

A rare case of extramammary Paget's disease at the lateral canthus as a mimic of basal cell carcinoma

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Introduction – Extramammary Paget's disease

- Rare intraepithelial adenocarcinoma, typically presents in the **6-8th decades of life**
- Commonly affecting **apocrine-rich areas**: vulva, scrotum, penis, perineum and perianal region
- **Ocular EMPD is extremely rare**
- Can be split into **primary and secondary forms**
 - **Primary** – arises from adnexal pluripotent cells
 - **Secondary** – epidermotropic spread from an underlying internal malignancy
- Can mimic benign skin lesions or inflammatory eyelid conditions, leading to **diagnostic delay**.

Case history

A male patient in his eighties presented to the oculoplastic clinic following a referral from his opticians.

Presentation:

- Crusted lesion on right lower eyelid near the lateral canthus
- Present for 2-3 years with recurrent bleeding episodes

Histopathology:

- Carcinoma in situ with pagetoid spread of highly atypical intraepidermal cells
- Immunohistochemistry: **CK7+, CK20+, EMA+, CEA+, CAM 5.2+**
- **Neuroendocrine markers and GCDFP-15 negative**
- Findings **suggestive of secondary EMPD**

Management and outcome

- Lesion excised with clear margins
- MDT recommended clinical surveillance for 3 years
- 3 months: no evidence of local recurrence

