

Pilomatricoma: a review of six pediatric cases with nine lesions

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SUMMARY: Turhan-Haktanır N, Demir Y, Şahin Ö, Bükülmez A, Tüzüner M. Pilomatricoma: a review of six pediatric cases with nine lesions. Turk J Pediatr 2009; 51: 44-48.

Pilomatricoma, although rare, is the second most frequent benign skin tumor in childhood. It is a hard, painless, slowly progressing subcutaneous tumor that is commonly located in the head-neck region, especially periorbital, and in the upper extremities. Histopathologic characteristics are distinctive. In this study, we performed a retrospective review of the records of our pediatric patients with pilomatricoma. We discuss herein the diagnosis and management of this uncommon skin tumor, and compare our experience with the previous literature. Pilomatricoma should be included in the differential diagnosis of superficial pediatric head and neck masses by any physician involved with pediatric tumors.

Key words: pilomatricoma, child, skin neoplasms.

Pilomatricoma is a benign tumor originating from hair follicle matrix cells. Although rare, it is the second most frequent benign skin tumor in childhood that mainly occurs in the head and neck region¹. Malignant transformation is exceedingly uncommon. Clinical diagnosis is not difficult but pathologic examination is necessary because it does not show spontaneous regression. Therefore, the preferred treatment is total surgical excision. For a physician concerned with diseases in children, clinical recognition of this lesion and planning the management are important. We aimed to retrospectively analyze the records of our pediatric patients with pilomatricoma, to discuss the diagnosis and management, and to compare our experience with the previous related literature.

Material and Methods

We performed a retrospective chart review of six patients (4 female, 2 male) with pilomatricoma who were admitted and treated in the plastic surgery clinic in our institution over a three-year period (2004-2006). The patients' ages were 8 to 14 years (mean 10).

Detailed data regarding clinical presentation, work-up, treatment, and results were reviewed. Data regarding age, sex, site of origin, and pathologic features were collected. Diagnoses were confirmed after the analyses of pathologic specimens by an experienced pathologist.

Results

Six tumors were found in the head and neck region, and the remaining three tumors were located in the upper extremity. Two of the patients with multiple tumors had lesions of the head.

The common appearance of the tumors was a hard, slowly growing, semi-mobile, lobulated, and well-circumscribed subcutaneous nodule. Minimum size of the lesions was 0.5x0.4 and maximum size was 2x1 cm. The tumors were solitary in three patients and dual lesions were seen in three patients. Three lesions were adherent to overlying skin (Figs. 1 and 2).

Histologically, the lesions were well demarcated and lobulated, and located in the dermis or subcutaneous tissue. Basaloid cells, shadow cells, and occasional foreign body giant cells



Fig. 1. Different locations of the pilomatricoma: infraorbital cheek (upper) and forearm (lower).

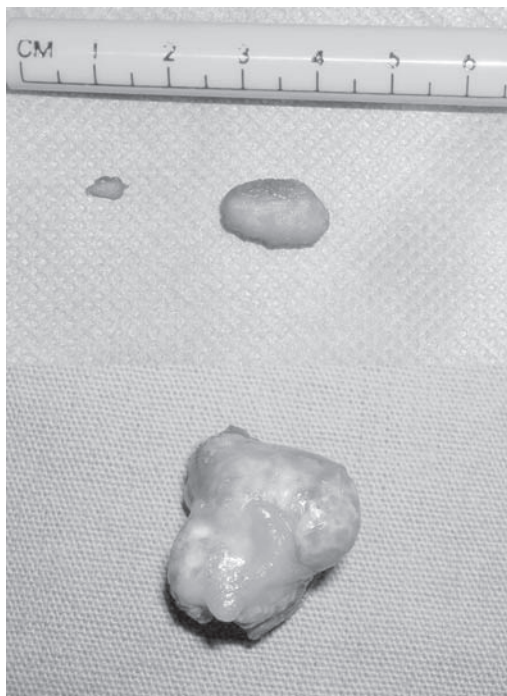


Fig. 2. Gross appearance of two pilomatricomas. The upper image shows a tumor excised with covering skin; the lower shows a well-circumscribed lobulated tumor that was not adherent to skin.

were identified in the tumors. Basaloid cells had intensely staining basophilic nuclei and scant cytoplasm, with indistinct cell margins. Shadow cells showed discrete margins and central unstained regions. Giant cells and large keratinocytes were also seen associated with shadow cells and keratin debris. Intracellular and stromal calcifications were frequently seen (Fig. 3).

