

## Multiple facial pilomatricomas

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**Abstract** Pilomatricoma is a benign and often misdiagnosed tumor arising from hair matrix cells. It is predominantly found in children below the age of 10 years and is characterized histologically by basaloid and eosinophilic ghost or empty cells. Pilomatricoma is generally solitary and multiple tumors are rarely reported. We report one such rare case of multiple facial pilomatricomas in a one-year-old girl.

**Key words**

Pilomatricoma, hair matrix, ghost cells, excision.

### Introduction

Pilomatricoma is a benign superficially located, appendageal tumor with differentiation toward hair cells. It usually manifests as a solitary, asymptomatic, firm nodule in the head and neck. It is more common in children, but occurrence in adults is increasingly being reported. Diagnosis is made by clinical examination and imaging and confirmed by histopathological analysis. Recommended treatment is complete surgical excision

### Case report

A one-year-old girl was brought by parents with history of swelling over left eyebrow. The swelling was noticed at 2 months of age and had been progressively increasing in size. There was no other significant past or family history.

On examination, there was about 2cm x 1.5 cm, hard lump over the left eyelid laterally (**Figure**

1). The lesion was adherent to skin but was mobile over the underlying orbital rim. There was loss of hair over the lesion. The lesion was non-reducible and there were no visible or palpable pulsations over it. Cough/cry impulse was also negative over the lump. Examination of right eyebrow revealed a similar but smaller (about 0.4 cm x 0.3 cm) lump laterally but without any hair loss or significant projection and hence not noticed by parents. Provisional diagnosis of bilateral external angular dermoid cysts was made and CT scan was advised to study the extent of the lesion. CT scan revealed the lesions to be well-demarcated, subcutaneous and hyperdense, with left lesion being markedly heterogeneous (**Figure 2**) and right lesion more densely calcified (**Figure 3**). None of the lesions showed intraorbital or intracranial extension. The patient was operated upon under general anesthesia and both the lumps were completely excised. Histopathological analysis of the specimens showed features of pilomatricoma. There were no perioperative complications. At 6 months of follow-up, there was no recurrence.

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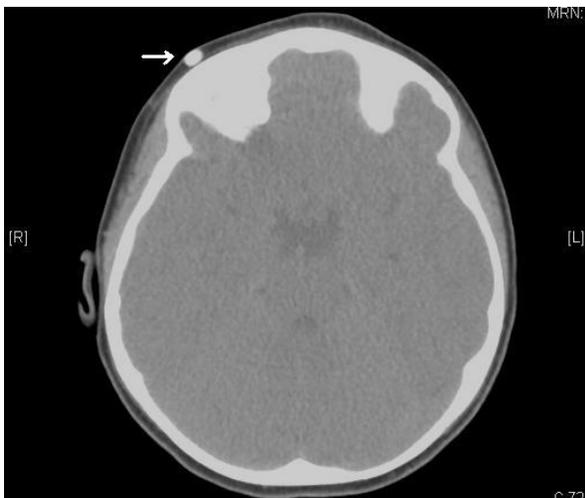
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**Figure 1** Pilomatricoma (red arrow) over left eyebrow laterally.



**Figure 2** Non-contrast CT scan showing left eyebrow heterogeneous lesion (white arrow) – pilomatricoma.



**Figure 3** Non-contrast CT scan showing right eyebrow hyperdense lesion (white arrow) – pilomatricoma.

## Discussion

Pilomatricoma (also called pilomatrixoma) is a benign tumor originating from the matrix of the hair root. This lesion was first described in 1880 by Malherbe and Chenantais who thought it to arise from sebaceous glands and called it calcifying epitheliomas of Malherbe. Lever and Griesemer, however, suggested the origin of the tumor to be hair matrix cells. The term pilomatrixoma was first used in 1961 by Forbis and Helwig, thus avoiding the word epithelioma, which carries the connotation of malignancy.<sup>1</sup>

Pilomatricoma typically presents as a firm, non-tender, subcutaneous nodule adherent to the skin but not fixed to underlying tissue. Most of the tumors are usually 2-3 cm in size though giant tumors of up to 18 cm are reported in literature.<sup>2</sup> Growth is usually slow and may occur over a period of months to years. Patients are usually asymptomatic, but some report pain during episodes of inflammation or ulceration. Pilomatricoma can present at any age, it demonstrates bimodal peaks in presentation, with up to 60 percent of cases occurring in the first two decades and a secondary peak in the sixth decade. Female preponderance has been reported with a male:female ratio of 2:3. The most frequent site is in the head and neck with more than 75% of the pilomatricomas being located on the scalp, face, neck or arms. No cases have been reported on the palms or soles, perhaps because of the lack of hair-bearing skin in the areas.

