CASE REPORT

Uncommon benign tumour of the skin: infraorbital pilomatricoma

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SUMMARY

Pilomatricoma is a relatively rare tumour of the skin derived from primitive basal cells of epidermis that differentiate into hair matrix cells. It comprises approximately 1% of all benign skin tumours. The most common sites for pilomatricomas are the head, neck and upper extremities. A 44-year-old woman, admitted to our department, underwent an orthopantomography examination, which revealed a well-circumscribed 1.6×1.4 mm radiopaque mass superimposed between the medial wall of the right maxillary sinus and lateral wall of the nasal cavity. Clinically, it was firm and mobile, and remained attached to the underlying skin when palpated. A cone beam CT scan was performed to ascertain its location and anatomic relations. Subsequently, it was totally excised under local anaesthesia. Histopathological evaluation of the lesion reported it to be a pilomatricoma.

BACKGROUND

Malherbe's calcifying epithelioma, or pilomatricoma, is an uncommon, painless, hard and slowly progressing benign lesion arising from epithelial hair matrix cells of the hair bulb. It is commonly located in the neck, cheek, periorbital region and scalp. Most cases are seen in children and adolescents. Pilomatricomas are the second most commonly excised superficial masses in children, after epidermoid cysts and excluding lymph nodes. In the periorbital area, they usually arise from the eyelids and eyebrows.^{1 2} Since these are uncommon tumours and their clinical features have rarely been reported, diagnosis can only be made with clinical examination followed by histopathological confirmation.³

Pilomatricomas have rarely been reported in the literature, however, Moehlenbeck *et al*⁴ reviewed 140 000 skin tumour cases and noted that pilomatricomas represented 0.12% of these cases. Kuang-Yu⁵ found 10 pilomatricomas among 22 000 surgical specimens, while Highman and Ogden⁶ found 11 tumours among 24 185 specimens. Lever and Griesemer⁷ reported 15 pilomatricomas among 29 488 surgical specimens. According to their study, the incidence of pilomatricoma is estimated to be 1 in 2000 surgical specimens and this result has also been verified in the studies carried out by Kuang-Yu,⁵ and by Highman and Ogden.⁶ In the aforementioned study, by identifying clinical and histological features in 15 cases, comments were reviewed on differentiation of such tumour from the epidermal cyst and on histogenesis.

The characteristic histological appearance is of shadow cells and basaloid cells. If complete surgical excision is performed, recurrence is rare.¹ In this case report, an uncommon benign tumour of the skin, infraorbital pilomatricoma, including its histopathological features and treatment method, is described.

CASE PRESENTATION

A 44-year-old woman presented to the Department of Oral and Maxillofacial Surgery, Faculty of Dentistry, Istanbul Aydin University, for a painless, removable and palpable mass under her right eye, towards the middle of her face, for 25 years.

According to clinical and radiological examination of the patient, a mobile, painless and palpable hard mass was detected around the right infraorbital foramen area. An orthopantomography image revealed a partially oval hypodense lesion in the infraorbital foramen (figure 1). A uniformly bounded radiopaque mass with a lobular appearance was seen in cone-beam CT (CBCT). According to CBCT, there was no connection between the bone and lesion—it appeared as a mixed-type lesion (figure 2).

DIFFERENTIAL DIAGNOSIS

The differential diagnosis includes multiple pilomatricomas, myotonic dystrophy, Gardner syndrome, Steinert disease, Turner syndrome.³ Pathologies such as trichoblastic carcinoma, basal cell carcinoma, dermoid cyst, sebaceous cyst, eyelid cyst, eyelid tumour, retention cyst, atheroma, abscess, cavernous vascular tumours, and benign hair follicle tumours such as trichoepithelioma and trichoblastoma, can be considered.^{3 8} Although pilomatricomas usually grow slowly, rapid growth has also been reported,⁹ also, pilomatricomas may resemble keratoacanthomas. They rarely show malignant transformation to pilomatrix carcinomas.¹⁰

Figure 1 Preoperative orthopantomography image of the patient.