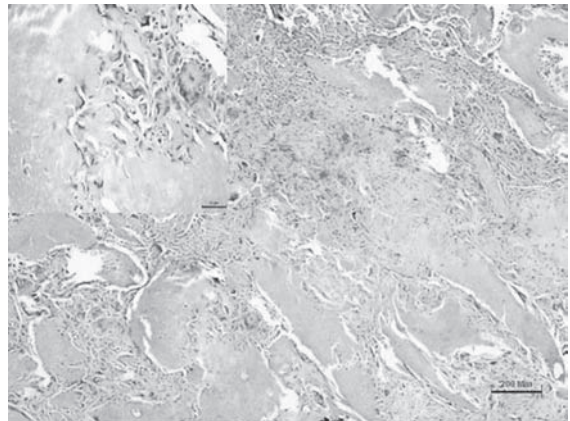


Fig. 3. Hematoxylin and eosin stain (x200 original magnification) (inset, original magnification x400) shows areas of dark basophilic cells and masses of eosinophilic “ghost” cells with scattered foreign body giant cells and melanin deposits between them.

All cases were treated with surgical excision. Tumors adhered to skin were excised with overlying skin (Fig. 2). No recurrences were noted during the follow-up period. The demographic and clinical characteristics of our patients are presented in Table I.

Discussion

The report of Malherbe and Chenantais² in 1880 was the first to describe pilomatricoma as a benign tumor of the skin. They considered it to be derived from sebaceous glands and arising in the subcutis and used the term “calcifying epithelioma” for this lesion. In 1961, Forbis and Helwig³ renamed this tumor as pilomatrixoma to correctly describe its origin from hair follicle matrix cells in their study with 228 such tumors. In 1977, the tumor was renamed as pilomatricoma, which is etymologically more correct⁴.

Pilomatricoma has biphasic age distribution. It primarily affects children and adolescents, with greater than 60% of cases occurring before the third decade (40% of cases before age 10 and 60% before age 20). The second peak occurs

Table I. Demographic and Clinical Characteristics of Our Series

Case No.	Age	Sex	Location of tumor	Tumor size (cm)	Clinical presentation	Treatment
1	8	F	Left forearm	1.5x1	Hard mass adhered to skin for 9 months.	Excision with covering skin
2	14	M	Left infraorbital	1x0.5	Hard enlarging mass for 6 months that was not adhered to skin.	Excision
3	8	M	Right forearm	1.5x1	Hard mass not adhered to skin for 2 years.	Excision
4	8	F	Left infraorbital	2x1	Hard mass appeared after an abscess that drained 5 years before. The tumor was adhered to skin.	Excision with covering skin
			Inferior to left medial cantus	0.8x0.3	Hard mass for 2 years, not adhered to skin.	Excision
5	13	F	Right infraorbital	2x1	Hard nodule for 4 years that was adhered to skin.	Excision with covering skin
			Left temporal	0.5x0.4	Hard nodule for 3 years. The tumor was not adhered to skin.	Excision
6	10	F	Right posterolateral neck	1x1	Hard, painful nodule for 1 year. The tumor was not adhered to skin.	Excision
			Left arm	0.5x0.5	Hard, painful nodule for 9 months. The tumor was not adhered to skin.	Excision

between the sixth and seventh decades. Although it is the second most common benign skin tumor in childhood¹, pilomatricoma is often not considered in the differential diagnosis of head and neck masses. This is probably due to the relative rareness of reports on pilomatricoma in the literature. A female preponderance has been reported, with a male:female ratio of 2:3⁵. In our series, there were four females and two males, or a ratio of 2:4. However, the relatively small number of our patients restricts producing a reliable rate. All races are affected, though most of the patients in the literature are white^{5,6}. Pilomatricoma does not show genetic inheritance and it is a solitary tumor in the vast majority of cases. However, in 2-3.5% of reported cases, multiple tumors occur concomitantly⁷. In our series, half of the patients (50%) had more than one tumor. This high rate of multiplicity shows discordance with the previous literature. A further study reviewing a wide series of Turkish patients may disclose this discordance. This tumor can also be familial and associated with Gardner syndrome, myotonic dystrophy, sarcoidosis, skull dysostosis, Rubinstein-Taybi syndrome, and Turner syndrome^{8,9}. None of our patients had any other associated systemic disease.

