

Primary periocular mucinous eccrine carcinoma : a case report.

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ABSTRACT:

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Mucinous eccrine carcinoma of the skin is a rare malignant sweat gland neoplasm of the head, neck and trunk. It may be diagnosed as a metastasis tumor or, which is highly unlikely, as a primary focus. The clinical diagnosis of primary mucinous eccrine carcinoma from other common tumors in the eyelid area may be difficult because this tumor does not have a pathognomonic clinical appearance, and is always asymptomatic. It can be presented as a painless, slow growing mass and is usually ignored by the patient. The essential treatment for primary mucinous eccrine carcinoma is complete surgical excision of this lesion. However, a histopathological examination should be performed first, and a systemic work up should be done to rule out the possibility of a primary tumor elsewhere. We report a patient who presented with primary mucinous adenocarcinoma of the eyelid at Institute of Dermatology and highlight their presentation, clinical course and therapeutic approach.

Key words: Mucinous eccrine carcinoma, eyelids tumor

บทคัดย่อ :

พีรพงษ์ สุดสงวน, ประภาวรรณ เขาวะวณิช รายงานผู้ป่วยโรค MUCINOUS ECCRINE CARCINOMA วารสารโรคผิวหนัง 2553; 26: 219-222.

สถาบันโรคผิวหนัง กรมการแพทย์ กระทรวงสาธารณสุข

Mucinous eccrine carcinoma เป็นเนื้องอกของผิวหนังที่มักพบบริเวณ ศีรษะ คอ และลำตัว ซึ่งการวินิจฉัยเนื้องอกชนิดนี้จากลักษณะทางคลินิกที่พบค่อนข้างทำได้ยาก เพราะเนื้องอกชนิดนี้ไม่มีลักษณะเฉพาะทางคลินิกที่ช่วยการวินิจฉัย และเนื้องอกชนิดนี้มักไม่มีอาการ โดยก้อนเนื้องอกจะโตขึ้นช้าๆ ไม่เจ็บจะมักถูกละเลยโดยผู้ป่วย การรักษาจำเป็นต้องผ่าตัดเนื้องอกออกทั้งหมดและตรวจเพิ่มเติมโดยละเอียดว่าเนื้องอกนี้กระจายมาจากอวัยวะระบบอื่นหรือไม่ รายงานฉบับนี้เป็นการนำเสนอผู้ป่วยชาย อายุ 43 ปี มาตรวจรักษาด้วยก้อนเนื้อขนาดเล็กบริเวณใต้ตาขวา ไม่มีอาการใดๆ ผลตรวจทางจุลพยาธิพบเป็น mucinous eccrine carcinoma

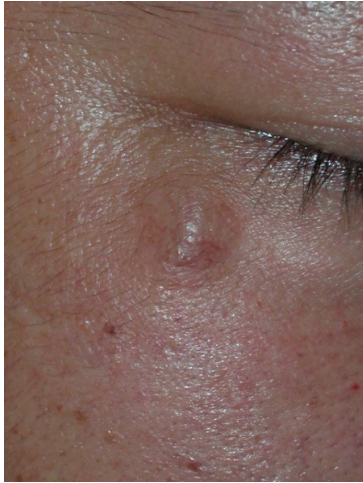
คำสำคัญ : เนื้องอกใต้เปลือกตา

Case report

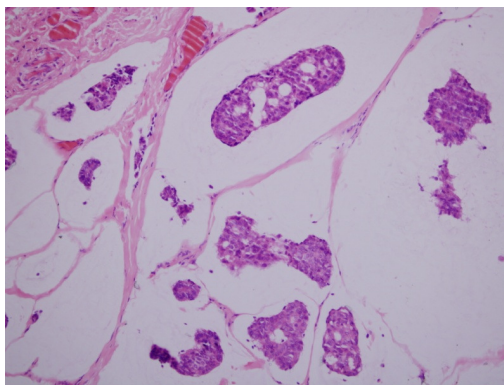
A 43-year old Thai male patient presented with a 6 month slow growing mass on lower the right eyelid. An examination revealed a 10 mm x 10 mm, painless, firm and fixed mass on the lateral third of the lower eyelid. (Figures 1) In order to obtain a definite diagnosis of the patient, an excisional biopsy was performed. The tissue pathology report indicated that it was a mucinous carcinoma. Consultation to rule out the metastasis and look for the primary origin in the salivary and thyroid glands, lungs, breast and gastrointestinal tract by internist, otolaryngologist and plastic surgeon, revealed negative results. The mass was excised.

A histopathological examination of the lesion showed the characteristic appearance of mucinous carcinoma as a primary malignancy. The tumor was

divided into numerous compartments by strands of fibrous tissue. Each compartment showed an abundance of pale staining mucin, surrounded with nests or cords of epithelial islands that also showed slight nuclear pleomorphism. (Figures 2) The mucin showed strong positive reactions to mucicarmin and Periodic acid-Schiff stain and resistance to diastase digestion. Some of the areas also showed solid epithelial lobules, and the surgical margin of the lesion was adequately excised. After ten months follow up, the patient has no recurrence of the mucinous carcinoma from any site, and has no new lesion. No lymph node metastasis and no occult primary malignancy was found.



Figures 1 Fixed mass on the lateral third of the lower eyelid.



Figures 2 Divided into numerous compartments by strands of fibrous tissue.

Discussion

Primary mucinous adenocarcinoma, arising from the eccrine sweat gland of the eyelid, is a very rare tumor. It was first described by Lennox in 1952⁽¹⁾. Fewer than 100 cases of primary mucinous adenocarcinoma have been reported in the English-language literature and about 20 cases in the Japanese-language literature⁽¹⁻⁷⁾. According to

cases in the English literature, primary mucinous adenocarcinoma of the skin primarily affects patients with an age range of 8 to 84 years (median 63 years). The male to female ratio is approximately 2:1. The racial distribution is 67% Caucasian, 32% Negroid, and 4% Asian. In most cases, the duration of disease varies from several months to 5 years (median 1.3 years). The eye, face, and scalp are involved in nearly 75% of cases, the local recurrence rate is approximately 30.1%, regional metastasis 11%, and distant metastasis 2.7%.²⁻³ They are usually asymptomatic and solitary, and present as a dome-shaped firm mass which often ignored by the patient for sometime⁽⁴⁾. Since this tumor has no pathognomonic appearance, tissue histopathology is needed to confirm diagnosis. The histology of a mucinous carcinoma is characterized by the presence of small epithelial islands of neoplastic cells, arranged in a duct-like structure, in a large pool of mucin (presumably sialomucin). Because the primary tumor in the periocular region is very rare, the diagnosis of primary mucinous carcinoma of the skin requires an exclusion of metastasis mucinous carcinoma, particularly from the salivary gland, thyroid gland, breast, lungs, upper and lower gastrointestinal tract. Primary mucous-secreting tumors on these sites can present with a solitary skin metastasis. Proper management in periorbital mucinous adenocarcinoma is a complete tumor extirpation. Commonly, a wide excision or Mohs' surgery is recommended, since conventional excision has a high incidence of local

recurrence that may occur after many years⁽³⁾. In our case, the patient presented with a single, painless, slow growing mass at the left lower eyelid for 2 years, which was similar to the clinical characteristics of the primary mucinous carcinoma described in previous reports. In our case, we performed an excisional biopsy. After surgery, the patient has a tumor free period for more than 10 months now. We suggested that the primary mucinous carcinoma should be one of the differential diagnoses of the eyelid tumor.

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