Mutations of the beta-catenin gene were detected in 75% of the pilomatricomas but the exact role of such mutations remains to be elucidated.<sup>3</sup> In one study of 10 pilomatricoma lesions, all immunostaining results were strongly positive for BCL2. This is a proto-oncogene that helps suppress apoptosis in benign and malignant

tumors; this data suggests that faulty suppression of apoptosis contributes to the pathogenesis of these tumors.<sup>4</sup> Recent investigations have further demonstrated that the proliferating cells of human pilomatricoma show prominent staining with antibodies directed against LEF-1 (a marker for hair matrix cells). Evidence also indicates that S100 protein can be used as biochemical marker in characterization of pilomatrixomas.<sup>5</sup> This data provides biochemical support of morphological evidence that these tumors are derived from hair matrix cells. Furthermore, investigators have shown that at least 75% of persons with pilomatricoma have mutations in the gene CTNNB1<sup>6</sup>; this data directly implicates faulty regulation of beta-catenin/LEF as the major cause of hair matrix cell tumor formation in humans.

Diagnosis may be arrived at by physical examination, imaging and histopathological examination.<sup>7</sup> Even though pilomatricoma is the commonest appendageal tumor in children, the diagnosis is frequently missed on physical examination alone by physicians unaware about this lesion<sup>8</sup> as happened with our case. Plain radiography often shows nonspecific calcification of the lesion. Ultrasound shows pilomatricoma to bear heterogeneous echo texture, internal echogenic foci in scattered-dot pattern, and a hypo-echoic rim or posterior shadowing. The classic CT radiological appearance has been described to be that of a non-contrast enhancing, sharply demarcated, subcutaneous nodule with amorphous calcifications. MRI shows a tumor with well-defined margins and high-signal intensity on T1- and T2-weighted images. Histologically pilomatricoma is a deep subepidermal tumor consisting of irregular islands of epithelial cells. The epithelial cells are organized in a characteristic biphasic architectural pattern with

keratinized ghost cells in the center surrounded by variable amounts of peripheral basaloid cells. The basaloid cells exhibit deeply staining basophilic nuclei, which often contain small nucleoli. The keratinized ghost cells in the center have lost their nucleus and thus have a central unstained area.<sup>9</sup> As the lesion ages, the number of basophilic cells decreases. Calcium deposits may be seen in up to 75% of lesions with von-Kossa staining.

In the vast majority of cases, pilomatricoma is a solitary neoplasm, however, multiple tumors occurring synchronously account for about 3.5% of reported cases.<sup>10</sup> Multiple or recurring tumors may be found in association with different conditions<sup>10</sup> which include Gardner's syndrome, myotonic dystrophy, Rubinstein-Taybi syndrome, sarcoidosis, trisomy 9 and Turner's syndrome. Myotonic dystrophy (Steinert disease) usually presents during the teenage years or adolescence, but the onset of pilomatricoma can range from many years before or many years after the onset of symptomatic myotonic dystrophy. Hence long-term follow-up is recommended in this subset of patients.

The differential diagnosis of pilomatricoma is varied. Firstly, pilomatricoma should be differentiated from sebaceous and dermoid cysts. Sebaceous cysts are firm, round, and mobile and may have a punctum. Dermoid cysts are firmly attached to underlying tissue and are often found in children. Other entities to be considered in the differential diagnosis include: inclusion cysts, giant cell tumor, foreign body reaction, eccrine spiradenoma, osteoma cutis, trichilemmal cyst, hydrocystoma, basal cell carcinoma, calcification in lymph node, fat necrosis, pyogenic granuloma, lymphadenopathy, fibroxanthoma, chalazion and keratoacanthoma.<sup>1</sup>

The treatment of choice and standard therapy for benign pilomatricoma is complete surgical excision. Recurrence rates after excision are insignificant and pilomatrical carcinoma should be considered if lesions recur after complete excision particularly in elderly age group. This malignant transformation is exceedingly rare and besides recurrence may be suggested by rapid growth, pain, itching, ulceration and bleeding. Pilomatrical carcinoma is a low-grade malignant lesion and carries a risk of distant metastases.

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