A retrospective investigation of twenty-five cases with periocular pilomatrixoma

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Research Article

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Abstract

Purpose

Pilomatrixoma is an uncommon benign skin tumor, developing from hair follicle matrix. It consistently presents in the head and neck region patients with pediatric predominance, from infancy to senior. Thus far, few case series studies have been focused on the periocular lesions that involve the eyelid and eyebrow. In the present study, we carried out a case series with 25 patients of periocular pilomatrixoma.

Methods

Records from the Hospital Information System (HIS) of our hospital were reviewed retrospectively between the years 2009 and 2020 to search for all patients that were diagnosed as pilomatrixoma of eyelid or eyebrow. All patients received excision in the Department of Ophthalmology and histological examinations in the Department of Pathology. Patients' information and characterization of tumors features were recorded and analyzed.

Results

These clinical conditions were mostly found in female patients in their early twenties. All tumors developed at upper eyelids or eyebrows and both right and left sides were equally affected. Basophilic cells and shadow cells were the basic components of the lesions, with or without calcification and foreign body giant cells. Complete surgical resection was the essential therapeutical approach for this disease and no recurrence or malignant transformations were noticed upon follow-up.

Conclusion

The findings of current case series analysis are in concordance with the clinical and histopathological features of periocular pilomatrixoma.

Introduction

Pilomatrixoma, a rare hair follicle matrix-derived benign tumor, always presents in the head and neck in pediatric populations[1]. In 1880, Malherbe and Chenantais first described it as "calcifying epithelioma of Malherbe". In 1949, Lever and Griesemer found the calcifying epithelioma arising from immature hair matrix cells[2]. In 1961, Forbis and Helwig used the term "pilomatrixoma" instead of previous "calcifying epithelioma" since they noticed the tumor developing from primitive epidermal germ cells that are differentiating toward hair matrix cells[3].

With the incidence of around 1% among benign skin lesions[4], pilomatrixoma is an uncommon lesion that presents at periocular regions, such as eyelid and eyebrow. Clinically, the diagnosis of periocular pilomatrixoma is somehow difficult given the similarity of their appearance to other dermal masses, such as chalazion[5, 6]. Therefore, histological examinations upon complete surgical resection are essential for the confirmation of such conditions[1].

Till now, most of the periocular pilomatrixoma are reported as case reports around the world. In English literatures, we noticed that two case series investigations of more than 10 patients have been reported in Israel[7, 8]. Another study of 13 cases of pilomatrixoma of the ocular adnexae was reported in German[9]. In the present study, we aimed for the first time to present a relatively large case series of periocular pilomatrixoma from one ophthalmic clinic in order to show the clinical and histopathological characteristics of such clinical condition in China.

Methods

Records from the Hospital Information System (HIS) of our hospital were reviewed retrospectively between the years 2009 and 2020 to search for all patients that were diagnosed as pilomatrixoma of eyelid or eyebrow. All patients received excision in the Department of Ophthalmology and histological examinations in the Department of Pathology. Patients' information includes gender and age, characterization of tumors features includes location, gross appearance, size (diameter in large), development duration, histopathological characteristics were recorded and analyzed. For histopathological examinations, all excised biopsies were sectioned and stained with hematoxylin and eosin. This study was adhered to the principles listed in the Declaration of Helsinki and was approved by the Ethics Committee of our hospital.

Results

Demographic data of patients

Totally 25 patients of periocular pilomatrixoma received excision in our department from 2009 to 2020. Age at the time of diagnosis ranged from 3 to 70 years (mean 25 years, median 15 years). There were 14 female patients (56 %) and 14 patients (56 %) younger than 20 years. The demographic data of all patients were listed in Table 1.

Table 1
The demographic data of patients and clinical characteristics of lesions

No.	Gender	Age (years)	Duration (months)	Location	Size (mm)	Calcifications	Giant cells
1	F	4	3	Right upper eyelid	4	+	+
2	F	11	5	Right upper eyelid	5	+	+
3	F	12	4	Right upper eyelid	3	+	+
4	M	22	2	Left eyebrow	3	+	+
5	F	7	24	Left eyebrow	5	+	-
6	F	15	2	Left eyebrow	2	+	+
7	F	51	4	Right eyebrow	5	+	+
8	F	30	4	Left eyebrow	4	+	+
9	M	44	3	Right eyebrow	5	+	+
10	F	10	2	Left eyebrow	6	+	+
11	F	40	12	Left upper eyelid	5	-	-
12	M	49	3	Left eyebrow	4	+	-
13	M	65	60	Left eyebrow	3	-	+
14	M	9	12	Right eyebrow	5	+	-
15	M	13	12	Right upper eyelid	5	+	+
16	F	61	2	Right upper eyelid	5	+	+
17	M	3	6	Right eyebrow	5	+	+
18	F	70	2	Right upper eyelid	8	+	+
19	M	4	48	Left eyebrow	5	+	-
20	M	9	1	Right eyebrow	6	+	+
21	F	13	1	Right upper eyelid	5	+	-
22	M	48	6	Right upper eyelid	4	+	+
23	F	10	3	Left upper eyelid	5	+	-
24	M	24	3	Left upper eyelid	8	+	+
25	F	4	6	Left upper eyelid	4	+	+
F: female; M: male; +: present; -: absent.							

