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Eye Lid Pilomatricomas

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Abstract: Pilomatricomas are uncommon benign tumors usually involving the head and neck region. They are commonly seen in children and adolescents with a female preponderance. We report four cases of eyelid pilomatricomas diagnosed between 2009 and 2012 and review the clinical and histopathological features. Clinically two cases were diagnosed as dermoid cyst, one case as pyogenic granuloma and one as epidermal cyst. The diagnosis of Pilomatricoma was done in all the cases after histological examination of the excision biopsy specimen.
Keywords: Pilomatricoma, Benign tumor, Subepidermal, Eyelids, Clinical features, Histology

INTRODUCTION

Calcifying Epithelioma of Malherbe is an uncommon benign tumor of the head and neck region in children and adolescents. The incidence has been reported as 1/1000 of cutaneous tumors in the head and neck region [1]. According to one study, the female to male ratio was 3:1 [2]. The tumor originates in the hair follicle matrix. It arises as a subepidermal tumor originating from the cells of hair matrix [3, 4]. The tumor was first described by Malherb in 1880 [2, 5]. The term 'Pilomatricoma' was first used by Forbis and Helwig in 1961 [4, 5]. Clinically Pilomatricoma presents as a firm non-tender subcutaneous nodule adherent to the skin, but not fixed to the underlying tissue [6]. It is often misdiagnosed clinically and the correct diagnosis can be established only after excision and histological examination [7]. The malignant variant, variably termed as malignant pilomatricoma or pilomatrical carcinoma is very rare [2, 8].

CASE REPORT

We studied four cases of Eyelid Pilomatricomas between 2009 and 2012 at Sarojini Devi Eye Hospital, Hyderabad. We analysed the clinical data including age, sex, site, size of the tumor and clinical diagnosis. We studied the histology and followed up the cases for one year after complete excision.

RESULTS

Among the four cases, three were female patients and one was a male patient. The age range was between 10 years to 26 years. In all the cases, the tumor was painless, slow growing present since the last three to six months. There was no history of trauma. No familial association was noted. All the patients had a single tumor. All four were present on the upper eyelid, three on the right upper lid and one on the left side. Two eyelid tumors were located on the middle portion of the eye lid, one on the inner portion and one on the outer portion of the eyelid. All the tumors were well defined and fixity to the skin was present in all the four cases. Three were firm and one was hard on palpation. The color of the tumor was brown in three cases and reddish brown in one case. Ocular examination was normal in all the cases. Two of the cases were clinically diagnosed as dermoid cyst, one was diagnosed as epidermal cyst and the fourth as pyogenic granuloma. The clinical data with histological correlation is shown in table 1.

Table 1: Clinical data with histological correlation

Sl. No.	Age (years)	Sex	Side	Size	Clinical diagnosis	Histology	Recurrene (One year)
1.	10	Female	Right	1x1cm	Dermoid	Pilomatricoma	Nil
2.	16	Female	Left	1.2x0.8cm	Dermoid	Pilomatricoma	Nil
3.	21	Male	Right	1.5x1cm	Pyogenic granuloma	Pilomatricoma	Nil
4.	26	Female	Right	1x0.7cm	Epidermal	Pilomatricoma	Nil

					cyst		

Excisional biopsy was done in all the cases. Histologically, the tumors were composed of sheets of basaloid cells, shadow cells or ghost cells (Fig. 1,2 & 3) with foci of calcification, small areas of keratinization and foreign body giant cell reaction (Fig.4) in the stroma. Foci of ossification were noted in one case. The tumors were followed up for a minimum period of one year and no tumor recurrence was noted in any of the cases.

