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Case Report

Recurrence and Management Challenges of Upper Eyelid Pilomatrixoma: A Rare Case Study with Atypical Presentation

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ABSTRACT

Pilomatrixoma is a benign tumor of the hair matrix cells and is usually confused with other entities since there are nonspecific findings. Most of the patients are young individuals, and involvement is usually in the head and neck region; however, rarely, involvement in the periorbital area has been reported. We present the recurrent case of pilomatrixoma in a 37-year-old male upper eyelid, as well as diagnostic and treatment difficulties that are commonly found in these tumors. The tumor was initially excised under local anesthesia. After three months, it recurred, which was further confirmed on histopathology as pilomatrixoma due to its characteristic features of basaloid cells and ghost cell formation. A good point here is that a recurrent tumor following good initial removal points to the necessity for very careful follow-up or, perhaps, more aggressive initial surgery. This underlines the fact that pilomatrixoma can present with a varied clinical spectrum; hence, histopathological confirmation is necessary for case definition. Pilomatrixoma, in some way, recalls the fact that, though the treatment is adequate at first, a chance of recurrence is probable and, thus, contributes to the body of experience in managing this rare disease.

Introduction

Pilomatrixoma is a benign neoplasm originating from the matrix of hair and is commonly termed calcifying epithelioma of Malherbe. Usually, this is seen to occur in the age group of children and adolescents, usually over regions of the head and neck. Occurrence at the site of the periorbital, especially at the level of the upper eyelid and the eyebrows, is strikingly rare, which may pose a challenge in differential diagnosis (1,2). Most pilomatrixomas are clinically misjudged due to their nonspecific presentation, and histopathological examination is post-excision dependent to make an accurate diagnosis (3,4). Pathologically, pilomatrixoma is recognized by unique cellular formations: shadow cells, basophilic cells, inflammatory responses, foreign body giant cells, and calcification that provide important diagnostic markers (4,5). The first part of the surgical excision is the overlying skin. Recurrence post-excision is rare, especially when performed with safe margins. The report describes a rare case of recurrent pilomatrixoma in the right upper eyelid after extensive surgical excision.

Case Description

A 37-year-old male patient reported a history of the development of the lesion over the superior lateral aspect of the right upper eyelid rapidly for three months. Past medical history was unremarkable, including no pain, swelling, redness, or history of trauma in the area. Clinical examination revealed a sessile, firm, non-tender, rock-hard nodular mass. The mass was fixed and sited at the inner surface of the eyelid, covered by normal tight-tone skin but with white nodules seen. It was encapsulated in a continuous fibrotic capsule of variable thickness around the lesion. Excision was taken up under local anesthesia. Within three months of successful excision, he again was seen with similar presenting complaints. The histopathological examination postoperatively revealed the classical features of pilomatrixoma, including small basaloid cells with evidence of keratinization, formation of a ghost cell, and colonization by melanocytes.

Discussion

The presentation of pilomatrixoma in our case is particularly noteworthy for several reasons. In our case, lesion growth was very rapid and adhesion to the internal tissues of the eyelids. It even caused suspicion of malignancy degeneration in the very first place. This would be unusual for a pilomatrixoma, which classically comes out as mass-like, firm, and nontender with ill-defined borders and is completely movable under the skin. The classical clinical picture involves a nodular mass, with the possibility of skin discoloration in the range of reddish to blue hues, due to the vessels beneath the epidermis being dilated. Chalky white nodules are also visible through the skin in some instances (6).

The demographic profile of our patient also provides an interesting deviation from the norm. However, most commonly, pilomatrixomas are predominant in the Caucasian population. Our presented case is relevant as it reminds readers that the Middle Eastern origin of the patient is known to have an impact on broad ethnicities without signaling strong racial predilection (4, 7). In addition, female preponderance in pilomatrixomas is well documented (4,8,9), and the finding of pilomatrixoma in our male patient also serves to build the knowledge that pilomatrixomas, though infrequent, are found in both genders.

Another issue of interest is the location of the anatomical and clinical presentation of the lesion of our patient. Pilomatrixomas usually occur in the upper eyelid or brow area and add to a proportion of head pilomatrixomas (4). Our case adds to the growing evidence of pilomatrixomas exhibiting a wide range of clinical variation in their presentation, including the possibilities alluded to by Choi et al. Pilomatrixoma histologically represents one of the characteristic clinical manifestations in a wide range of eyelid tumors. A pilomatrixoma can be presented as subcutaneous nodules and pedunculated or sessile prolapse tumors of the eyelid with a necrotic surface and as a cyst (8). The varied presentations of pilomatrixoma underline the importance of considering it in the differential diagnosis of any atypical eyelid lesion.

Another critical aspect of this case is the differential diagnosis process. This is due to the fact that the lesion exhibits a benign and malignant tumor spectrum, for which the differential diagnosis includes sebaceous adenoma, epidermal inclusion cyst, dermoid cyst, sebaceous gland carcinoma, basal cell carcinoma, and Kaposi sarcoma, to name a few (10, 11). Since so many clinical features overlap with other clinical conditions, histopathological and immunohistochemical investigations are warranted to come to an accurate diagnosis.

The other pathological findings show basaloid cells and ghost cells, which are the features of pilomatrixoma (4, 9). However, ghost cells are not specific to pilomatrixomas and may be found in some other conditions, such as epidermoid cysts, chronic inflammation of the hair follicle with hyperkeratosis, and a few other chronic dermatoses. This would, therefore, justify a review histologically at the highest level to differentiate pilomatrixoma from those that bear a close resemblance. In our case, though the histology was characteristic, the patient did show a recurrence—a course relatively rare, following surgical excision in about 2.6% of cases (12, 13, 14, 15). This has led to the assertion that the management of pilomatrixoma is quite complicated and requires proper planning in surgery that should encompass wide-margin excisions to reduce any possibility of recurrence (16, 17).

Further, the occurrence of two recurrences in our case post-surgical resection challenges the very biological behavior of the pilomatrixomas and intrinsic factors that might be responsible for its recurrences. This lesion has been traditionally described as a benign entity. However, the extremely aggressive behavior of this in this case sets out a vast field of much more complex pathophysiological mechanisms. Molecular and genetic studies could elucidate, in pilomatrixomas, the varied clinical features and high tendency for tumor recurrence.

Conclusion

Pilomatrixoma of the eyelid, while rare, should be a consideration in the differential diagnosis of eyelid masses. This case highlights the need for comprehensive surgical management and the potential necessity for repeat excision in the event of recurrence. The unusual presentation and recurrence, in this case, add valuable insights into the diverse clinical spectrum and management challenges of pilomatrixoma.

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Conflict of Interest

The author declares no conflict of interest.

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