

CASE REPORT

Giant peduncular pilomatricoma (calcifying epithelioma of Malherbe): An interesting case

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ABSTRACT

Giant peduncular pilomatricoma (calcifying epithelioma of Malherbe) is a rare tumor first reported by Malherbe & Chenantais. In 1961, the term Pilomatrixoma was proposed by Forbis & Helwig which was later corrected to a more etymologically correct term Pilomatricoma. We report a 65 year-old Egyptian man, presenting with gradually enlarging mass on the base of the nose & glabellar area. Gross examination revealed hard non tender, protuberant mass with broad base, fixed partly to the skin, but freely mobile over the underlying structures. There was no lymphadenopathy. Punch biopsy was conclusive of pilomatricoma. The protuberant mass was excised & the wound closed with a partial thickness skin graft. The patient has no symptoms or signs suggestive of recurrence or metastases after 1 year of follow up.

INTRODUCTION

Malherbe and chentais¹ are credited with the first report of pilomatricoma. In 1880, they described calcifying epitheliomas, initially thought to be tumors of the sebaceous glands. Malherbe corrected this view in 1905.² Since then it has been called “calcifying epithelioma of Molherbe”. After several studies suggested the cell of origin to be the outer sheath cell of the hair follicle, Forbis and Helwig³ in 1961 proposed the term “Pilomatrixoma”. This new term was later corrected to “pilomatricoma”, which is more etymologically correct.⁴

This tumor usually appears as a benign, single, firm, stony hard, slow-growing subcutaneous or intradermal nodule, less than 3cm in size.⁵ Lesions usually are the color of the normal skin, but red purple lesions have been observed.⁶ It may appear at any age, with a bimodal peak, the first peak being 5-15 years and the second being 50-

65 years. It is more common in females (female-to-male ratio of 1.5:1 overall but 2.5:1 in patients younger than 20 years).^{7,8}

Approximately 50% of the lesions occur on the head and neck especially the cheek, pre-auricular area, eyelids, forehead, scalp, lateral and posterior neck, the remainder occurring in decreasing frequency in the upper extremity, Trunk, and lower extremity.⁹

CASE REPORT

A 65 year-old Egyptian man, otherwise in good health, presented with a 3 years history of a gradually enlarging mass on the base of the nose and glabellar area. Physical examination revealed a 7×5×3-cm hard non tender, protuberant mass on the base of the nose and glabellar area. It had a broad base with well-defined margins, fixed partly to the skin but freely mobile over the underlying structures. The lower half of the mass is chalky

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white with irregular surface while the upper half of the mass is fleshy red with somewhat smooth surface (Fig.1). The patient stated that the upper



Fig. 1 Showing protuberant mass on the base of the nose and glabella area with irregular surface chalky white lower half.

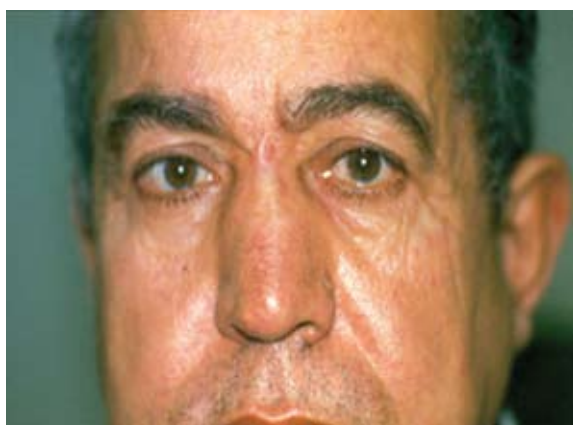


Fig. 2 Post surgical treatment clinical photo.

part of the lesion is the newly developed lesion. There was no lymphadenopathy. Further history, examination, hematology, biochemistry were normal. Plain X-rays showed massive calcifications in the mass with no definite bony involvement. CT revealed a large, well-defined protuberant intradermal mass with abundant calcifications. The margins of the mass were clean. Punch biopsy done from the lesion was conclusive of pilomatricoma. And the sections revealed that the tumor had both basophilic and eosinophilic elements. The basophilic areas were composed of small basaloid cells

that were in contiguity with the eosinophilic areas. The basoid cells were without significant atypia. These cells blended with eosinophilic shadow cells that had central unstained areas, corresponding to the lost nuclei (Fig. 3). There were areas of focal calcification (Fig. 4). But there was no definite focus to suggest an aggressive or malignant nature. The overlying epidermis was atrophic. The protuberant mass was excised with a 3-mm margins and the wound closed with a partial thickness skin graft obtained from the clavicular area. The patient has had no symptoms or signs suggestive of recurrence or metastases after 1 year of follow up (Fig. 2).

