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Case Report

Primary mucinous adenocarcinoma of the eyelid: A case-series☆

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ABSTRACT

Purpose: Primary mucinous adenocarcinomas (PMAs) of the eyelid are rare clinical entities typically arising from the peri-orbital area. The purpose of this case-series is to report 3 cases of PMA as well as to discuss the pathological and immunohistochemical features of these tumors.
Material & methods: Three cases of PMA of the eyelid were identified from 2 tertiary ophthalmology referral centers. Clinical and histopathological features of the cases were reviewed. Immunohistochemistry was performed for cytokeratin (CK) 7, CK20 and p63.
Results: PMA of the eyelid was identified in the three male patients, ages 63–73 years old. Immunohistochemistry demonstrated positive staining for CK7 and GCDP-15 and were negative for CK20. One of the 3 cases was stained for p63, and it was found to be positive.
Conclusion: Due to the difficult clinical diagnosis of this often benign appearing lesion, it is imperative that physicians send all specimens for histopathological and immunohistochemical correlation. Advances in IHC including CK7 and CK20 as well as p63 are important for the diagnosis of this rare eyelid tumor, though are not yet totally definitive in their ability to distinguish PMA from other lesions with similar profiles.

1. Background

Primary mucinous adenocarcinomas (PMA) of the eyelid are rare entities clinically, and even more so in the literature. Since first described in 1952 by Lennox et al. [1], a total of 55 cases have been identified in English literature from 1979 until 2010 [2].

Clinically, these lesions are often unremarkable to patients due to their indolent nature. They have been described in a variety of ways including papillomatous, pedunculated or fungating. As well, colors can vary widely ranging from flesh to tan, grey, red or blue in hue. Other descriptors that have been used include smooth, bumpy, crusted, spongy, or firm [3]. Oftentimes, the tumors can have been present for a year or more prior to initial presentation to a healthcare professional, a trend also demonstrated in our cases.

Histopathologically, PMAs have been classically described as a neoplasm consisting of basaloid cells surrounded by large clear pools of loosely woven mucinous stroma [4]. It is acknowledged that the appearance of this particular tumor can vary, contributing to the difficulty in making such a rare diagnosis. Historically, the tumor was classified as eccrine [3]. The contemporary view held by the World Health Organization, however, is that PMAs have both eccrine and apocrine differentiation [5,6].

Immunohistochemical (IHC) characterization of these tumors has evolved in time with the eventual goal being to help reduce the panel of screening tests that patients undergo before classifying the mucinous adenocarcinoma of the eyelid as a primary one.

The treatment options for these tumors consist of primary excision ensuring negative margins due to the high rate of recurrence of these tumors (>40%), or Mohs micrographic surgery done in the case of deeply invading tumors in order to provide a pleasant cosmetic outcome.

2. Materials + methods

2.1. Case 1

A 67-year-old gentleman was evaluated in the oculoplastics clinic for a recurrent cyst on the right lower eyelid in the lateral commissure. The patient had cysts removed in this area both one, and five years prior to the current presentation. No pathology was previously performed. Other than controlled hypertension, the patient had no significant past medical history.

Clinical examination revealed a cystic lesion at the lateral commissure of the right lower eyelid. The lesion was excised and a pathological

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correlation was sought. The histopathological correlation revealed a mucinous adenocarcinoma that was not completely excised.

Subsequently, a re-excision was performed that involved the lateral aspect of both the upper and lower eyelids. Specimens were taken for frozen section analysis and were reported as being negative for tumor tissue. The lids were repaired. The final pathology report noted that though the margins appeared to be uninvolved, this could not be guaranteed.

Five years later, the patient again returned due to a recurrence of the right lower eyelid. It appeared cystic in nature, trans-illuminated, and was dumbbell shaped (See Fig. 1 #1). At the time, excision was refused. Nine months later, the patient again returned due to the enlarging nature of the lesion – that had now reached 13 mm in width. Again, tissue was sent for frozen section and the margins were considered negative. The eyelid was repaired.

