CASE REPORT

Complicated Giant Pilomatrixoma of the Parotid Region

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Abstract

Objective Pilomatrixoma (calcifying epithelioma of Malherbe) is a benign soft tissue tumour arising from dermis or subcutaneous tissue which should be considered in differential diagnosis of preauricular lesions especially when skin fixation is present.

Case Report Twenty-three year old male referred to our clinic with complaint of left preauricular swelling over 18 months which enlarged and became painful in the last 2 months. Because the lesion showed signs of infection, surgery was planned after medical therapy was completed. FNAB suggest pleomorphic adenoma as preliminary diagnosis. US or MRI showed no specific feature.

Treatment and Prognosis Total excision, superficial parotidectomy with facial nerve sparing was performed after regression of infectious signs. Postoperatively no recurrence was detected.

Conclusion Pilomatrixomas are benign tumours but have diagnostic difficulties according to clinical and cytologic findings. This rare lesion should be kept in mind to avoid misdiagnosis as malign parotid tumours, particularly in the presence of skin change.

Keywords Pilomatrixoma · Giant · Complication · Parotid region · Skin neoplasm

Introduction

Pilomatrixoma or calcifying epithelioma of Malherbe is a benign, cutaneous tumour originating from pluripotent precursor cells of hair follicles. Malherbe and Chenantais described pilomatrixoma as a benign tumour of sebaceous glands in 1880. But later, it was understood that the origin of the tumour is from hair follicles indeed. [1].

Pilomatrixoma is commonly located at head and neck region especially at periorbital and preauricular areas. Although it can be seen in all ages and sexes; it is most often seen in first two decades of life and females. Pilomatrixoma is a relatively uncommon lesion but clinically it can be mixed up with other lesions and cause misdiagnoses. Differential diagnosis of tumour is difficult because familiarity to lesion by clinicians is insufficient and few publications are available in English literature [2–5].

Clinical presentation is a solid mass in dermal or subcutaneous tissue and it can be misdiagnosed as malign neoplasm especially in head and neck region. Because of the difficulty in differential diagnosis particularly in preauricular region, further imaging techniques as CT or MRI is necessary. Fine needle aspiration biopsy (FNAB) is helpful in preoperative diagnosis of pilomatrixoma but possible false diagnosis as carcinoma should be kept in mind [4, 5].

Malign transformation of pilomatrixoma is very rare so treatment with effective curettage or simple excision is sufficient. Additional treatment is unnecessary if no recurrence is seen in initial tumour [6].

In our case, a mass in parotid region complicated with skin changes. We emphasize that pilomatrixoma as well as malign parotid gland tumours must be considered in preliminary diagnosis. Increase in awareness of this uncommon lesion, surgery with facial nerve sparing and repair of

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possible skin defect in the same session are the goals of this article.

Case Report

Twenty-three year old male referred to our hospital with left preauricular swelling which started 1.5 years ago and enlarged during last 2 months. Over an infected mass, fistulisation to cheek skin with purulent discharge was detected. Patient was prescribed 10 days of oral antibiotic following 10 days of intravenous treatment. After this long treatment, infection and purulence regressed and physical examination revealed solid painless mass measuring about 4×5 cm over left masseter muscle which was freely mobile at base and fixed to overlying skin. Skin over the lesion showed hyperemic colour changes; serous discharge was detected from fistule spontaneously and with pressure as well. Facial nerve functions were normal (Fig. 1).

Patient history showed no special feature except smoking (1.5 packages/day). Routine biochemical tests were in normal limits.

US imaging identified heterogenous solid tumoural mass containing multiple calcifications located inferiorly in left parotid gland. Neck MR reported as hypointense capsulated mass anterior to the gland with contrast enhancement in capsula and septa (Fig. 2).

In FNAB, proliferated epithelial cells, macrophages, stromal fragment with seroproteinous ground suggested pleomorphic adenoma and according to the biopsy superficial parotidectomy approach with facial nerve sparing was planned.

This procedure was performed under general anesthesia after informed consent was taken from the patient. A superficial parotidectomy was completed with exposure of facial nerve. Total excision of the lesion along with the

Fig. 2 Coronal MR imaging. 4×3 cm mass with partial contrast enhancement anterior to *left* parotid gland

involved cheek skin with safe margin was done. Caseous material leaked from the lesion perioperatively. Skin defect in cheek, 2×2 cm, was closed by Z plasty to restore cosmetic form (Fig. 3).

Pathological examination of the specimen was reported as pilomatrixoma (Fig. 4). Postoperatively, sialoceles which was regressed with repeated aspiration and pressure dressing persisted for 4 weeks. Postoperative control in 7th month, no recurrence or residual tumour was detected (Fig. 5).

