Eyelid Pilomatricomas in Young Adults: A Report of 8 Cases

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GOAL

To review the clinical and histopathologic features of pilomatricoma of the eyelids and eyebrows

OBJECTIVES

Upon completion of this activity, dermatologists and general practitioners should be able to:

1. Discuss the most common characteristics of patients with pilomatricoma.
2. Describe the clinical presentation of pilomatricomas.
3. Discuss the histopathology of pilomatricomas.

CME Test on page 34.

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Pilomatricomas are common benign childhood tumors, which usually occur in the head and neck region. We present 8 patients and review the clinical and histopathologic features of pilomatricoma of the eyelids and eyebrows in young adults. From 1992 to 2000, diagnosis of eyelid pilomatricoma was performed in 8 young adults (13–36 years). The female-male ratio was 3:1. The mean age at presentation was 22 years. Each of our 8 patients had a single tumor, 4 on the upper eyelid and 4 on the eyebrow. Ossification existed in 4 cases. No tumor recurrence has been noted at 3.9 years. Pilomatricomas are uncommon tumors on the eyelid and brow region of young adults. These lesions are frequently misdiagnosed when evaluation is based on clinical evidence alone: only 4 of our cases were correctly diagnosed prior to excisional biopsy.

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The first complete work on pilomatricomas was published by Malherbe and Chenantais[1] in 1880. They described calcifying epitheliomas, initially thought to be tumors of the sebaceous glands. In 1949, Lever and Griesemer[2] suggested that the origin of the tumor was hair matrix cells. The term *pilomatrixoma* was proposed by Forbis and Helwig[3] in 1961, thus avoiding the word *epithelioma*, which carries the connotation of malignancy. The term was later corrected to *pilomatricoma*, to be more etymologically correct.[4] Locally aggressive behavior in some cases of pilomatricoma was first suggested by Gromiko[5] in 1927. The malignant variant of pilomatricoma was not seriously considered

Clinical Features of Patients Examined in This Study*

Patient No.	Age, y	Sex	Location	Size, cm	Evolution Time, mo	Preoperative Clinical Diagnosis	Histology	Postoperative Follow-up, y
1	24	F	Left upper eyelid	1.0	1	Epidermal cyst	CH, calcified eroded	9
2	13	F	Left upper eyelid	1.2	7	Pilomatricoma	Calcified, FBGC	6
3	24	F	Right eyebrow	0.4	3	Epidermal cyst	Calcified	6
4	16	F	Right eyebrow	1.2	8	Epidermal cyst	Increased vessels	3
5	14	F	Right upper eyelid	2.0	6	Dermoid cyst	Increased vessels	3
6	36	M	Right eyebrow	2.0	12	Pilomatricoma	Calcified	2
7	29	M	Left upper eyelid	0.7	5	Pilomatricoma	—	1
8	18	F	Right eyebrow	0.9	60	Pilomatricoma	Increased vessels	1

*F indicates female; M, male; CH, cutaneous horn; FBGC, foreign body giant cells.

until a case was reported in 1980 by Lopansri and Mihm.[6] Pilomatrical carcinoma is rare.

Pilomatricoma is a subepidermic tumor that arises from the external radicular shaft of the hair root[7] as a firm solitary lesion of the face and upper extremity. It generally measures 0.5 to 3.0 cm in diameter and is typically found in young children. Pilomatricoma may present not only as a benign lesion or low-grade malignant lesion with a tendency to recur locally but also as a highly malignant tumor.

Patients and Methods

Eight cases of patients with pilomatricoma seen between 1992 and 2000 were analyzed retrospectively (Table). The age, sex, location and size of the tumor, evolution time, clinical diagnosis, histology, and follow-up time after surgical treatment were noted.

Results

The female-male ratio was 3:1. The female age range was 13 to 24 years, and the mean was 18 years. The mean male age was 32.5 years. The mean age at first presentation was 22 years, but in many patients, the history went back some time. The time to diagnosis ranged from 1 month to 5 years, although 50% of the patients had had the tumor for 6 months or less. No patient had familial or hereditary factors.

The main patient complaints were of a tumor or swelling under the skin. All patients had a single tumor, located on the upper eyelid (Figure 1) in 4 cases and on the eyebrow in the other 4 cases. Five tumors were located on the right, and 3 were on the left. The eyelid tumors were located 1 on the inner, 2 on the middle, and 1 on the outer portion of the eyelid, with none affecting the free-edge. The eyebrow tumors were all in the middle part of the right eyebrow.

All 8 tumors were clearly demarcated and fixed to the skin. The texture of the tumor was cystic in 5 cases (one had a keratotic appearance of a cutaneous horn) and firm-calcified in the other 3 cases.

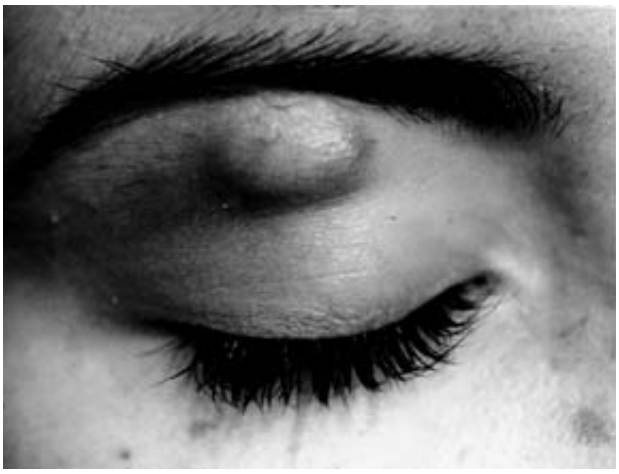

Figure 1. Preoperative appearance of a firm, well-defined, subcutaneous mass (1.2 cm in diameter) arising from the medial third of the eyelid of patient 2.

