Masquerades less known: Case report of benign hair follicle tumors

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Benign eyelid tumors derived from hair follicles are rare and frequently misdiagnosed as basal-cell carcinoma when evaluation is based on clinical evidence alone. The discrepancy between the clinical aspect and the histological diagnosis can be as high as 30–40%, even for experienced clinicians. Unfortunately, this masquerade is not only clinical but also histological. Patients may be subjected to unnecessarily extensive surgery. We present a case report of three patients clinically diagnosed as malignancy but histopathologically proven as benign hair follicle tumors.

Key words: Brookes tumor, hair follicle tumors, trichoblastoma, trichoepithelioma, tricholemmoma

Tumors of the eyelids, benign or malignant can either arise from specific anatomical structures like Meibomian and Moll’s glands, tarsal plate or from the skin and ocular adnexa. Effective management of malignant tumors requires scrupulous attention to the completeness of removal. Conversely, the much rarer benign hair follicle tumors of the eyelids may be safely excised with a narrow margin of clearance. We present a case report of three patients to give an insight of benign tumors in which just an excision of tumor alone is sufficient to avoid large reconstructions of the eyelid.

Case Reports

Case 1

A 45-year-old female presented to us with swelling in right eye for 4 years, increasing in size for the past 6 months. History of right eye upper lid mass excised 5 years back. Ocular examination was normal except for a 1 × 1 cm nontender swelling extending to tarsus with telangiectatic vessels on surface with distorted meibomian gland orifices with segmental loss of eyelashes in the right upper lid [Fig. 1a]. Suspecting meibomian gland carcinoma mass excision biopsy with 4 mm margin was done with direct closure. Pathological studies showed structure of hyperkeratotic squamous epithelium with mild papillomatosis with thickened eosinophilic basement membrane. The down growth of epithelial cells shows increasing clear cell differentiation with heavy pattern and infiltration up to deep stroma S/O trichilemmoma [Fig. 1b].

Case 2

A 48-year-old female presented to us with a growth in left lower lid for 20 years which has increased in size for 6 months. On examination, a multilobulated, multicolored mass of 10 × 18 mm attached to skin seen in lower lid with vessels on surface and surface ulceration [Fig. 2a] and eyes were otherwise normal. Provisionally diagnosed as malignant melanoma, a mass excision biopsy was done with 4 mm margin clearance. Pathology showed structure of squamous epithelium and sub epithelium nests of basaloid cells with palisades. Areas of keratinous cysts with focal cribriform pattern and tumor cells showed atypical features of increased mitosis and vesicular nuclei. No evidence of deep stromal invasion S/O trichoepithelioma with minimal atypical features [Fig. 2b].

Case 3

A 65-year-old male presented with progressive, painless growth in left upper lid for 5 years. Left eye had a blackish lesion with irregular borders with central ulceration and vessels on surface [Fig. 3a]. Provisionally diagnosed as basal cell carcinoma/malignant melanoma an excision biopsy was done. Histopathological examination (HPE) showed acanthotic squamous epithelium and subepithelial tumor tissue arising from follicular germinative cells arranged in nests and rows with bottom heavy pattern. Stromal eosinophils with interspersed melanin pigmentation seen with focal area of intratumoral necrosis S/O Pigmented trichoblastoma (TBL) with low grade features [Fig. 3b].

Discussion

Tumors of the skin and the eyelid can present clinical and histopathological diagnostic problems. In all cases the diagnostic considerations must include the case history, the shape, color, consistency, and slit-lamp appearance of the tumor and the result of histopathological examination of the completely excised preparation. The conclusions reached should determine the (further) treatment and follow-up. Hair follicle tumors can be benign or malignant growths.

Benign hair follicle tumors

- Basaloid follicular hamartoma
- Fibrofolliculoma
- Pilar sheath acanthoma
- Trichofolliculoma

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• Trichoepithelioma
• Pilomatrixoma
• Trichoblastoma
• Trichoblastic fibroma
• Trichoadenoma
• Trichilemmoma
• Trichilemmal cyst (pilar cyst)
• Proliferating trichilemmal cyst
• Pilomatrixoma.

