Egyptian Journal of Ear, Nose, Throat and Allied Sciences (2015) **16**, 193–199

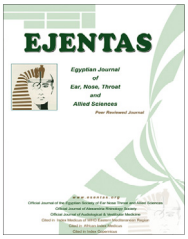
HOSTED BY

ELSEVIER

Egyptian Society of Ear, Nose, Throat and Allied Sciences

Egyptian Journal of Ear, Nose, Throat and Allied Sciences

www.ejentas.com



CASE REPORT

Ossifying pilomatrixoma with marrow formation of the left cheek region – Case report with review of literature

Ashwin Ashok Jaiswal [a,*], Amrish Kumar Garg [a], Ravindranath Membally [b], Tanuja Anand [c], Manoj Kumar Mohanty [d]

[a] *Department of ENT & Head Neck Surgery, J.L.N. Hospital & Research Centre, Bhilai (CG), India*
[b] *Department of Pathology, J.L.N. Hospital & Research Centre, Bhilai (CG), India*
[c] *Department of Anaesthesia, J.L.N. Hospital & Research Centre, Bhilai (CG), India*
[d] *Department of Paediatric Surgery, J.L.N. Hospital & Research Centre, Bhilai (CG), India*

Received 4 January 2015; accepted 5 February 2015
Available online 24 February 2015

KEYWORDS

Pilomatrixoma;
Ossification;
Marrow;
Neoplasm;
Otorhinolaryngologist;
Benign

Abstract Pilomatrixoma is a rare, benign, circumscribed, calcifying epithelial neoplasm that arises from the hair follicle on any part of the body. It may present to the otorhinolaryngologist as a palpable mass in the head and neck region. They are usually found in girls during the first two decades of life. These tumors may contain calcification, which, when present, is helpful in suggesting the diagnosis. We present a rare case of ossifying pilomatrixoma in a 15 year old female who presented with a painless hard slow growing swelling over the left cheek region. FNAC could not provide any clue whereas USG and CT scan showed the swelling to be in subcutaneous plane, superficial to the parotid gland. Complete excision of the mass along with the overlying adherent skin in toto with preservation of facial nerve was done. The diagnosis was confirmed after histopathological examination of the excised specimen which revealed pilomatrixoma with extensive ossification and marrow formation.

1. Introduction

Pilomatrixoma, also known as pilomatricoma, is a rare, benign tumor that originates in the cells of the hair follicle matrix.[1] The tumor was first described in 1880 by Malherbe and Chenantais, who believed that it arose from the sebaceous gland and called it as calcifying epithelioma of Malherbe.[2] In 1922, Dubreuilh and Cazenave described its unique histopathology consisting of basaloid cells and shadow/ghost cells. In 1942, Turhan and Krainer determined that the origin of this neoplasm is from hair

* Corresponding author at: Department of ENT & Head Neck Surgery, J.L.N. Hospital & Research Centre, Sector 9, Bhilai, Dist. Durg, Chhattisgarh 490009, India. Tel.: +91 9407983605; fax: +91 0788 2227814.
E-mail addresses: ash1978jaiswal@yahoo.com, ash1978jaiswal@gmail.com (A.A. Jaiswal), drakgarg19@yahoo.co.in (A.K. Garg), mrnath101@gmail.com (R. Membally), anand.tanuja@yahoo.in (T. Anand), manoj65us@yahoo.com (M.K. Mohanty).
Peer review under responsibility of Egyptian Society of Ear, Nose, Throat and Allied Sciences.

http://dx.doi.org/10.1016/j.ejenta.2015.02.002

cortex cells.[3] Later, the term pilomatrixoma was coined by Forbis and Helwig in 1961 thus avoiding the term epithelioma which carries the connotation of malignancy and it was suggested that the cells of origin are the outer root sheath cells of the hair follicle.[1]

Clinically, pilomatrixoma presents as a single, slowly growing, well-circumscribed, 5–30 mm in size, deep dermal and subcutaneous, firm to hard, superficial, mobile nodule. Usually they are solitary nodules but multiple and familial cases are also reported. The majority of these tumors arise during the first two decades of life; however, a second peak of occurrence is seen in older patients.[4] Overall, there is a female predominance. Even though the pilomatrixoma is more common in the head and neck, it tends to skip from the clinical diagnosis and diagnosis is usually made by histopathological examination of the excisional biopsy.[5,6] Hence, we report a unusual case of ossifying pilomatrixoma with marrow formation along with extensive review of literature.

