CASE REPORT

Pilomatrixoma in Head and Neck Region

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ABSTRACT:

Pilomatrixoma is a rare, benign, circumscribed, calcifying epithelial neoplasm that arises from the skin on any part of the body. Here, we are presenting a case of pilomatrixoma in head and neck region, in a 40-year-old male patient. On clinical examination it was diagnosed as sebaceous cyst. Later, it was excised surgically. On microscopic examination, the lesion was identified as pilomatrixoma.

Since, the pilomatrixoma is a rare lesion and the dentist is tending to forget this entity, we discuss the clinical presentation, histopathological findings, and other characteristics of pilomatrixoma.

Key words: Pilomatrixoma: Hair follicle tumor: Malherbe's epithelioma: Von Kossa stain

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

Pilomatrixoma presents as a single, slowly growing, subcutaneous, firm to hard, superficial, mobile nodule.[1] Even though the pilomatrixoma is more common in head and neck, it tends to skip from the clinical diagnosis, because dentists are generally not familiar with it and there is a lack of discussion of pilomatrixoma

in the literature. Hence, we report a case of pilomatrixoma and discuss its clinical and histopathological findings.

Case Report:

A 40-year-old male patient complained of a slow growing swelling over the left cheek area since 4-years. There was past history of trauma in that particular area. On examination, the mass in the left cheek area, measuring about 2.5x1cm in dimension, soft to firm in consistency, lobulated, intermittently painful and freely mobile over the deeper structures was noticed. A provisional diagnosis of sebaceous cyst or fibroma or lipoma was made. Excision of the mass was performed under local anesthesia and sent for histopathological examination.

On microscopic examination, the haematoxylin and eosin stained section had pleomorphic appearance with variable cellularity and an admixture of dark-staining cellular and light-staining paucicellular areas. The more cellular areas consisted of dark-staining, basophilic small cells arranged as irregularly shaped islands at periphery and more eosinophilic ghost (shadow) cells arranged at the centre. The ghost cells contained abundant eosinophilic cytoplasm with/without small hyperchromatic nuclei. Other cellular components seen were histiocytes, few multinucleated giant cells, and areas of calcification. Von kossa technique was applied to demonstrate the calcium salts. Based on the results of the histopathologic analysis, we were able to make a diagnosis of pilomatrixomas. The patient's postoperative course was uneventful and there has been no recurrence of the lesion until now.

Discussion

In the 1970s, Moehlenbeck reviewed 140,000 skin tumors and found that pilomatrixoma accounted for only 0.12% of them. The head and neck region is involved in about 40% to 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]

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.[4]

Lever and Griesemer classified pilomatrixoma as a hamartoma, but Dubreuilh and Cazenave proposed two alternative hypothesis. 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.[4] The second hypothesis states that pilomatricoma is of ectodermal origin and that evidence exists to show that they originate from the hair germinal matrix.[2,4] Pilomatricoma formation represents a disturbance of the hair follicle cycle in which limited cytologic differentiation of pilar keratinocytes occurs but further development into mature hair fails to take place. There is a study suggesting that apoptosis is the main mechanism leading to the development of the dead shadow cells and is most probably responsible for the biological behaviour of pilomatricoma.[2]

The etiology of pilomatrixoma is subject to controversy. Rosa Guinot-Moya, Eduard Valmaseda-Castellon, Leonardo Berini-Aytes, Cosme Gay-Escoda done study on 211 lesions and they recorded antecedents of trauma, insect bites or surgery, intradermal reaction of vaccine, history of alopecia areata as the etiology.[5] Moehlenbeck FW observed and stated that there were no familial presen-tations in their retrospective study.[6] In our case, the etiology was found to be a trauma.

Approximately 60% of pilomatrixoma occur in patients younger than 20 years, and the distribution is somewhat in bimodal age group.[2] It is more common in women (1.5 to 2.5:1) than men and even most authors have reported a female preponderance.[7] It presents as a solitary cystic swelling that is typically painless and therefore often ignored. It eventually progresses to become a calcified lesion.

The present case occurred in a 40 year-old male patient of Indian origin in the left cheek area. It presented as a solitary, slow-growing, hard mass; there were no other associated findings or similar swellings elsewhere in the body.

Multiple pilomatrixoma are uncommon, as most authors report an incidence in the range of 2 to 10%. Multiple lesions have occurred in association with myotonic dystrophy, Gardner syndrome, Turner syndrome, Steinert disease, Rubinstein-Taybi syndrome and sarcoidosis.[1,2,3] In our case, the patient did not had any family history of pilomatrixoma and also did not demonstrate any of the features of these associated syndromes.

In view of the rarity of pilomatrixoma, high chances of missing the diagnosis of pilomatrixoma on clinical examination. The differential diagnosis includes dermal and subcutaneous masses (sebaceous cyst, epidermoid cyst, basal cell epithelioma, gaint cell tumor and neurofi-broma), calcified lesions (calcified epidermoid cyst, foreign body reactions or calcified hematomas).[5]

It is difficult to make the diagnosis on clinical examination alone, hence excisional biopsy has to be done. Histopathologic examination will reveal a sharply demarcated dermal nodule. At the periphery are the nucleated basaloid cells and at the center are non-nucleated ghost cells. The basaloid cells generally have small, uniform nuclei, scant cytoplasm, and indistinct cell borders surrounded by fibrillary material. Sheets of ghost cells, which evolve from basaloid cells, represent dead cells that have retained their shape. A transitional area is noted between the basaloid area and the ghost cells. Calcification is predominantly seen in the area of the ghost cells. Giant cells, which represent a foreign-body reaction to the ghost cells, are also seen in the area where keratin is abundant.[2]

Ossification in pilomatrixoma is not known. Tomas and Kothare were reported ossification in 3 cases out of 8 cases.[8] In the present case, the calcifications were appreciated with the use of Von Kossa stains.

