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Ophthalmic Pathology for Dermatopathologists: Unique aspects of pathology of the eyelid and conjunctiva

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Skin Tumors

Conjunctival Tumors

Specialized Adnexal Gland Tumors

Orbital and Lacrimal Gland Tumors

Soft Tissue Tumors

Conjunctival Tumors
Anatomical considerations:

1. Aggressive behavior of “benign” tumors
2. Perineural spread
3. Bad location
4. Skeletal muscle involvement
Misdiagnosis of ocular sebaceous carcinoma

Overdiagnosis of conjunctival nevus

Misunderstanding of conjunctival melanosis

Misclassification of adnexal neoplasms

Missing conjunctival SCCIS and sebaceous carcinoma

Ignorance of issues and entities
57 year-old F referred to Moh’s surgery with diagnosis of SCC
Moh’s procedure stopped as the surgeon could not clear the margins.
She realized it “must be a different tumor”, stopped the procedure and send the debulk for histology.
Moh’s surgery debulk
62 year-old male referred to Moh’s surgery with diagnosis of BCC. Moh’s procedure stopped as the surgeon could not clear the margins. She realized it “must be a different tumor”, stopped the procedure and send the debulk for histology.
Debulk from Moh’s surgery
60 year old male initially diagnosed as SCC
“clinical picture did not fit”
Ocular Sebaceous Carcinoma

Clinical masquerader

- 65% Meibomian
- 25% Zeiss
- 10% caruncle or skin

Bad prognosis:
- Meibomian and Zeiss glands
- >2 cm
- >6 months
- High histological grade

- 0.2-0.7% eyelid tumors
- >40 y.o.
- M:F = 1-1/5
- Radiation
- Locally aggressive
- 15-30% metastasize
- 15-80% 5 year survival
Ocular Sebaceous Carcinoma

- Misdiagnosis
  - SCC
  - BCC
  - Missed

Histological masquerader

Key histological features:
- In situ:
  - Pagetoid spread
  - Involvement of hair follicles and sebaceous glands
- Invasive
  - Poorly differentiated
  - Infiltrative growth
  - Nests with comedo necrosis
- Cytology
  - Bubbly vacuolated cytoplasm

Diagnostic adjuncts:
- Oil red O
- Cytoplasmic bubbly EMA

Not associated with Muir Torre syndrome

Poorly differentiated, No sebaceous cells!!!
Metastatic sebaceous carcinoma
Infiltrative growth
Follicular spread
Infiltration of sebaceous glands
Comedonecrosis
Bubbly cytoplasm
Pagetoid spread
Cytoplasmic bubbly EMA
Cytoplasmic Oil-Red-O
WHO (1980):

- Melanocytic nevi
- Conjunctival melanosis
  - primary or secondary
  - congenital or acquired
  - with atypia or without atypia
- INVASIVE melanoma

50% of conjunctival neoplasms

“the term melanoma in situ could unnecessarily alarm both clinician and patients, particularly since many PAM lesions have little propensity to evolve into melanoma”.

Conjunctival nevus

- 28% of conjunctival tumors
- Bulbar > caruncle > plica semilunaris
- Young caucasians (mean age 32 years)
- Flat to slightly raised pigmented macules or papule
- 30% amelanotic

- Junctional
- Compound (70%)
- Subepithelial

- Movable
- Well-circumscribed
- Does not involve cornea
- Contain intralesional cysts
“Juvenile conjunctival nevus”

- Confluent and pagetoid growth
- Reverse maturation
- Inflammation

Primary acquired melanosis

- 10% conjunctival tumors
- Acquired
- Middle age or older
- 30% amelanotic
- Bulbar

Without atypia

With atypia
**Clinical behavior of epithelioid and non-epithelioid PAM with atypia**

<table>
<thead>
<tr>
<th>Category</th>
<th>Non-epithelioid (Low-risk)</th>
<th>Epithelioid (High-risk)</th>
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<tbody>
<tr>
<td>1 Recurrence</td>
<td><img src="#" alt="Graph Bar" /></td>
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<td>&gt;1 Recurrence</td>
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<td>Invasion</td>
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<td>Metastases</td>
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Low-risk and High-risk Histologic Features in Conjunctival Primary Acquired Melanosis With Atypia: Clinicopathologic Analysis of 29 Cases