2. Case report

A 15 year old female patient presented to OPD of J.L.N. Hospital & Research Centre, Bhilai, with a slowly progressive swelling over the left cheek region since 3 years. On examination, well circumscribed swelling of size approx 3 cm × 3 cm was seen over the left cheek region located 2 cm above the mandibular margin, 3 cm above and anterior to the angle of mandible and 3 cm below the zygomatic arch, the skin over the swelling was tethered with no visible pulsations. On palpation it was non tender, stony hard in consistency, the skin over the swelling was adherent to the swelling (Fig. 1). On intraoral examination, no bulge was seen on the buccal mucosa. There were no lymph nodes enlarged in the neck. The patient also had alopecia areata of left eyebrow. USG on left cheek region showed a large linear hyperechoic lesion of size 2 cm with a strong posterior acoustic shadowing obscuring the left parotid parenchyma. FNAC was tried but could not be done as needle could not be introduced into the stony hard swelling. Taking into consideration its consistency contrast enhanced CT scan on the head & neck was done which showed a large non enhancing lesion (of attenuation 1138–1140 HU) of size 11 mm × 23 mm × 24 mm in the subcutaneous tissue plane above the angle of mandible occupying the left cheek region with extensive calcification within soft tissue and maintained fat planes (Fig. 2(a–d)). A differential diagnosis was considered and the patient was planned for excisional biopsy under GA. The main concern was preservation of the facial nerve branches. Complete excision of the mass along with the overlying adherent skin in toto with preservation of the facial nerve was done. The wound was sutured meticulously with 6.0 prolene. Excised tumor of approx size 3 cm × 3 cm with stony hard consistency was sent for HPE (Fig. 3(a–d)). Grossly it was a hard tissue mass with the skin adhered on it which on cut section showed extensive bone formation. Histopathological examination showed irregular islands of epithelial cells which consist of basophilic cells as well as shadow cells. The stroma surrounding the epithelial island showed extensive ossification, at places in the ossified stroma hemopoietic cells were seen enclosed by bony trabeculae suggesting marrow formation (Fig. 4(a–d)). The postoperative course was uneventful and facial nerve functions were normal. Immunohistochemistry revealed pancytokeratin positive in ghost cells and Ki 67 negative for basaloid cells (Fig. 5(a and b)) favoring ossifying pilomatrixoma.

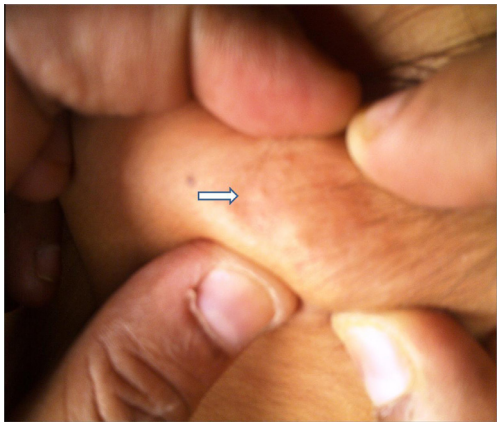


Figure 1 Clinical photograph showing, well circumscribed swelling of size approx 3 cm × 3 cm was seen over the left cheek region located 2 cm above the mandibular margin, 3 cm above and anterior to the angle of mandible and 3 cm below the zygomatic arch. On palpation the skin over the swelling was adherent to the swelling {*White arrow*}.

3. Discussion

Pilomatrixoma, or calcifying epithelioma of Malherbe, is a rare benign tumor of hair-follicle origin that usually arises in the head and neck region, most commonly in the first two decades of life.[7] However, the onset age varies from 4 months to 86 years.[2,8] Approximately 60% of pilomatrixoma occurs in patients younger than 20 years, of which in 40% of cases it occurs before the age of 10 years and in 60% of cases before the age of 20 years, whereas the distribution is somewhat in the bimodal age group.[2] It is more common in women (1.5–2.5:1) than men and even most authors have reported a female preponderance.[9] Our case was a 15 year old female patient.

In the 1970s, Moehlenbeck reviewed 140,000 skin tumors and found that pilomatrixoma accounted for only 0.12% of them.[10] The head and neck region is involved in about 40–77% of the patients with this tumor, followed by the cheek, scalp and periorbital areas; but it is not known to occur on the palms, soles, or genitalia.[2] In the head it occurs predominantly in the cheek followed by around the eyes, scalp and least of all in the preauricular region. Usually solitary, 2–3.5% are found in multiple.[2] In our case, a solitary swelling was situated over the left cheek region.

The etiology of pilomatrixoma is subject to controversy. Based on these anatomical features, several authors have suggested that the most likely mechanism for development of pilomatrixoma is inclusion of epidermic elements in abnormal locations during embryonic life and subsequent growth after birth.[11] Lever and Griesemer classified pilomatrixoma as a hamartoma, but Dubreuilh and Cazenave proposed two alternative hypotheses. The first is that, these tumors arise from branchial clefts. This theory seems unlikely because pilomatrixomas can develop at any age and it does not account for the lesions on the extremities.[11] The second hypothesis states that pilomatricoma is of ectodermal origin and that evidence exists to show that they originate from the hair germinal