Macroscopic appearance and histopathology

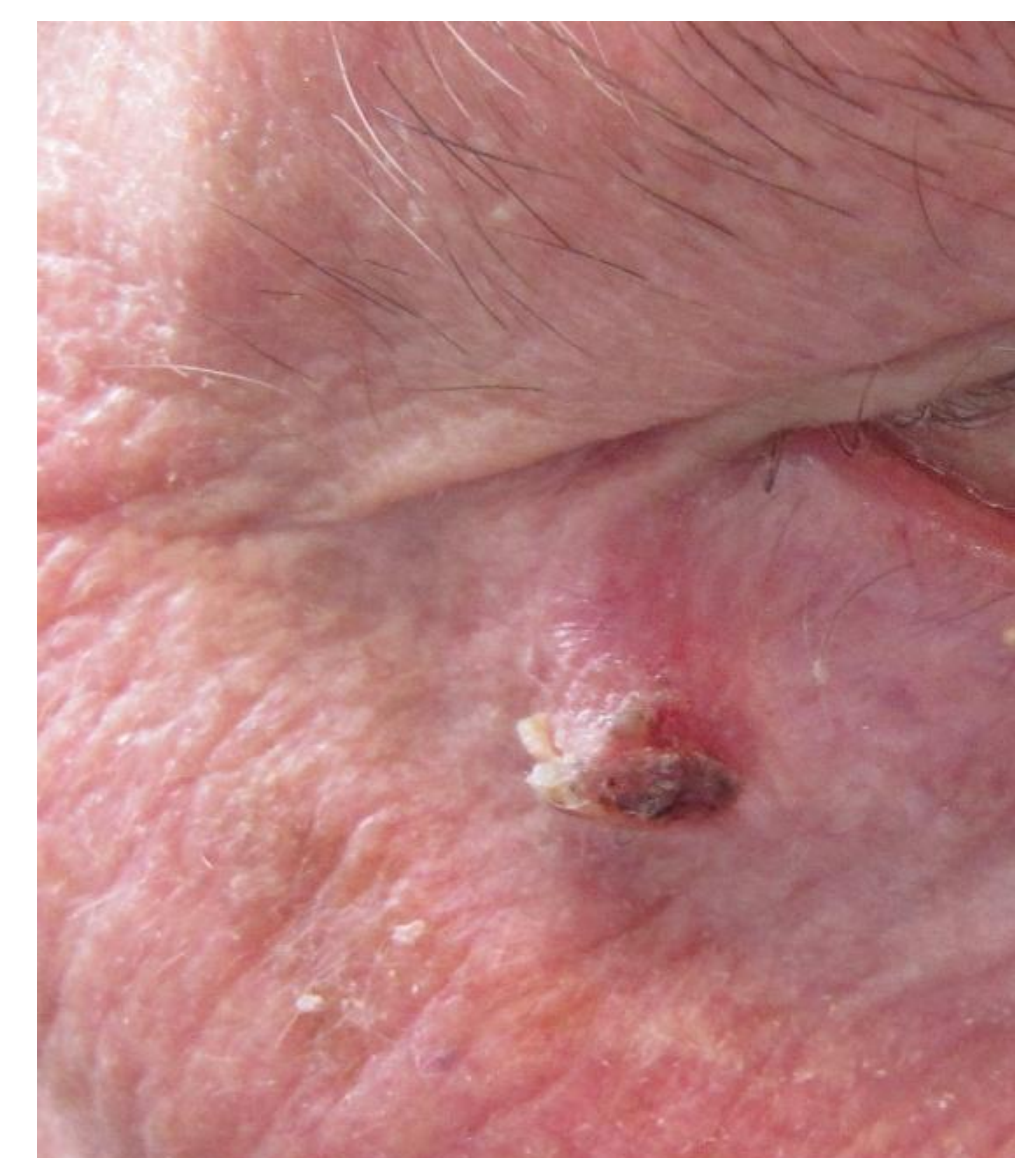


Fig.1: Photograph of the lesion inferior to the right lateral canthus on the right lower eyelid.

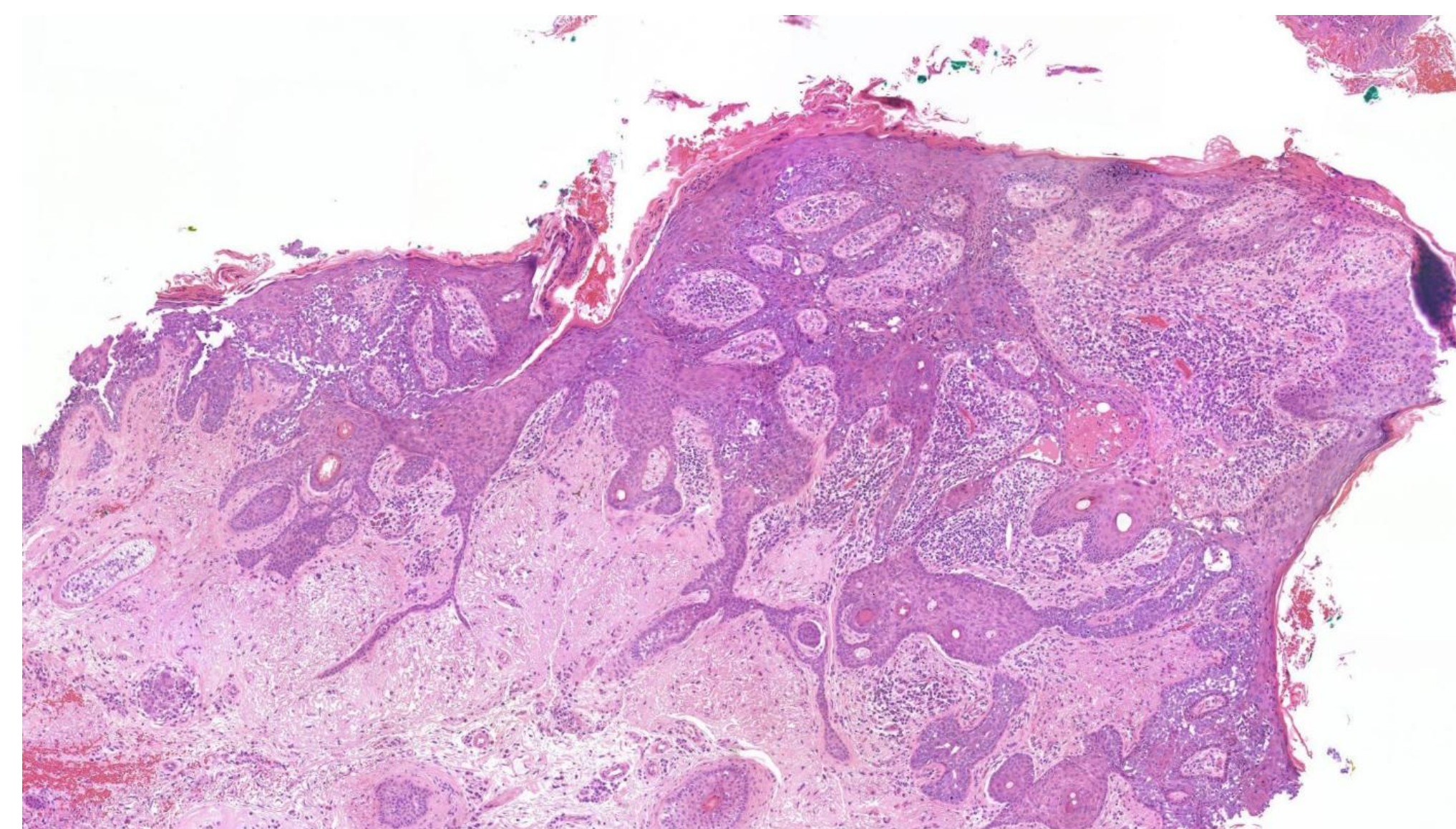


Fig.2: Histological appearance of the lesion (section stained with haematoxylin & eosin). Magnification at 5x. Basaloid cells can be seen clearly within the lesion.

