Case Report

Calcifying Tumor of Malherbe

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Abstract

Tumours of the eyelid are not very common. Frequently we come across squamous cell carcinoma. We are reporting a rare case of calcifying epithelioma of eye. The usual presenting region is the hair bearing areas of body and upper extremity. A 15 years old male presented with swelling over upper lid. Clinically diagnosed as molluscum contagiosum. But histopathologically confirmed as calcifying epithelioma of the eye. Epithelioma situated over the eyelid is very rare.

Keywords: Calcifying epithelioma, Pilomatricoma

1. Introduction

Calcifying epithelioma is a rare benign tumor known to occur in children. Only 11 to 50% cases are known to be diagnosed preoperatively. The incidence has been reported as 1/1000 of cutaneous tumors occurring commonly in head and neck region. It accounts for up to 0.2% of all routine dermatological specimens. It predominantly affects females, the male to female ratio being 1:3. Even though modalities like ultrasound, MRI and cytopathology are used the diagnosis may be missed. A confirmed diagnosis is obtained after histopathology alone. A misdiagnosis leads to unnecessary extensive surgery. The tumor involves the skin with calcification in centre of epithelial cell clusters arising from skin appendages. It shows the tendency of calcium deposits to occur in sites of mitochondrial inactivity in tumor masses because of their large size and diminished blood supply. There is faulty suppression of apoptosis in pathogenesis of pilomatricoma. They are seen in hair bearing areas of the face. Other cases have been reported but this is the only case we came across at our tertiary care centre. We present this because of the diagnostic difficulties it presents and because histopathology plays important role in further treatment. An incomplete surgical excision can be followed by local recurrence and hence a complete excision must be done, however there is no need for an extensive surgery as the possibility of malignant transformation is rare. We assert that pilomatrixoma should be considered in the differential diagnosis of firm skin nodule on the eyelid or brow in the children and the middle-aged patients.

2. Case History

A 15 years male presenting with swelling over upper lid of right eye since 5-6 months. The swelling was painless, progressive and gradually increased in size. The clinical diagnosis was molluscum contagiosum. Histopathologically confirmed as calcifying epithelioma of the eyelid.

Grossly received single soft tissue 0.5 cm with central white area surrounded by black area at the periphery. Histopathological examination showed basophilic masses, with shadow cells.
3. Discussion

Pilomatricoma is also known as Calcifying Epithelioma of Malherbe and was described in 1880. It is defined as tumor with differentiation towards hair cells, especially hair cortical cells. It occurs as solitary lesion present on face & extremities. Grossly it measures 0.5-3 cms in size. It usually occurs in first two decades of life. Rapid enlargement may occur because of haematoma formation or perforation. As a rule it is not hereditary. It also occurs in myotonic dystrophies and can be present with Gardner’s syndrome. It is well encapsulated and well demarcated by a rim of connective tissue. Irregular islands of epithelial cells are seen in dermis. At periphery basophilic cells and shadow cells with keratin are present. The peripheral basophilic cells are embedded in symplasmic mass, while the shadow cells lack nucleoli. As disease progresses the number of shadow cells increase. Seventy five percent of tumor cells show calcification. The calcification is seen as basophilic fine granules or large deposits replacing shadow cells. Ossification may take place in 15-20% of cases. Strong reaction with PAS stain is seen due to keratin. Mutation with beta-catenin is seen. The epithelial and stromal responses of chronic conjunctivitis are non specific and can be initiated by low grade persistent or intermittent infection of lid margin and lacrimal sac. Chronic inflammation sets in due to conjunctival exposure resulting from proptosis or from ectropion of the lower lid, vitamin A deficiency, tear function disorders, allergies, toxic topical medications, embedded foreign body and neoplasms. During chronic inflammation goblet cells increase in number, epithelium undergoes hyperplasia, resulting in focal crypt like epithelial folding, pseudoglands of Henle become isolated from surface to form subepithelial retention cysts containing entrapped secretions. In time secretions become inspissated to form calcific concretions. Stromal inflammation shows formation of follicles and papillae. Papillae are seen as paving block or giant papillae causing irregularities.

Recently imaging modalities have been used for diagnosis of pilomatrixomas. On ultrasonography, pilomatrixomas are heterogeneously hyper-echoic with internal echogenic foci and a peripheral hyper-echoic rim or completely echogenic with strong posterior acoustic shadowing in the subcutaneous layer. On magnetic resonance imaging (T1W1) pilomatrixoma gives homogenous intermediate intensity signal and does not enhance on contrast.

These should be differentiated from sebaceous cysts, dacryoliths, trichilemmal cysts and pilomatrix carcinoma. Dacryoliths are small calculi which form in the lacrimal sac. They appear as laminated, basophilic structures and are associated with actinomyces like Arachnia propionica. Trichilemmal cysts show palisading pattern which is not seen in pilomatrixoma. Their shadow cells show a central unstained area, while those of pilomatrixoma show central blue areas of calcification. Pilomatrix carcinoma, is a low-grade malignant lesion with a tendency to recur and carries a risk of distant metastases. Infiltration of surrounding tissue, necrosis, high number of atypical mitotic figures, perineural or vascular invasion are the typical histological features.

The tumor does not regress spontaneously. Treatment is recommended to avoid a foreign body reaction and inflammation with eventual scarring. Surgical excision is the preferred method of treatment. We performed an excision following which the patient is fully recovered with no signs of recurrence at the end two years.
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