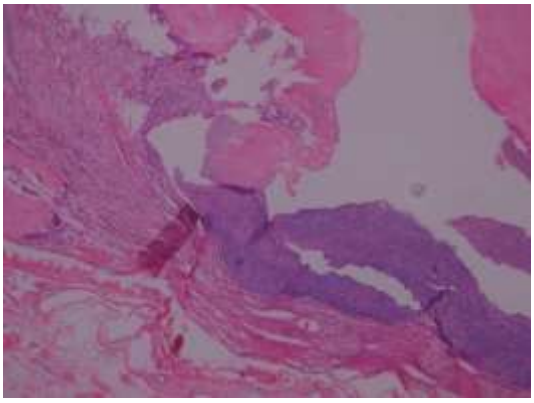

Fig.1: Section shows basaloid cells & ghost cells

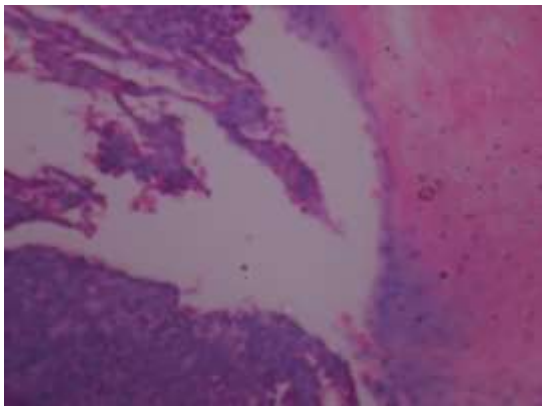

Fig. 2: Higher magnification

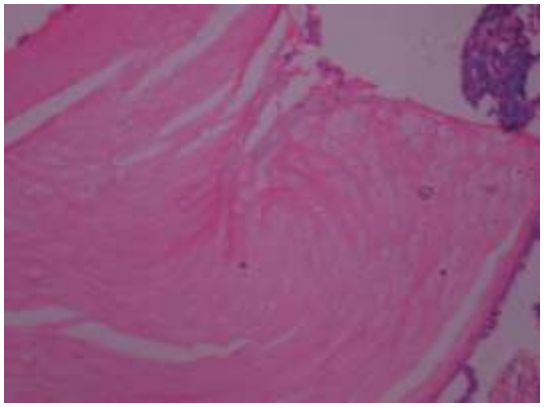

Fig. 3: Section showing ghost cells

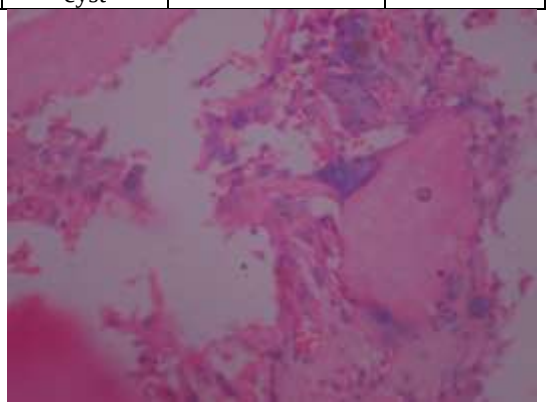

Fig. 4: Section showing foreign body giant cells

DISCUSSION

Pilomatricoma is also known as Calcifying Epithelioma of Malherbe [1]. It is a tumor arising from the external hair shaft of the hair root [2]. Its distribution corresponds to the density of hair follicles at the particular site [2, 3]. Hence the coomon occurance in areas like scalp and face. It has never been reported in areas like palms and soles, because of the lack of hair follicles in these areas [4]. Pilomatricoma may appear at any age, but the peak presentation occurs bimodally in first and sixth decades [2, 3]. However, all our patients were in the second and third decades. Female preponderance had been reported with a male to female ratio of 2:3 [4]. In our study of four cases, the male to female ratio is 1:3. Pilomatricomas are usually solitary, though multiple tumors occurred in 3.5% of the cases according to Enrique Menica-Gutierrez *et al*. Pilomatricoma appears more frequently in patients with myotonic dystrophy than in general population [2, 3]. They may be related to Sarcoidosis or Gardner's syndrome [3, 4]. In our study, all the four patients presented with solitary tumors which were firm to hard on palpation and non-tender [6]. The tumor is often misdiagnosed clinically and the correct diagnosis is established after excision and histological examination [7].

The tumor is commonly diagnosed clinically as dermoid cyst, inclusion cyst, pyogenic granuloma, capillary hemangioma and chalazion. The skin over dermoid cyst looks normal and can be moved freely over the lesion. Inclusion cysts have a diffuse yellow color and are softer and fluctuant than pilomatricoma. Capillary hemangiomas can have a reddish blue color and will be soft and compressible. Chalazion may become inflamed and may be yellowish in color [6]. Granuloma pyogenicum is a hard and rapidly enlarging lesion. Histologically, the tumor is well encapsulated and shows basaloid and shadow cells at the periphery with areas of keratinization, fibrosis, calcification, foreign body granuloma and ossification in the stroma. Ossification may appear in 15 to 20% of the cases [1,

2]. In our study, ossification was seen in one case out of four cases. Radiological imaging is of little diagnostic value for pilomatricoma. Complete surgical excision including the overlying skin is the treatment of choice [4]. Recurrence is very rare with a reported rate of 2.6%. [2]. Our cases were followed up for one year after excision and there was no recurrence in any of the cases. The malignant variant, pilomatrical carcinoma is very rare and is suggested histologically by nuclear atypia and atypical mitoses [8]. They can metastasize to the lungs, bone and other viscera and the outcome is poor in such cases [2].

CONCLUSION

We reported four cases of Eyelid Pilomatricomas occurring in adolescents and young adults. Pilomatricoma should be considered in the differential diagnosis of eye lid and eye brow tumors in children and adolescents. The tumor is commonly misdiagnosed clinically and histological examination is required for the correct diagnosis of the tumor.

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