Gazi Tıp Dergisi / Gazi Medical Journal

2005: Cilt 16: Sayı 2: 88-89

OLGU SUNUMU - CASE REPORT

PRIMARY MUCINOUS ADENOCARCINOMA OF THE EYELID

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Primary mucinous adenocarcinoma of the eyelid is an uncommon malignant cutaneous tumor. A firm, slow-growing cystic mass, which had been noticed two years previously, developed in a 54-year-old man's right lower eyelid. Full-thickness eyelid excision of the tumor with tumor-free edges was performed, and no recurrence occurred for more than 2 years. We present this unusual lesion to emphasize that all eyelid nodules, including benign-appearing ones, should be biopsied to exclude malignancy such as primary mucinous adenocarcinoma.

Key Words: Eyelid, Mucinous Adenocorcinoma, Tumor

GÖZ KAPAĞININ PRIMER MÜSINÖZ ADENOKARSINOMASI

Göz kapağının primer müsinöz adenokarsinoması nadir görülen bir malign deri tümörüdür. Sağ alt göz kapağında sert, yavaş büyüyen kitle şikayeti ile başvuran 54 yaşında hastaya cerrahi sınırlarda tümör olmayacak şekilde tam kalınlıkta eksizyon uygulandı. Primer müsinöz adenokarsinoma tanısı ile 2 yıl izlenen hastada herhangi bir nüks rastlanmadı. Bu vaka sunumundaki amacımız, benign görünümlü göz kapağı nodülleri de primer müsinöz adenokarsinoma gibi malign lezyonlardan ayırıcı tanısının yapılabilmesi için biyopsi ile değerlendirilmesi gerektiğini vurgulamaktır.

Anahtar Kelimeler: Göz Kapağı, Müsinöz Adenokarsinoma, Tümor

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Primary mucinous adenocarcinoma of the skin is a rare malignant eccrine sweat gland tumor of the head, neck and trunk.1 In the face it usually occurs in the lower eyelid of elderly people. It was first described by Lennox in 1952.2 To date fewer than 100 cases have been reported. This tumor has been reported in the ophthalmology and dermatology literature, but not to our knowledge in the plastic surgery literature.

Typically, a patient with a primary mucinous carcinoma of the skin usually has a long-standing history since the lesions are painless and slow-growing and are ignored by the patients. While rarely causing death, recurrence following primary excision is common and widespread metastases may occur. We present this unusual malignant lesion that in this case has no recurrence.

CASE REPORT

A 54-year-old man presented with a 0.9 × 1 cm painless, firm lesion, cystic in appearance, situated over the lateral third of his right lower eyelid (Fig. 1). He had first noticed the lesion 2 years before and it had grown slowly. On clinical evaluation, firm swelling of the inferior was noted without any erythema or diplopia. It had a rubbery consistency, transilluminated well, and extended to the gray-line margin without invasion of the conjunctival surface. Visual acuity was normal. The lesion was removed by full-thickness eyelid resection followed by direct approximation of the eyelid margins. The tissue specimen was thoroughly examined by a dermatopathologist, and the diagnosis was mucinous adenocarcinoma. Microscopic images were strikingly characteristic with cords and nests of small basaloid cell proliferations embedded in mucin pools, those separated by fibrous septae (Fig. 2). The mucin found was consistent with sialomucin and stained with PAS/Alcian blue pH:2.5 histochemistry. The margins of the excision specimen were free of tumor. A thorough workup with upper gastrointestinal series, abdominal and pelvic computerized tomography scan, and full colonoscopy with biopsies revealed no occult primary tumor.

The lower eyelid was reconstructed using a switch-flap from the upper lid. The surgery was successful, and a good functional and cosmetic result was obtained. The patient had no recurrence during the 2-year follow-up.