Follow-up at six months was unremarkable for recurrence.

2.2. Case 2

A healthy 73-year-old man was referred to the oculoplastics clinic because of a mass on his right upper eyelid (See Fig. 1 #2). The lesion was noted one year prior to consultation and had been slowly enlarging during that period. The mass appeared to be cystic in nature and trans-illuminated. Grossly on clinical exam, it was bi-lobed in shape. There was no palpable enlargement of the lymph nodes.

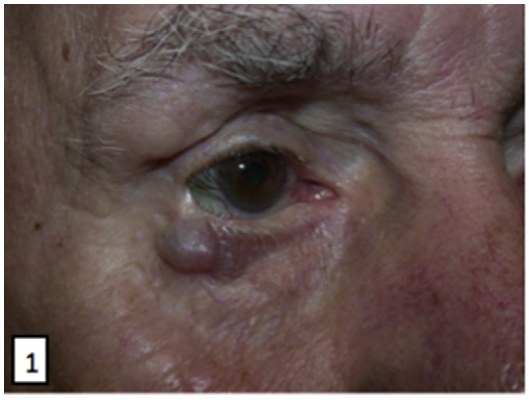
The patient returned to the operating room where the lesion was excised. Frozen section analysis revealed clear tissue margins. The upper lid was repaired with a Cutler-Beard procedure. The final Pathology report noted that there was a complete excision of the lesion.

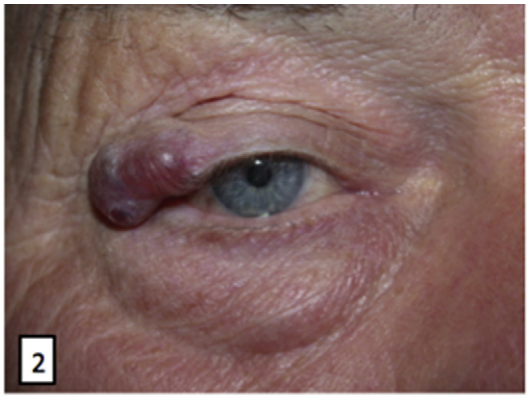
He returned for the second stage of the Cutler-Beard procedure six weeks later. The exam was otherwise unremarkable. The patient lived several hours from the hospital, and was followed-up by the referring physician.

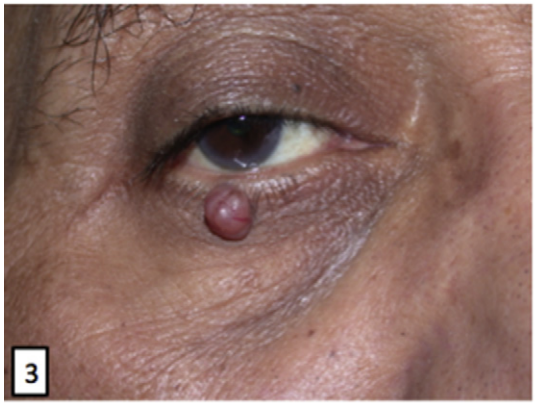
2.3. Case 3

A 62-year-old man was referred to the oculoplastics clinic due to the presence of a lesion of the right lower lid that had been increasing in size over a period of one year. Clinical examination revealed a lesion measuring 7 mm in diameter on the center of the lid that appeared cystic in nature (See Fig. 1 #3).

An excision of the lesion was performed and a specimen was submitted to Pathology. The lesion was reported as a mucinous adenocarcinoma. There was no confirmation that the margins were negative.







Case 1: A sessile bluish tumor, cystic in appearance with poorly demarcated borders on the lower eyelid is seen

Case 2: A bilobed, cystic tumor that transilluminated was seen on the upper eyelid.

Case 3: A polypoid lesion on the lower eyelid

Case	CK7	CK20	GCDP-15	P63
1	+	-	+	+
2	+	-	+	NA
3	+	-	+	NA

Fig. 1. Clinical Photographs and Summary of Immunohistochemical Markers.