Malignant hair follicle tumors
• Trichilemmal carcinoma
• Trichoblastic carcinoma
• Marginal proliferating trichilemmal cyst
• Pilomatricoma carcinoma.

Some specific follicle tumors are seen in syndromes associated with internal malignancies, for example, trichilemmomas in Cowden disease. Also, many tumors have a counterpart which, although rare, may be locally aggressive and have the potential to spread and metastasize. Trichilemmoma is a benign tumor that arises from the outer layers of the hair follicle in adults. Clinically, they appear as a small nodule with either smooth skin-colored papules or wart-like lesion with irregular rough surface that can be mistaken for verruca or cutaneous horn. Histologically, the lesions show lobular acanthosis of glycogen-rich clear cells with often palisading cells with distinct basal membrane in its periphery. Hair follicles may be seen in the lesion.[6]

Trichoepitheliomas are benign tumors of hair follicle origin and appear in three varieties: solitary, multiple, and desmoplastic. Multiple trichoepitheliomas can occur alone as multiple familial trichoepitheliomas or in association with cylindromas and syringomas as Spiegler–Brooke syndrome.[9] They appear as a firm elevated nonulcerated skin-colored nodule. Histologically, the tumor shows multiple horny cysts with fully keratinized centers surrounded by islands of proliferating basaloid cells that are sometimes difficult to distinguish from basal cell carcinoma (BCC). The stroma of the tumor is well demarcated from the surrounding dermis. When high differentiation exists, abortive formations of hair papillae and hair shafts can be seen.[7]

TBLs are benign well-margined skin neoplasms of follicular germinative cells. These tumors are typically sporadic, symmetric, solitary, small (often <2 cm), arise from the hair follicle and grow slowly. Variants: Clear cell, pigmented, adamantinoid, rippled-pattern. Rippled-pattern TBL is characterized by peculiar arrangement of the basaloid cells in linear rows parallel to one another similar to the palisading of nuclei of Verocay bodies seen in schwannomas. Cutaneous lymphadenoma is an adamantinoid variant of TBL. Depending on the degree of differentiation toward other follicular components and the relative prominence of stromal and epithelial elements this group was classically subdivided into TBL numbers, trichoblastic fibroma, and trichogenic TBL. Trichogenic TBL is the most mature form and includes complete hair follicle. The principle histopathological differential diagnosis of TBL is from BCC. BCC can be much more irregular, invasive, and destructive than TBL. In BCC, there are clefts between the epithelium and stroma, whereas in TBL the clefts are within the periepithelial stroma and the surrounding dermal collagen. Mitoses, apoptotic bodies, mucinous stroma, or amyloid deposits are typically present in BCC. In contrast to BCC, papillary mesenchymal bodies are characteristically present in TBL. Mitoses and apoptotic bodies are rare in TBL. Bcl-2 expression in TBL is limited to the basoloid keratinocytes in the outermost layer, but BCC shows diffusive staining.[8] In TBL, CD34 and CD10 are expressed only in the peritumoral stromal cells, while in BCC intraepithelial staining is observed.[8,9] CK20-positivity of Merkel cells may be abundantly found in TBL but not in BCC.[10] Recently, expression of PHLDA1,[11] a marker of follicular stem cells, has been reported in TBL but is absent in BCC. Malignant alteration and aggressive features in TBLs can lead to BCC or trichoblastic carcinoma,[12] but this is rare.

As for our knowledge is concerned, there is no case series of benign hair follicle tumors published in Indian literature. Benign hair follicle lesion can be safely removed without recourse to wide margins. In all our three patients excision was done and they were asymptomatic since 1 year. Confirmation by incisional biopsy of the nature of any large lesion suspected of being a malignant tumor is essential in order to avoid performing an unnecessarily extensive or time-consuming excision where frozen section is not available. Since this is underdiagnosed, as ophthalmologists we must be aware of
this clinical and histological masquerade when assessing tumors and inflammatory conditions of the eyelid. This lesion is reported infrequently among oculoplastic pathology. Though it closely mimics malignant lesions of eyelid, it cannot be proved unless there is a high index of suspicion especially in trichoepithelioma.

Conclusion
This article aims at recreating an awareness among oculoplastic surgeons and ocular pathologists about the masquerades of malignant lesions and thus avoids extensive lid reconstruction.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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