Case report

PRIMARY MUCINOUS ADENOCARCINOMA OF THE EYELID: A CASE REPORT

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Abstract This study reported a rare case of primary mucinous adenocarcinoma involving the eyelid. We examined a 58-year-old man with a history of gradually enlarging mass in the lateral third of the left lower eyelid. Surgical excision of mass and histopathology studies were performed. Histopathologic and systemic evaluation revealed a primary mucinous adenocarcinoma. No evidence of recurrence or metastasis was noted 36 months postoperatively. Primary mucinous adenocarcinoma is an uncommon tumor of the skin that should be added to the differential diagnosis of eyelid tumors. Since it is a very rare condition, complete physical examination and investigations need to be done carefully in order to rule out the primary from metastatic tumor. Chiang Mai Med Bull 2004; 43(3):127-131.

Keywords: Eyelid tumor, eyelid mass, mucinous adenocarcinoma

Mucinous adenocarcinoma 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 adenocarcinoma 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 adenocarcinoma is complete surgical excision of this lesion. However, a histopathological examination should be performed first, and a systemic work-up completed to rule out the possibility of a primary tumor elsewhere. This would in order to confirm the diagnosis prior to undertaking the excision.

We report a patient who presented with primary mucinous adenocarcinoma of the eyelid at Maharaj Nakorn Chiang
Mai Hospital and highlight their presentation, clinical course and therapeutic approach.

**Case report**

A 58-year old Thai male patient presented with a two year slow growing mass at the left eyelid. An examination revealed a 5 mm x 7 mm, painless, firm and fixed mass situated over the lateral third of the lower eyelid (Fig 1). It also showed a visual acuity of 6/6 OU. The other anterior and posterior segment examinations were conducted within normal limits.

In order to obtain a definite diagnosis of the patient, an incisional biopsy was performed. The resultant tissue pathology report indicated that he had a mucinous type of adenocarcinoma. To rule out any metastasis – and look for the primary origin in the salivary and thyroid glands, lungs, breast and gastrointestinal tract – internal medicine, otolaryngologist and plastic surgeon consultants undertook examinations that gave negative results. The mass was then completely excised using frozen section control, and the eyelid reconstructed by the Tenzel's flap technique. No intraoperative complication was found afterwards.

A histopathological examination of the lesion showed that the characteristic appearance of mucinous adenocarcinoma supports primary malignancy. The tumor was seen to divide 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 (Fig. 2). The mucin showed

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**Figure 1.** Clinical findings show 5 x 7 mm mass at left lower eyelid.
strong positive reactions to mucicarmine 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 then considered adequate, with the postoperative result being good.

After three years of examination, the patient has shown no recurrence of the mucinous adenocarcinoma from either site, nor have any new lesion been identified. No lymph node metastases has occurred, and no occult primary has been found.

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 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 ages range of 8-84 years (median 63 years). The male-to-female ratio is approximately 2:1. The racial distribution is 67% white, 32% black, 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

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**Figure 2.** The histopathology of the tumor shows epithelial islands floating in a large pool of mucin separated by thin fibrovascular septa in the dermis layer. “Adenoid cystic” pattern is seen in most of the tumor. (Hematoxylin-eosin stain, x 40)
They are usually asymptomatic and solitary, and present as a dome-shaped firm mass, often ignored by the patient for some time. Since this tumor has no pathogenomonic appearance, tissue histopathology may contribute to confirmation of the diagnosis. The histology of a mucinous adenocarcinoma 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 adenocarcinoma of the skin requires the exclusion of metastasis mucinous adenocarcinoma, particularly from the salivary gland, thyroid gland, breast, lungs, and upper and lower gastrointestinal tract. Primary mucus secreting tumors on these sites can present with a solitary skin metastasis.

Proper management in periorbital mucinous adenocarcinoma is 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. In our study, 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 adenocarcinoma described in previous reports. In our case, we performed a wide excision with frozen section control. The patient was followed up as tumor free for more than 3 years after surgery.

We proposed that wide excision with frozen section control should be considered as a treatment option for patients with primary mucinous adenocarcinoma, and suggest consideration of primary mucinous adenocarcinoma in the differential diagnosis of the eyelid tumor.

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References

รายงานผู้ป่วยมะเร็งปฐมภูมิชนิด MUCINOUS ADENOCARCINOMA บริเวณเปลือกตาที่พบในจังหวัดเชียงใหม่

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บทคัดย่อ มะเร็งปฐมภูมิชนิด mucinous adenocarcinoma ที่เปลือกตาเป็นเนื้องอกของเปลือกตาที่พบได้น้อยมาก ผู้เขียนได้รายงานโรคมะเร็งปฐมภูมิชนิด mucinous adenocarcinoma ในผู้ป่วยชายไทยอายุ 58 ปีที่มีตัวเรื่องเกิดขึ้นที่บริเวณเปลือกตาล่างด้านซ้าย ผู้ป่วยได้รับการตรวจร่างกายและตรวจร่างกายท้องปฏิบัติการโดยละเอียดไม่พบว่ามีการกระจายของมะเร็งมาจากบริเวณอื่น ผู้ป่วยได้รับการรักษาโดยการผ่าตัดเอ็นเก่าออกหมด แม้ว่าโรคนี้จะพบได้น้อยมากแต่เป็นโรคหนึ่งที่ควรระวังวินิจฉัยต่างโรคภูมิคุ้มกันที่เกี่ยวข้องกับการเปลือกตา การตรวจร่างกายและตรวจทางท้องปฏิบัติการโดยละเอียดมีความจำเป็นในการช่วยแยกภาวะมะเร็งปฐมภูมิจากมะเร็งที่กระจายมาจากบริเวณอื่น เชิงการวิจัย 2547;43(3):127-131.

คําสําคัญ: มะเร็งปฐมภูมิของเปลือกตา ผู้ป่วยมะเร็งปฐมภูมิชนิด mucinous adenocarcinoma