Staining

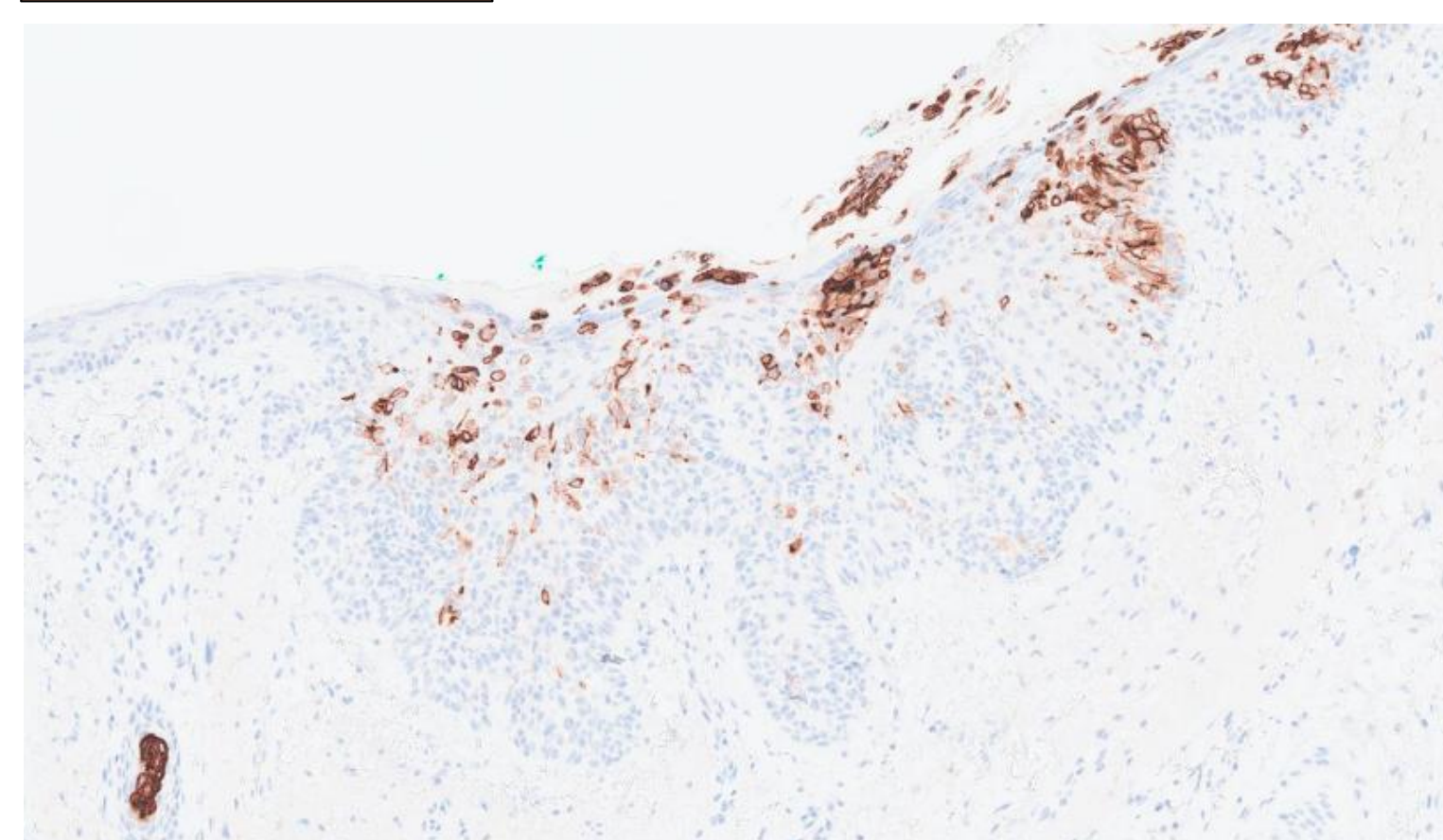


Fig.3: Staining of the lesion reveals **positive** expression of **CEA** by the lesion cells.