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Figure 2 Preoperative cone-beam CT images of the patient.

TREATMENT

After careful evaluation of the clinical and radiographic findings, it was decided to excise the lesion with prediagnosis of peripheral osteoma or calcified fibroma. A skin incision was made in the infraorbital area up to the removable and palpable mass, following blunt dissection of subcutaneous tissues. The fibrous capsule around the mass was also dissected and no relationship with the bone was seen. Subsequently, the mass taken from within the soft tissue was observed to be a totally calcified and bony structure (figure 3). The infraorbital neurovascular sheath was preserved during the operation and the deep tissues were closed with 3/0 resorbable sutures; the skin was closed using 5/0 silk sutures (figure 4).

The bony mass was sent for histopathological examination with the prediagnosis of peripheral osteoma and calcified fibroma. Microscopic examination showed basophilic cells and enucleated ghost cells, and that the mass was formed by keratinous debris. There were numerous foci of calcification, giant cells and vessel-rich bone marrow, and the fibrous capsule was monitored around (figures 5 and 6). The final diagnosis was pilomatricoma.

OUTCOME AND FOLLOW-UP

After an uneventful 7-day postoperative period, the sutures were removed and, at 24-month follow-up, the patient had no symptoms and no recurrence was detected (figures 7 and 8).

Figure 3 Intraoperative images and exposed mass.

Figure 4 Primary closure.

DISCUSSION

Pilomatricoma is a benign lesion, formed by the hair follicle matrix of the hair root. It was first described by Malherbe and Chenantais¹¹ as calcifying epithelioma—a benign subcutaneous tumour arising from the sebaceous gland. Later ultrastructural and electron microscopic studies showed that these lesions in fact arise from pilomatricoma cells. Forbis and Helwig¹² proposed the term pilomatricoma due to the lesion's histogenesis. Pilomatricomas are considered to be ectodermal in origin. This theory supports the idea that pilomatricomas arise from the germinal matrix of hair. In 1922, Dubreuilh and Cazenave¹³ described the unique histopathological characteristics of this neoplasm, including islands of epithelial cells and shadow cells. In 1961, Forbis and Helwig¹² proposed the term pilomatricoma to describe the condition to avoid a connotation of malignancy and to denote its origin from hair matrix cells.

Despite being better defined, pilomatricomas continue to be frequently misdiagnosed and are not usually considered in differential diagnoses. They typically present as a superficial, firm, solitary, slow-growing, painless mass of the dermis.¹ The overlying skin may exhibit a bluish discolouration or ulceration. Treatment consists of surgical excision. Recurrence is rare after complete resection. Malignant pilomatricomas have rarely been reported.³

Pilomatricoma is usually a solitary lesion affecting all ages, however, it is most often seen in children and young adults.^{14–16}

Figure 5 Histopathological image (red arrow: masses of pillar cells derived from epithelial cells; green arrow: large areas of trabecular calcification; black arrow: vessel-rich fatty bone marrow).

Lever and Griesemer⁷ reported that 9 from 15 cases occurred before the age of 20 years, and 4 cases occurred between the ages of 21–50 years. It has been reported in 40% of patients in the first decade of life and 20% of those in the second decade of life.¹⁷ In a retrospective study carried out by Lan *et al*,¹ the rate of incidence in those under 10 years of age is 26.8%, and the rate of incidence in those between the ages of 11 and 20 years, 22.3%, with the rate of incidence in patients between the ages of 50–70 years being 27.9%. It is reported that these results do not comply with the literature results reviewed during the earlier mentioned retrospective study.

Pilomatricomas are mostly (40–77%) seen in the head and neck region, followed in frequency by the upper extremities, the trunk and, finally, the lower extremities.^{14–18} Up to 10–17% of cases attach to the periocular tissues and up to 5% are multifocal. The most frequent occurrence in the head and neck area is on the neck, followed by the frontal, temporal periorbital and preauricular areas.^{19–20} Lan *et al*¹ reported occurrence being on the neck and cheeks, followed by scalp and periorbital area. The most common involved site in the paediatric group (under 18 years) was the neck and cheek, and in adult patients, the scalp and neck.^{19–20}

Pilomatricomas generally present as subcutaneous red to blue masses that are fairly well circumscribed, mobile and

Figure 6 Histopathological image (blue arrow: basophil masses and keratinous debris).