DISCUSSION

Carcinomas of the skin are the most common malignancies, but eccrine-derived neoplasms, such as mucinous adenocarcinoma, are rare. Mucinous adenocarcinoma was described earlier,1 and it was first formally described and reviewed by Mendoza and Helwig in 1971.3 In their reports, 14 cases were reported on the face, head, or trunk. The patient age at presentation ranged from 8

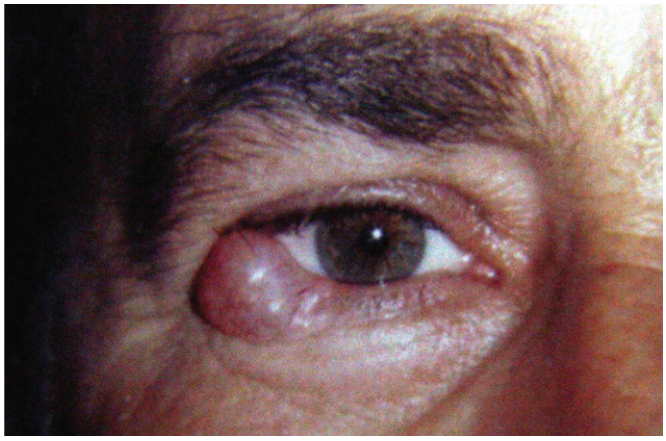

Fig. 1: Painless, firm, cystic mass at the lateral margin of the right lower eyelid.

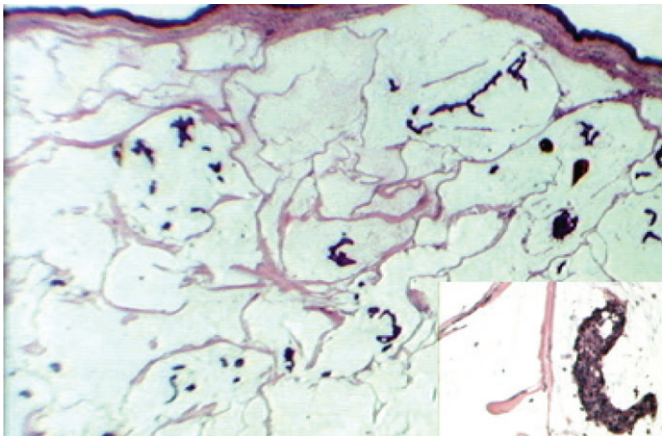

Fig. 2: Mucinous adenocarcinoma with characteristic cords and nests of basaloid cells embedded in pools of mucin separated by thin fibrous septae.

to 76 years. While six of their lesions recurred following excision, there was only one case of metastasis, and seven cases of favorable prognosis. Wright and Font 4 reported the first series of mucinous adenocarcinoma of the eyelid with 40% recurrence in 21 cases. One death due to local extension was also described in this report. Bilateral primary mucinous carcinoma of the eyelid was reported by Bertagnolli et al. Two other cases of primary mucinous adenocarcinoma of the upper eyelid 5,6 and one of the lower eyelid 7 were reported with no recurrence during the follow-up. Another two cases of primary adenocarcinoma of the eyelid were reported with deep extension that needed exenteration 8 or en bloc removal of the aggressively invading tumor including the orbit, ethmoid and sphenoid sinuses.9 Recurrence of insufficiently resected primary mucinous adenocarcinoma was also reported.

The preoperative diagnosis of these tumors is difficult because they do not have any pathognomonic clinical appearance that is bluish, pink or red, and range from firm to indurated to cystic. They are usually asymptomatic, and present as a dome-shaped firm mass, often ignored by the patient for some time1. They can also present as a subcutaneous mass. They have to be differentiated from basal cell carcinoma, papilloma, keratoacanthoma, Kaposi's sarcoma, hemangioma and pyogenic granuloma.10 The diagnosis of primary mucinous carcinoma of the skin requires the exclusion of metastatic mucinous carcinoma, particularly from the gastrointestinal tract, salivary glands, lacrimal glands, thyroid gland, lungs or breast.11 In fact, the incidence of metastatic mucinous carcinoma is much greater than that of primary mucinous carcinoma. For this reason, a meticulous systemic evaluation should be performed to exclude a distant primary mucinous tumor. In our patient, we decided on the diagnosis of primary tumor after a detailed systemic evaluation, especially of the gastrointestinal system.

Complete tumor extirpation is sufficient to treat these lesions. Since the range of healthy skin margins of the excised periorbital tumors is limited, recurrent periorbital tumors are difficult to treat. As a result, any conventional excisional biopsy technique or Mohs surgery is used and the tumor-free margins around the tumor should be reached. A favorable prognosis is expected if the lesion is totally removed at an early stage without any metastasis.

In conclusion, we emphasize that with all eyelid nodules, including benign-appearing ones, physicians should keep in mind primary mucinous adenocarcinoma of the eyelid.

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