Most pilomatricomas are located in the head and neck region⁹, followed in decreasing frequency by upper limb, trunk, and lower extremity. In the head and neck, cervical, temporal, eyelid, and preauricular regions are the most frequently reported locations⁸. No such tumor has been reported in regions that are deficient in hair. In our series, six of the total nine tumors were in the head and neck region. Of them, four were in the periorbital area. This distribution of pilomatricoma was consistent with the previous literature.

Clinically, pilomatricomas present as solitary, hard, painless, well-circumscribed dermal or subcutaneous masses measuring 0.5 to 3 cm in diameter. These lesions, which may be soft and cystic-like early in the course, are characteristically rock-hard when completely developed. They are usually asymptomatic; however, pain may be elicited when pressure is applied. In our series, only one patient, who had two lesions, complained of pain. In the other patients, the tumors were painless.

Pilomatricomas have a unique type of pivot movement with unilateral pressure that helps to identify these lesions: the disk-shaped

nodules are anchored deep in the dermis or subcutis, and when pressure is applied to one end, the other springs up like a teeter-totter. In 1978, Graham and Merwin¹⁰ described the tent sign that has been used to evaluate pilomatricoma clinically. To discern the multifaceted nature of the lesion, the covering skin may be stretched¹¹. Rigid and malignant nodules that are attached to the deeper skin would not exhibit this pivoting characteristic. Telangiectasias, a hemangioma-like darkish red shade, or vivid blue or black discoloration on the skin overlying the pilomatricoma may be seen^{5,12}. Development is usually slow and may take months to years. No lymphadenopathy has been reported at the time of diagnosis. No clinical or histological sign of malignancy or lymphadenopathy was detected in our cases.

Radiologic imaging is considered of little diagnostic value for pilomatricoma. Because of their rather superficial location, routine radiographic imaging seems unnecessary. However, calcified foci may be detected with plain X-ray films. Larger tumors or unusual locations may necessitate cross-sectional imaging for planning the proper surgical intervention. None of our patients required imaging because of the superficial localizations.

Differential diagnoses of pilomatricoma include: epidermal inclusion cyst, ossifying hematoma, giant cell tumor, dermoid cyst, chondroma, foreign body reaction, degenerating fibroxanthoma, eccrine spiradenoma, hydrocystoma, metastatic bone formation, osteoma cutis, osteochondroma, trichoepithelioma, trichilemmal cyst, basal cell epithelioma, steatocystoma simplex, cylindroma, branchial cleft cysts, and cervical adenitis secondary to mycobacterium^{7,13,14}.

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 shadow 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 shadow cells in the center have lost their nucleus and thus have a central unstained area. Calcification, especially in the form of basophilic stippling, is seen in many of the shadow cell regions, and overt calcification can be identified in a minority of lesions. Foreign body giant cell inflammation can

also be identified in regions where keratinized debris is abundant. The overlying epidermis is usually normal; however, the tumor is separated from the epidermis by a layer of fibrous tissue that may create the appearance of adherence to the overlying skin⁸. Often, multiple lobulations of the basaloid component project into the adjacent dermis in which the lesion resides. This lobulated architectural pattern, in association with the biphasic basaloid and shadow cell population, allows for the identification of the tumor as pilomatricoma¹⁵. Histopathologic findings of our patients were all typical for pilomatricoma.

Because spontaneous regression is not expected, the treatment of choice is complete surgical excision. Overlying skin may be tightly adhered to the tumor, requiring excision of that skin along with the tumor. However, the tumor never adheres to deep structures, and separation from the underlying subcutaneous tissue is commonly easy. If the boundaries are not well-defined or the tumor is adhered to the surrounding tissues, a malignancy should be suspected. In these cases, wide excision with margins of 1 to 2 cm is recommended^{16,17}. The recurrence rate is extremely low and most recurrences occur from incompletely excised tumors. We applied excision of the covering skin with the tumor in three lesions, which were found fixed to the overlying skin.

In conclusion, pilomatricomas are hard, painless, slowly progressing tumors that are located subcutaneously. The common locations are the head-neck region, especially periorbital, and the upper extremities. These benign tumors are typically seen in children. Histopathologic characteristics are distinctive with basaloid cells, ghost cells, foreign body giant cells, and intralesional calcifications. The preferred treatment is complete excision. If the tumor is adhered to overlying skin, excision should include the skin. This method is remedial. Recurrence of pilomatricoma is not common. These tumors should be included in the differential diagnosis of superficial pediatric head and neck masses by any physician concerned with pediatric tumors.

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