Figure 7 Follow-up (12 months) image of the patient.

firm-to-gritty on palpation.²¹ Clinical features as documented by Duran and de Buen,²² and later also by Perez and Nicholson,²³ should raise clinical suspicion—they include onset in childhood or early adulthood, average size of 10 mm or less, consistency ranging from firm to cystic, moderate pattern of growth, pink to purple hue with subepithelial yellowish tinge and intact overlying skin with telangiectatic vessels. Pilomatricomas are usually solitary nodules also between 2 and 10% can be seen as multiple. The diameter of pilomatricomas vary between 0.5 and 3 cm in most cases. These tumours slide slowly over the underlying layers. They are generally asymptomatic and develop slowly in a process taking months and years.³

The diagnosis of pilomatricoma is only made based on clinical features, and the correct preoperative diagnosis rate varies between 0% and 30%. The value of radiological imaging in the diagnosis of pilomatricoma is not significant.¹⁶ In MRI examination, T2-weighted, high signal band imaging, shows a band formed with basoloid cells with further evidence in the histological examination.²⁴ Ultrasonography is also considered a relatively quick and non-invasive method for calculating the depth of larger masses.²⁵

Malign transformation of pilomatricomas is rare. Less than 20 cases have been reported in the literature.^{15–16} There has been incidence of malign pilomatricoma in middle-aged and older patient populations.³ It is in the histological features of pilomatricoma, such as proliferation of active basoloid cells, that atypical mitosis is seen, along with nuclear polymorphism, invasion of blood vessels and infiltration of the underlying structures. The tumour is seen to be fairly well circumscribed and firm-to-gritty in histopathological examination. It comprises of a number of islands of epithelial cells, the periphery of which are lined with basophil cells and in the centre of which shadow cells are lined. Calcification occurs in 75% of cases. Eosinophilic keratinous material layers are seen extensively in necrotic sites and this material may form a foreign giant cell reaction. Basal or squamous cell epitheliomas can be considered a type of skin or subcutaneous cyst in histopathological differential diagnosis.^{3–8–9}

Fine-needle aspiration cytology has been identified for the preoperative diagnostic examination. However, if there are no shadow cells in aspiration, the results may be incorrect.^{15–16} Since spontaneous regression has not been observed, standard treatment of pilomatricoma is complete surgical excision. Wide

Figure 8 Follow-up (24 months) cone-beam CT images.

excision performed by keeping a 1–2 cm safe border is recommended to reduce the risk of malignant changes and local recurrence.¹⁵ Sometimes, the skin overlying the tumour must be excised due to the tumour attaching to the dermis.^{15 16} Recurrences are rare after surgery, with an incidence of 0–3%. It must be differentiated from pilomatrix carcinoma in cases of repeated local recurrence.¹⁸ There have been rare cases describing distance metastasis of pilomatrix carcinoma.^{8 10}

Learning points

- ▶ Malherbe's calcifying epithelioma, or pilomatricoma, is an uncommon, painless, hard and slowly progressing benign lesion arising from epithelial hair matrix cells of the hair bulb.
- ▶ Despite being well defined, pilomatricomas continue to be frequently misdiagnosed and are not usually considered in differential diagnoses.
- ▶ Pilomatricoma is a benign tumour with atypical forms and, unfortunately, no tumour-specific diagnostic features exist, necessitating careful histopathological examination.
- ▶ Treatment of pilomatricoma is total surgical excision. Recurrence is rare.

Contributors BE and SK were responsible for treatment of the patient. BE, SK and TBA followed up of the patient. SK, BE and TBA were responsible for preparation of

manuscript. NA contributed to pathological evaluation. BE was responsible for critical revision.

Competing interests None declared.

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