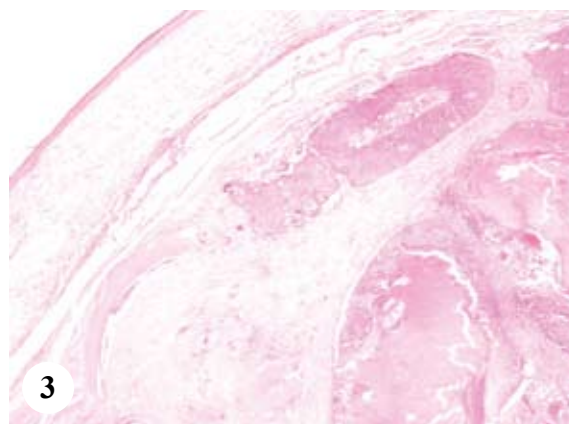


Fig. 3 H&E of the skin section showing both basophilic and eosinophilic elements of the tumor. Focal calcification can be identified.

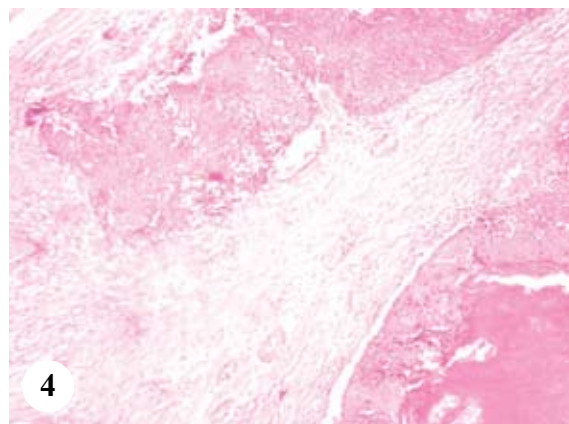


Fig. 4 High power showing the basaloid cell and the eosinophilic shadow cells.

DISCUSSION

Pilomatricoma is a relatively rare benign skin neoplasm that in general presents as a single dermal or subcutaneous nodule.¹⁰ Four clinical variants have been described: an eruptive type,¹¹ a perforating type,¹² a familial type associated with myotonic dystrophy¹³ and Gardner syndrome,¹⁴ and a recurrent invasive pilomatrix carcinoma.¹⁵ Any pilomatricoma over 3cm in size is considered large. And the term “giant pilomatricoma” has been used for lesions greater than 5cm in diameter, although the term is very subjective.¹⁶ The clinical diagnosis of pilomatricoma is a difficult one. In an extensive case series review by Julian and Bowers⁷ and another by Macleod and Scobie,¹⁷ they found a rate of incorrect diagnosis at the time of initial consultation of 79% and 75% respectively. Of foremost importance is an awareness of the occurrence of the lesion and its varied presentations. Palpation reveals a stony, hard consistency, which is a typical feature of pilomatricoma. The tumor slides freely over the underlying layer, and the overlying skin may have a reddish or bluish discoloration.¹⁸ The histological features of pilomatricoma are those of shadow cells, basophilic cells, and foreign body cells.¹⁶ Intracellular and stromal calcifications are noted in about 70% of cases.¹⁹ The histological features that distinguish a pilomatrix carcinoma from a pilomatricoma are the asymmetry of the tumor, the infiltrative border, the predominance of basaloid cells with necrotic foci and the invasion of vessels and nerves. The basaloid cells are large with hyperchromatic and vesicular nuclei and large nucleoli and have a high mitotic index with atypical mitoses.¹⁶ The characteristic radiological findings of pilomatricoma have been described as well-defined skin tumor with large amount of amorphous calcifications in the tumor

on conventional radiographs and CT scans.²⁰ Simple excision with narrow margins is usually satisfactory for pilomatricoma but wide local excision is recommended for pilomatrix carcinoma.^{16,21} The present case has showed no recurrence or metastases for 1 year since the resection. In conclusion, though we encountered a particularly large pilomatricoma, it did not have histopathological or radiological evidence of aggressive behavior.

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