Discussion

Pilomatrixoma is an uncommon benign tumour arising from hair follicles. Several publications report its

Fig. 1 Preoperative clinical view of patient from *left* and *front* aspects. Fistulisation and hyperemia was seen in overlying skin

Fig. 3 Tissue specimen removed preoperatively

Fig. 4 **A** Tumour which is surrounded by fibrous tissue adjacent to the parotid gland (H and E, original magnification $\times 40$). **B** Tumour showing irregular blands of epithelial cells composed of basophilic cells and shadow cells (H and E, original magnification $\times 100$)

Fig. 5 Front and left lateral view of the patient at postoperative seventh month

incidence between 0.03 and 0.1 % [5]. It is commonly presented in head and neck region, followed by trunk, upper and lower extremities. Most frequent sites are periorbital (36 %), preauricular (28 %) and upper cervical (23 %) areas when it is located at head and neck

region. Majority of the cases are in pediatric age, 90 % of them are under 10 years of age with a female predominance [2, 4]. Our case was detected in the head and neck region which is the most common site of occurrence.

Some studies suggest that one of the causative factor is polyoma virus as well as concomitant with myotonic dystrophy, gardner syndrome, xeroderma pigmentosum and basal cell nevus syndrome [7]. Our patient had no special feature in his history, family history or physical examination to propose a genetic anomaly.

Clinically pilomatrixomas are slowly growing, asymptomatic, mobile, solid nodules or cystic mass in dermis or subcutaneous tissue with overlying normal skin. In very superficial lesions, skin changes as bluish or reddish can be seen. Preoperative diagnosis varies between 0 and 30 % [4, 6, 8].

Lack of familiarity to lesion by clinician and confusing with other tumours due to location of mass as in preauricular region result in difficulty in preoperative diagnosis. Firm mass like cartilage or bone is its characteristic feature. Other typical features are slow growing, freely mobile under skin and blue reddish colour change in overlying skin. This colour change occurs in 24 % of cases [4]. In our patient, the lesion enlarged during 1.5 year period after he noticed a swelling posterior to his chin. This slow progression caused delayed admission to hospital.

Occasionally, fast growing, ulcerated and painful cases as well as multiple located pilomatrixomas are detected [4, 9, 10]. Tumour size usually varies between 0.5 and 3 cm but in a few publications tumours larger than 3 cm was presented [6, 8, 11]. Fistulisation of overlying skin is a rare feature and the big size of tumour (5 × 4 cm in diameter) makes our case one of giant pilomatrixomas presented in literature.

Primarily, malignancy is considered in differential diagnoses. Other differential diagnoses are sebaceous cyst, ossifying hematoma, branchial remnant, preauricular sinus, adenopathy, chondroma, giant cell tumour, dermoid cyst, degenerated fibroxanthoma and foreign body reaction. Especially in preauricular location, it can be mixed up with tumours and inflammatory lesions of superficial lobe of parotid gland. In general, the tumour is easily recognised clinically if clinicians have prior exposure to the same lesion. But unfortunately, most clinicians are unaware of the lesion so correct prediagnosis is inconsistent (0–30 %) [12]. FNAB of our case was reported as pleomorphic adenoma so we planned our surgery as benign tumour of parotid gland.

Pilomatrixoma is diagnosed by FNAB as well as US and CT imaging. But in differential diagnosis, CT, MRI or US has no pathognomonic feature. For accurate diagnosis, histopathological examination is necessary. Pathological study is done to search for basophilic and shadow cells, and demonstrate calcium deposits in parenchymal tumoural cells and/or connective tissues [13].

Treatment of pilomatrixoma is complete surgical excision and in the preauricular region it is associated with the possibility of facial nerve damage, especially if the tumour

is large as in this case. Early exposure of the facial nerve and its complete display at the time of surgery allowed this large tumour to be safely and completely excised with no postoperative facial nerve injury. In malignant form which has a high potential of recurrence, total excision with a 1–2 cm safe surgical margin is suggested in literature [14]. Recurrence rate ranges between 0 and 3 % in publications [2, 4]. Pilomatrixoma has a good prognosis, but malign transformation was reported after repeated recurrent tumour excision [15]. So patient should be followed for recurrence. Our patient showed no recurrence till his control at 7th month.

Conclusion

Pilomatrixomas are benign lesions treated with total excision with low recurrence rate. Clinically it is difficult to diagnose a pilomatrixoma if clinicians have no prior exposure to the same lesion. Patients should be followed up for local recurrence. It is mostly seen in head and neck region so, clinicians should keep pilomatrixoma in mind in the differential diagnosis of head and neck masses.

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