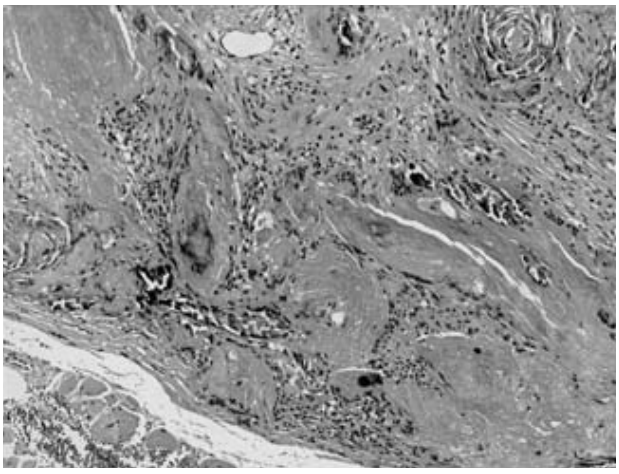

Figure 2. Tumor in patient 6 containing abundant keratin surrounded by epithelial cells arranged in bands. There are areas of focal calcification (H&E, original magnification ×200).

In one case, the tumor was particularly superficial, calcium eroded through the surface and extruded. The sizes of the tumors at examination varied from 0.4 to 2 cm in diameter, with a mean size of 1.2 cm. Rapid enlargement of the tumor was referred to by one patient as a result of internal bleeding, and subsequent hemosiderin pigmentation was seen, simulating a pigmented lesion. The color of the skin covering the tumor was normal in one case, purple in one case, brown in one case, and yellow-white in the other 5 cases.

Four of the tumors (50%) were correctly diagnosed preoperatively, and 4 (50%) were diagnosed simply as epidermal and dermoid cysts. Treatment was by excisional biopsy. No pilomatricoma behaved aggressively. No tumor recurrence has been noted in our patients after a mean follow-up of 3.9 years.

On macroscopic examination, all tumors were encapsulated and solid. Microscopically, shadow and basophilic cells were present in different proportions in all cases (Figure 2): partial or definitive ossification in 4 cases (50%), focus of horn pearl or keratin in one case, foreign body giant cells in stroma in one case, and an increased number of blood vessels in 3 cases.

Comment

Pilomatricomas appear at any age, with peak presentation bimodally in the first or sixth decade, but they are uncommon in young people such as the 8 patients we present (13–36 years old). The most common site in our patients was the head and neck region, as has also been reported by other authors. The periorbital region is involved in 17% of the cases.[8] All of our patients' tumors were located on the upper eyelid or eyebrow, mostly on the right side (62.5%) and on the middle portion of the eyelid/eyebrow. It has been suggested that the distribution of pilomatricoma corresponds to the density of hair follicles at a particular site.[9] The hairy scalp has about one half the density of follicles of the face, which is the most richly supplied area of the whole body.

Pilomatricomas are usually solitary tumors. Multiple lesions are found in 3.5% of cases[10] and may be associated with myotonic dystrophy. They also may be related to sarcoidosis or Gardner syndrome.

Pilomatricoma presents as a hard nodule, either deeply subcutaneous and invisible or superficial with possible erosion through the skin surface. This may lead to a false diagnosis of malignancy. The diagnosis is rarely made clinically. Differential diagnosis must be done with other benign and malignant conditions that may appear as solitary firm skin nodules, especially those that occur on the head and neck. In the eyelid, differential diagnosis most frequently includes dermoid cyst in young patients and epidermal cyst in adult patients. Other differential diagnoses include benign tumors (chalazion, epidermoid cyst, and keratoacanthoma) and malignant tumors (basal-cell carcinoma and metastasis).

Histopathologically, pilomatricoma is characteristically a mass composed of viable basaloid cells; shadow cells; and foci of calcification and, occasionally, ossification. The shadow cells represent areas of necrosis of the previously viable basal cells. Foci of calcification and ossification gradually develop in the necrotic areas. Small blood vessels also are increased in number, and the overlying

dermis and epidermis are atrophied. One of our patients presented with these characteristics, which are signs of a potentially malignant form. Rarely, a pilomatricoma may perforate the epidermis,[11] which may have been the cause of one of our cases developing a cutaneous horn. The reported incidence of calcification ranges from 69% to 85%, and bone metaplasia is observed in 15% of cases, presumably secondary to conversion of fibroblasts into osteoblasts.[3] The mechanism of occurrence of calcification and ossification in pilomatricoma remains unclear.[12] In our series, 4 cases (50%) showed calcification, but no bone metaplasia was observed.

Spontaneous regression has never been observed. Complete surgical excision, including the overlying skin, is the treatment of choice, and no patient has developed secondary lesions at a new site. Reported recurrence rate is 2.6%.[3] Because the circumocular area is a cosmetically sensitive area, correct clinical diagnosis as a benign tumor is important in its treatment, especially in women, the population most frequently affected in young people.

Pilomatrical carcinoma is an extremely rare tumor occurring in middle-aged patients, more often in men (male-female ratio of 4:1 in contrast to 2:3 for benign lesions[13]) and more often in larger lesions (up to 20 cm). They can metastasize to the lungs, bone, and viscera with a subsequent poor outcome.[14]

Conclusion

We described 8 cases of eyelid pilomatricomas, emphasising the number and ages of the patients affected (13–36 years). This tumor is commonly misdiagnosed preoperatively (75%) when evaluation is based on clinical evidence alone, and should always be considered in the differential diagnosis with other eyelid lesions.[15]

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