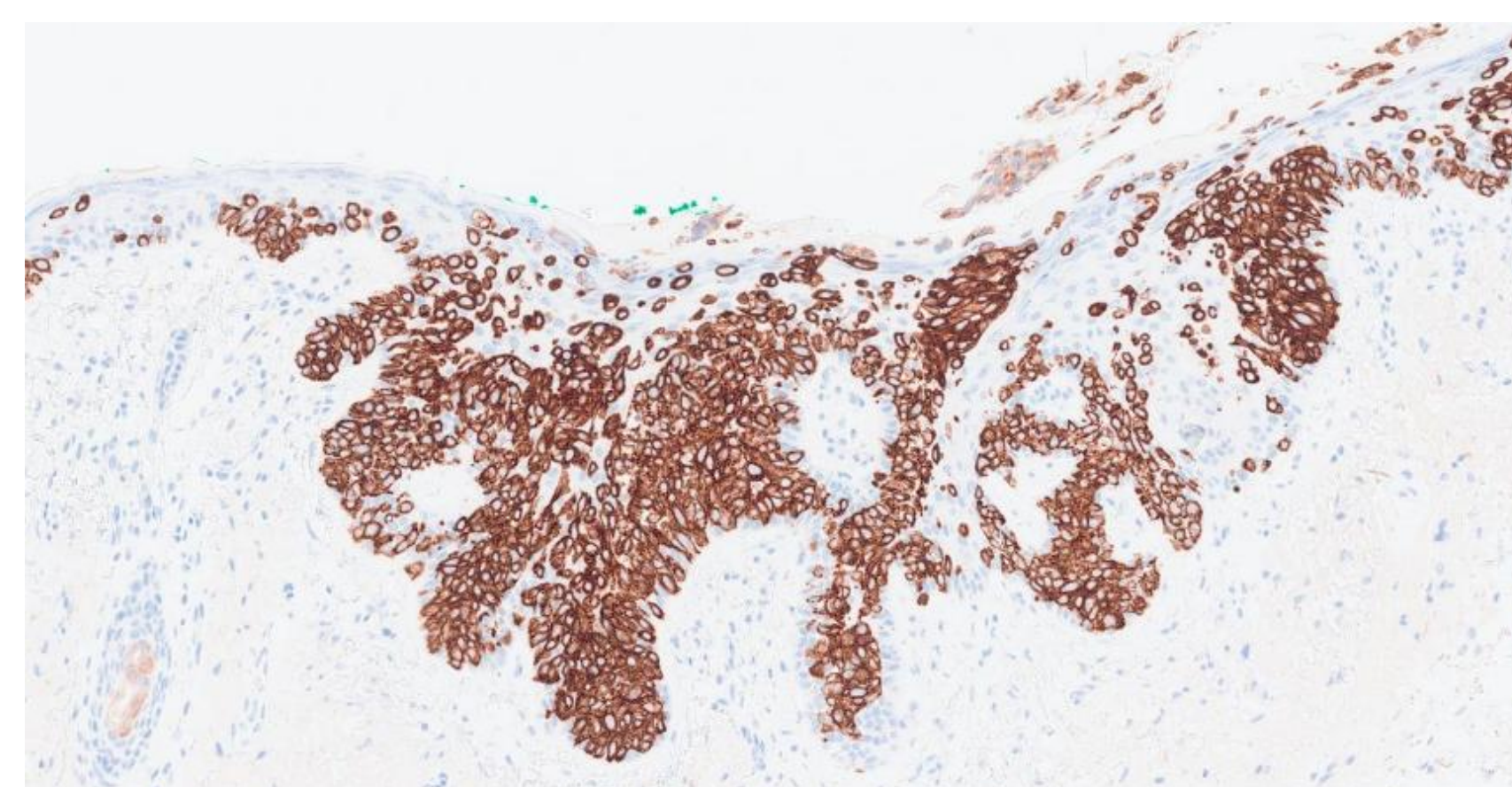


Fig.4: Staining of the lesion reveals **positive** expression of **CK7** by the lesion cells.

