EUROPEAN OPHTHALMIC PATHOLOGY SOCIETY (EOPS)
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Material submitted: One histology H&E slide

Title of Case: A hairy eyelid problem

CLINICAL HISTORY: Male 27 yo, who presented with a 4-5 months history of a growing red/dome shaped lump from the right upper lid

MACROSCOPY: A wedge of skin and conjunctiva 13mm superior to inferior, 11mm from medial to lateral and up to 5mm thick. There is a pale tan firm red nodule at the superior most aspect of lash line measuring 12x10mm.

MICROSCOPY: Lobulated and circumscribed dermal based neoplasm. Composed of islands of basaloid cells exhibiting abrupt keratinization, without intervening granular layer (trichilemmal keratinization). Presence of Ghost / shadow cells noted.
Basaloid cells show occasional isolated mitotic activity; however, abnormal mitoses were not seen. Intermediary cells progressively more eosinophilic cytoplasm, with pyknotic nucleus. Tumour keratin elicit inflammatory response with foreign body giant cells, granuloma, cholesterol clefts and focalacute inflammation. No calcificationwas seen in the sections examined.

DIAGNOSIS–Benign calcifying epithelioma of Malherbe (pilomatricoma/pilomatrixoma), completely excised. Pilomatrixoma/Pilomatricoma.

DISCUSSION – Pilomatrixomafirst described by Malherbe and Chenantais in 1880. It is a rare benign adnexal tumour originating from the hair matrix and cortex. It is an uncommon neoplasm representing approximately 1% of all pathologically diagnosed benign skin tumours. Tumours can occur at any age, but about 60% of cases are diagnosed in the first two decades of life with a female preponderance. Rare cases are presented in the eyelid. Pilomatricoma presents as slow growing, dermal based, asymptomatic nodule measuring 0.5 - 3 cm. Clinically it presents as a flesh colored to white, firm papules / nodules that may have an overlying pink to blue hue. Pain, pruritus or discharge can occur at times. Occasionally, skin ulceration may be seen simulating squamous cell carcinoma.
Rare appearances may include bullous, perforating, pigmented lesions, keloid-like.
Head and neck are the most common sites of involvement, followed by upper and lower extremities and then trunk.

Under the microscope it has a lobulated appearance and appears as a circumscribed dermal based neoplasm. Composed of islands of basaloid cells exhibiting abrupt keratinization, without intervening granular layer (trichilemmal keratinization). Presence of Ghost / shadow cells noted. It has been described the uncommonly presence of metaplastic bone formation includingextra medullaryhaematopoiesis. Hemosiderin, melanin and amyloid deposition has been reported in isolated cases.

Although pilomatricoma are usually not hereditary, familial cases have been recognized. Familial forms have been seen in myotonic dystrophy Curschmann-Steinert (an uncommon autosomal dominant disorder characterized by hypotonia, muscle wasting, cataracts, hypogonadism, progressive mental retardation and frontal baldness), familial adenomatous polyposis related syndromes (including Gardner syndrome), Turner syndrome, Rubinstein-Taybi syndrome, trisomy 9, gliomatosis cerebri and others.

Complete excision is curative, recurrence is unlikely and diagnosis can generally be established by histopathological examination.

Malignant transformation (PilomatrixCarcinoma) rarelydevelop. The few cases described have been reported in older adults(case reports document occurrence in children and young adults, mostly de novo). It is rare and they arise from existing pilomatricoma. Histologically it shows pleomorphic basaloid cells, prominent nucleoli, high mitotic activity, necrosis, lymphovascular invasion (rare). It shows locally aggressive behaviour, infiltrating borders (2) .Local recurrence is common. Metastases may rarely occur, involving regional lymph nodes and lungs

DIFFERENTIAL DIAGNOSIS

Pilomatrixoma is rarely suspected clinically and can be easily mistaken by a cyst, sebaceous carcinoma, chalazion and other tumours. Limited sampling is the most common source of mistakes in view of the different components and reactions seen in these tumours.

REFERENCE

1. SepidehSiadati, Ashley A Campbell, Timothy McCulley, Charles G Eberhart. Clinicopathological features of 19 Eyelid Pilomatrixoma. Ocul Oncol Pathol, 2021 Oct 25;8(1):30-34
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