Four distinct morphological stages of pilomatricomas have been proposed

1) Early: small and cystic lesions
2) Fully developed: large and cystic neoplasms
3) Early regressive: foci of basaloid cells, shadow cells and lymphocytic infiltrate with multinucleated giant cells
4) Late regressive: numerous shadow cells, absence of basaloid or inflammatory cells, calcification and ossification may be present[9]

In the present case the pilomatrixoma was small and cystic; hence it is an late regressive. Standard treatment recommends the surgical tumor resection with safety margins in order to minimize the risk of recurrence of the malignant variants. [7, 10] Relapses are rare but if it occur it may be due to incom-plete removal of the lesion, occurring in 2-6%.[7,5]

The malignancy of pilomatrixoma is rare. In the lit-erature some 60 cases of malignant transformation have been documented till date. The principal indicators of malignancy are cellular pleomorphism, frequent mitotic figures and atypias, central necrosis and infiltration of the skin, soft tissues and lymphatic and vascular ele-ments. The malignant version of pilomatrixoma is more frequent in males (with a 3:1 predominance over fe-males) and in elderly individuals (60% are over 40 years of age). Once the diagnosis has been confirmed, management consists of radical excision with adequate resection margins, due to the high associated relapse rate (49% of controlled cases). [5] In our case no malignant variants were diagnosed and there has been no recurrence in the follow up of 3years of the patient.

Black et al report the clinical behavior of pilomatrix carcinoma in adults to resemble that of basal cell carcinoma in its potential to metastasize.[11] Distant metastases are rare, with only 6 cases described to date in the literature. Patient survival in such cases has been 3-18 months from the time of diagnosis of metastatic disease.[5]

Conclusion:

Pilomatricoma is a painless, firm, slow-growing, benign skin neoplasm that derives from hair follicle matrix cells. It is typically found in the neck, cheek, periorbital region and scalp. It is more common in children and adolescents. The characteristic

histopathologic appearance consists of ghost cells and basaloid cells. Recurrence after complete surgical excision is rare. The dentist should be familiar with this entity and should expand their differential diagnosis of a superficial mass in the head and neck region.

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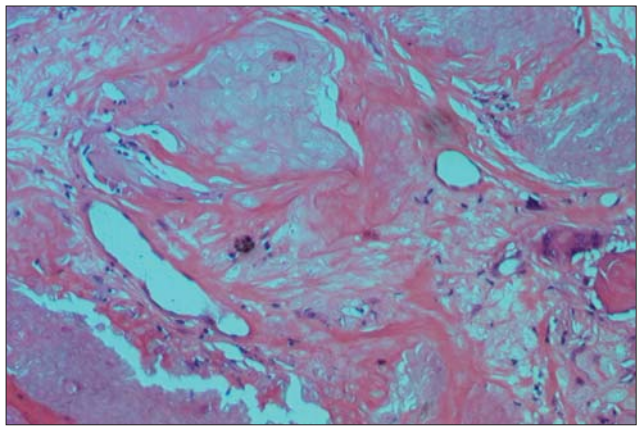

Fig 1: Photomicrograph of pilomatrixoma showing an admixture of dark staining cellular and light staining paucicellular areas. (hematoxylin and eosin, magnification-X 400).

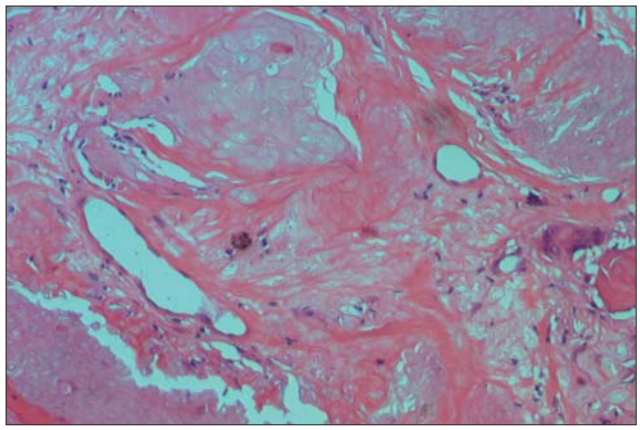

Fig.2: Photomicrograph of pilomatrixoma showing a ghost cell and giant cell. (hematoxylin and eosin, magnification- X400).

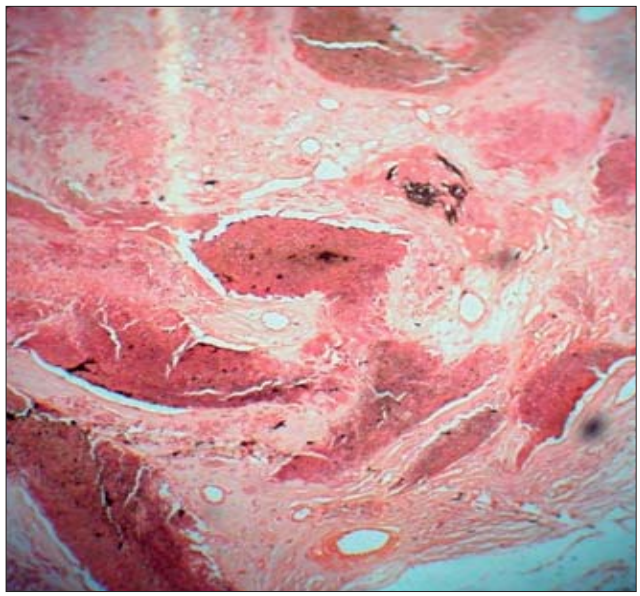

Fig 3: Photomicrograph of pilomatrixoma showing a calcification (Von Kossa stain, magnification- X100).