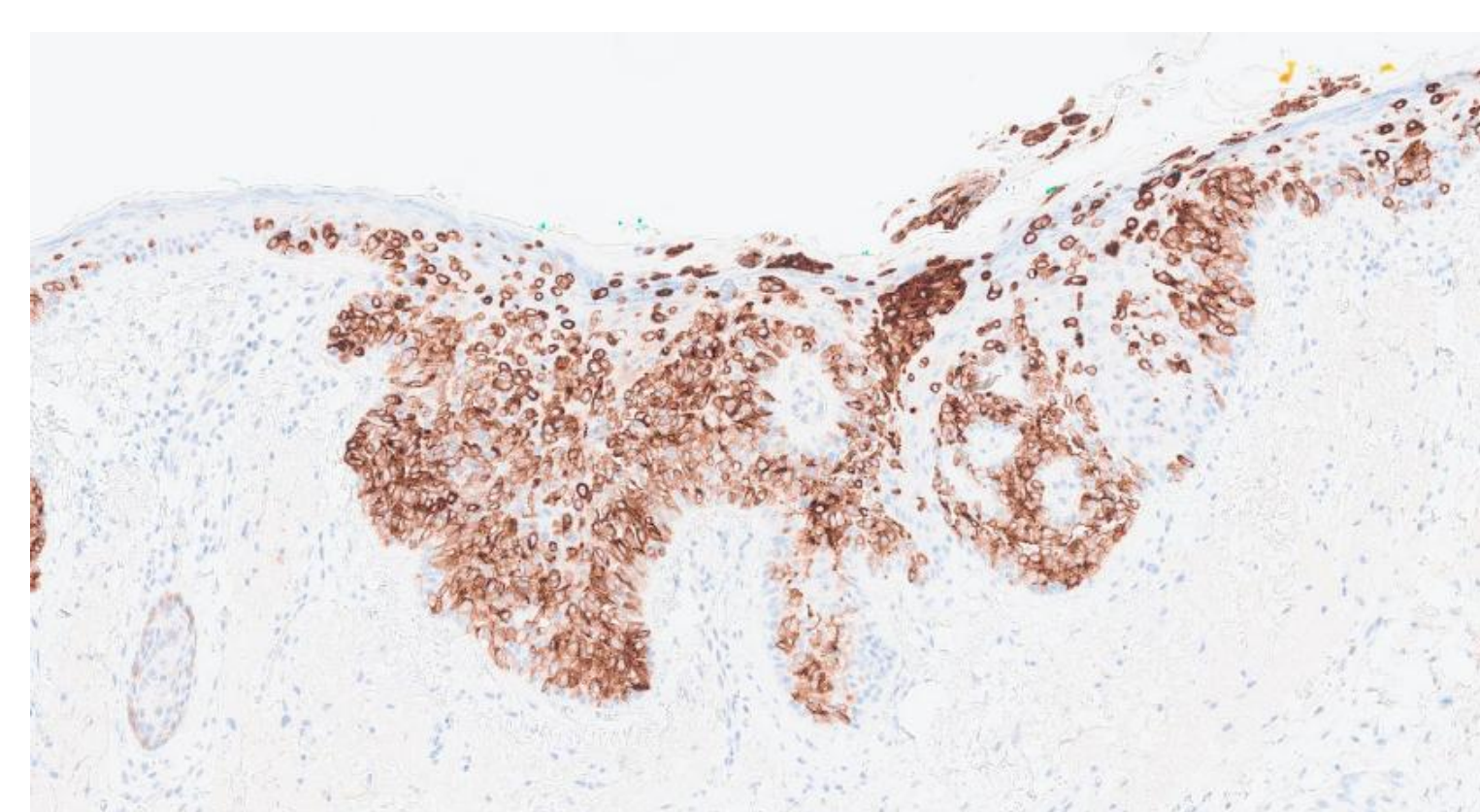


Fig.5: Staining of the lesion reveals **positive** expression of **EMA** by the lesion cells.