*Mitsuihiro Sugiura*,† *Kathryn A. Colby,*‡ *Martin C. Mihm, Jr.*§ and *Artur Zembowicz*†

Non-epithelioid

- Minimal visible cytoplasm (high nuclear/cytoplasmic ratio)
- Small hyperchromatic nuclei without nucleoli
- Lentiginous growth pattern
Epithelioid PAM

- More cytoplasm visible (low nuclear/cytoplasmic ratio)
- Prominent nucleoli (9 cases, 56%)
- Pagetoid spread (9 cases, 56%)
High-grade PAM=Melanoma in situ

- 50% cases had 2 or more recurrences
- 94% cases associated with invasion into the lamina propria
- 25% metastatic disease; 1 death
Conjunctival melanoma

Poor prognosis:
• Non-bulbar
• Epithelioid
• Large size
• High mitotic rate

• Not movable
• Irregular, poorly circumscribed
• Can involve cornea
• No intralesional cysts

• Bulbar>caruncle>eyelid
• Middle age to older
• 30% amelanotic
• De novo>PAM>nevus
• 15-30 % metastasize
• 70 % 5 year survival
Unusual features in adnexal neoplasms arising in specialized ocular adnexa
Endocrine mucin-producing sweat gland carcinoma

- Low-grade
- Reminiscent of papillary solid CA of the breast
- Older individuals
- Precursor to mucinous carcinoma

- Lower or upper eyelid
- Nodule
- Sometimes multiple
- Can involve both eyes
Stages of development of mucinous carcinoma

- In situ
- Endocrine mucin-producing sweat gland carcinoma
- Invasive mucinous carcinoma
56 year old F with a long history of atopic conjunctivitis treated with steroids and cyclosporin
Biopsy performed to rule out infection
Biopsy 8 months later
Diagnostic pitfall

- Small cell squamous cell carcinoma in situ arising the context of atopic conjunctivitis treated with cyclosporine
Phakomatous choristoma of the eyelid: a report of a case


Choristoma (hamartoma) of lenticular tissue

Skin-covered nodule on the nasal aspect of lower eyelid

Presenting at birth or in first 6 months of life

Size: 6-20 mm

Searching the old consultation files of the Children’s Hospital in Boston, we encountered a case of an eyelid tumor in a newborn, which had remained unclassified for 35 years. In April 1967, the case had been sent for consultation by Dr. F. C. Blandi, Professor of Ophthalmology at the University of Iowa, to Dr. G. F. Vawter, Professor of Pathology at Children’s Hospital Medical Center in Boston, MA. Referring to a series of congenital tumors of the parotid gland recently published by Dr. Vawter in his letter requesting consultation, Dr. Blandi wrote:

‘...I saw some time ago a congenital tumor of the lacrimal glands which may be similar to your tumors... Any comment you would care to make on this would be greatly appreciated.’

In response, Dr. Vawter wrote:

‘I have puzzled over this for sometime, not having personally seen a lesion of lacrimal or salivary gland quite like this before. Nor have those colleagues to whom I have showed it. In short this differs considerably, on the surface, from the lesions I reported... A most fascinating problem.’

From the current perspective, it is evident that this above exchange of letters took place at a time when additional cases of this rare tumor have been reported in the literature. We present this case, as it represents an excellent example of this tumor type, to remind the young readers of Dr. Vawter and Dr. Blandi who made significant contributions to pediatric and ocular pathology, respectively, throughout their outstanding careers.

Case report

The patient was a male infant who presented at the University of Iowa’s Department of Ophthalmology for a congenital tumor of the lower eyelid of suspected lacrimal gland origin. Clinical information about this patient is limited to the information in the letter of Dr. Blandi. We were not able to obtain more clinical information about this patient, as his medical records were no longer available.

Materials and methods

The patient’s tissue blocks were retrieved from the
You can find more examples of interesting conjunctival lesions on www.dermpedia.org