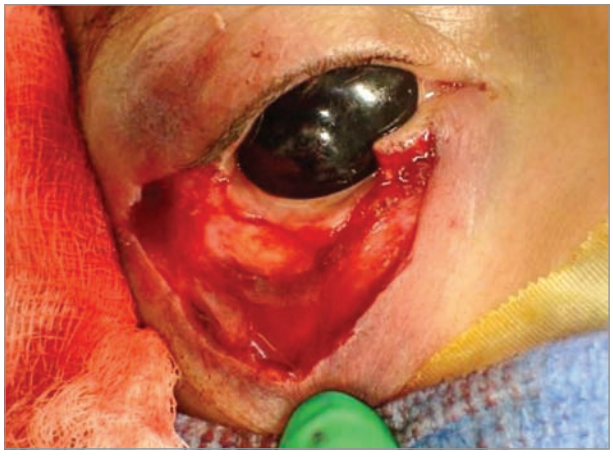


**Figure 2.** Large right lower-eyelid defect (5.0 cm × 2.5 cm) after achieving negative margins.


### ARTICLE INFORMATION

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