Clinical characteristics of lesions

The duration of lesion development ranged from 1 to 60 months (mean 9.2 months, median 3.5 months). Among the 25 patients, 8 (32 %) presented lesion at right upper eyelid, 5 (20 %) at right eyebrow, 4 (16 %) at left upper eyelid and 8 (32 %) at left eyebrow. Thus, lesion on the right eye was involved in 13 (52 %) cases and the eyebrow was also involved in 13 (52 %) cases (Table 1).

All the patients have presented a slowly growing, spherical, well-circumscribed single nodule. Most of the nodules had normal overlying skin, except only 1 case with ulcerated overlying skin. The tumor size (diameter in large) ranged from 2 to 8 mm (mean 4.76 mm). None of the patients have presented multiple pilomatricomas at involved periocular sites (Table 1). The CT (computerized tomography) scan under nonenhanced mode indicated that all lesions were well-circumscribed with calcification (Fig. 1).

The follow-up histopathological observations confirmed the diagnosis of pilomatrixoma. All the tumors were composed of typical basophilic cells in the peripheral and shadow cells in the center (Fig. 2A). Meanwhile, calcifications (Fig. 2B) were found in 23 cases (92 %) and foreign body giant cells (Fig. 2C) were detected in 18 cases (78 %) (Table 1).

All the patients received complete excision in the Department of Ophthalmology of our hospital. No intraoperative or postoperative complications was occurred. Moreover, no recurrence or malignant transformations were observed upon follow-up.

Discussion

Periocular pilomatrixoma is indeed an infrequent clinical condition in ophthalmic clinics[10]. To the best of our knowledge, the present study analyzed the largest case series of periocular pilomatrixoma in English literatures in recent years.

Pilomatrixoma were mostly found in female patients in the first 2 decades of their lives[1]. Our findings here were in accordance with the conclusions before (female: 56 %,younger than 20 years: 56 %). The periocular lesions mostly develop at upper eyelids and eyebrows, without left or right predominant. We also found all tumors located at upper eyelids or eyebrows and both right and left sides were equally affected (right vs. left: 52 % vs. 48 %)[7–9].

In general, the gross appearance of pilomatrixoma is a kind of firm, well-circumscribed nodule with or without epidermal ulceration. Although the size of tumors varied from each other, most of the periocular pilomatrixomas were smaller than 1 cm[11]. Upon CT scan, the lesions are always well-circumscribed and isodense relative to adjacent skeletal muscle with different degrees of calcification[12].

The tumors were composed of two kinds of typical tumors: basophilic cells at peripheral site and shadow cells at central site. Other pathologic features including calcification and foreign body giant cells[13]. The incidence of calcification ranges from 69 % to 85 % as reported before and the foreign body giant cells

are always located around the calcified epithelial cell mass[3, 10]. We observed 23 cases (92 %) had calcification and 18 cases (78 %) had foreign body giant cells in their excised biopsies.

Recently, multiple pilomatricomas were noted, which are mostly related to systemic diseases like Gardner syndrome (GS), Turner syndrome (TS), etc[14]. However, to the best of our knowledge, no multiple pilomatricomas has been reported to derive from ocular adnexae till now. In our analyzed cases, no multiple pilomatricomas was found, either.

Conclusion

In summary, we analyzed 25 Chinese patients diagnosed as periocular pilomatricoma from one ophthalmic clinic in the past decade. It was confirmed that the demographic and clinical characteristics of this disease were consistent with the current knowledge.

Declarations

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Ethics approval: This study was adhered to the principles listed in the Declaration of Helsinki and was approved by the Ethics Committee of Jiaxing Hospital of Traditional Chinese Medicine.

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Figures

Figure 1
CT scan under nonenhanced mode indicated well-circumscribed lesions were located at right eyebrow (A) and left upper eyelid (B).

Figure 2
Histopathological features of periocular pilomatricoma (stained with hematoxylin and eosin). (A) Basophilic cells (white arrow) and shadow cells (black arrow). (B) Calcifications (white stars). (C) foreign body giant cells (white arrows).