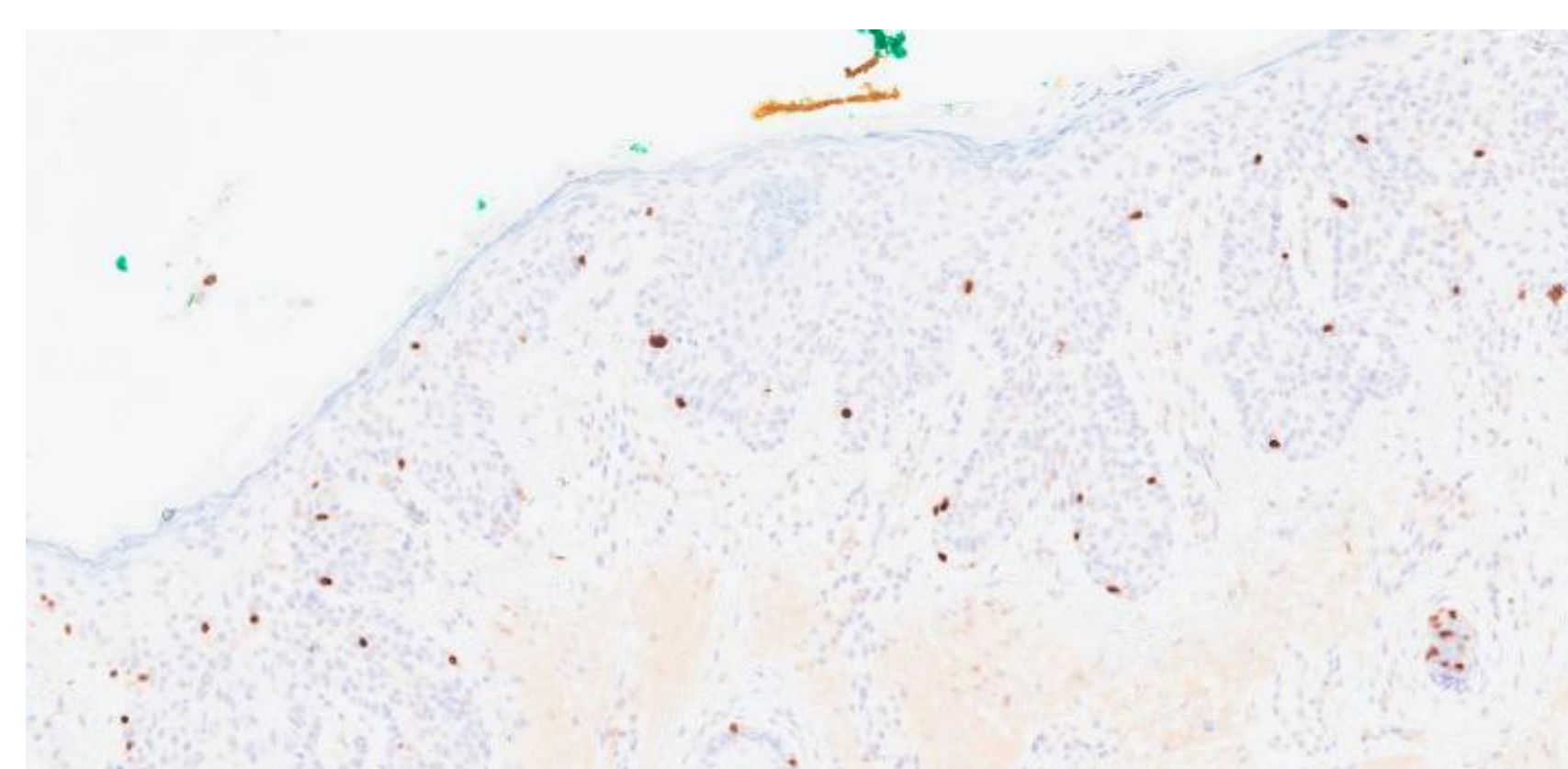


Fig.6: Staining of the lesion reveals **negative** expression of **Sox10** by the lesion cells.

Discussion

Management considerations:

- **High recurrence rates** due to subclinical extension and discontinuous growth – incomplete excision
- **False-negative margins common**
- Mohs micrographic surgery preferred to ensure complete excision

Immunohistochemistry profiles:

- **Primary:** CK7+, GCDFP-15+, CK20-
- **Secondary:** GCDFP-15+ and:
 - Colorectal: CK20+, sometimes CK7+
 - Urothelial: Both CK20+ and CK7+

In our case: secondary extension likely from the glands of Moll

- Negative Sox10 excludes a differential diagnosis of melanoma

Differential diagnosis

- Melanoma in situ
- Pagetoid Bowen's disease
- Sebaceous carcinoma in situ

Careful **long-term surveillance** required

Early detection critical

Conclusion

- Extramammary Paget's disease can present in the eyelid as a BCC mimic and should be fully excised with clear margins, given its propensity for recurrence.
- Immunohistochemical markers are essential for establishing a diagnosis of EMPD and stratifying it as primary or secondary, possibly helping identify an origin in secondary EMPD.

References

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