International Journal of Otorhinolaryngology and Head and Neck Surgery
Jha S et al. Int J Otorhinolaryngol Head Neck Surg. 2022 Apr;8(4):409-411
http://www.ijorl.com
pISSN 2454-5929 | eISSN 2454-5937

DOI: https://dx.doi.org/10.18203/issn.2454-5929.ijohns20220813

Case Report

Giant pilomatrixoma on the back of neck: a unique case

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Received: 08 February 2022
Accepted: 03 March 2022

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ABSTRACT

Pilomatrixomas are benign adnexal tumours, commonly occurring in the head and neck region. We present a unique case of giant pilomatrixoma on the posterior triangle of the neck, its diagnosis and subsequent treatment and its potential to become a primary differential diagnosis of slow-growing, benign masses. We report a case of 10×10 cm slow-growing painless mass in the nape of the neck with overlying ulcerations and conflicting radio-pathological pre-operative reports and its subsequent diagnosis as pilomatrixoma. Histopathological examination revealed the presence of ghost cells and basaloid cells, confirming the diagnosis. We stress the importance of recognizing this pilomatrixoma variant, whose clinical picture is confusing and obscure**.** We also reiterate the need for close follow up of small lesions due to the small but grave risk of malignant transformation and recurrence. Pilomatrixoma has variable presenting features and poses a diagnostic dilemma. Surgical planning can be done with the help of proper radiological features, and complete excision of the mass is required to achieve a cure.

Keywords: Pilomatrixoma, Hair follicles, Benign tumour

INTRODUCTION

Pilomatrixomas, a benign tumour of hair follicles, predominantly occur in the head and neck region with low malignant transformation potential. It is uncommon, it affects women more and tends to happen in the first two decades of life.[1] It is synonymously known as calcifying epithelioma of Malherbe, the name kept after Malherbe and Chenantais, who first described this tumour in the 19th century. A single lesion or multiple slow-growing tumours are firm to hard, typically small, with most of them being less than 3 cm. These can present asymptomatically or are associated with pain and ulceration. Pilomatrixoma may be related to genetic disorders like Turner's syndrome, myotonic dystrophy, trisomy 9 and Sotos syndrome.[2]

Ultrasonographic features of pilomatrixoma are well defined hypoechoic lesions with internal echogenic foci or heavily calcified lesions and posterior acoustic shadows.[3] Computed tomography or magnetic resonance imaging categorises the extent of lesion and surrounding structure infiltration. Primary differential diagnoses are hemangiomas, plexiform neurofibroma, dermatofibroma and malignant basal cell carcinoma, cutaneous lymphoma, and leiomyosarcoma.[4]

The treatment of choice is surgical excision of the lesion, and the diagnosis is ultimately achieved and confirmed by histopathological examination of the excised tumour. Pilomatrixomas are diagnosed histopathologically by characteristic basaloid cells, multinucleated giant cells and ghost cells.

We present a peculiar case of gigantic pilomatrixoma of 10×10 cm in the back of the neck, which attained massive size due to negligence and faith in indigenous medicines. The unusual age, gender and presentation length, and ambiguous clinical picture suggest a malignant transformation in a long-standing mass. Our case is unique in establishing pilomatrixoma as a probable differential diagnosis in long-standing head and neck masses with conflicting clinical features and radio-pathological reports.

CASE REPORT

A 35-year-old male patient presented to the ear, nose and throat outpatient department (ENT OPD) of our tertiary care centre with a solitary mass in the posterior triangle of the neck for three years. Initially, it was growing slowly. However, it achieved a considerable increase in size in the past year. The patient only felt pain and did not have a fever or other constitutional symptoms and functional disability. The patient reported skin changes over the mass for six months associated with serous discharge. During the entire course of the disease, the patient followed the advice of a non-qualified local practitioner or some indigenous treatment providers.

On examination, the mass measured 10×10 cm extending from the nape of the neck superiorly to the clavicular region inferiorly and the posterior border of sternocleidomastoid to the midline of the rear neck side by side (Figure 1). It was round in shape, firm in consistency, non-compressible or fluctuant, non-pulsatile and non–transilluminate. It had multiple overlying ulcers with serous fluid oozing out of it. Fine needle aspiration cytology (FNAC) of the lesion suggested non-Hodgkin's lymphoma, round cell tumour or small cell carcinoma. There were no clinically enlarged lymph nodes in the neck or any part of the body. Hepatosplenomegaly was not revealed on systemic examination.

Figure 1: 10×10 cm mass present in the posterior triangle of the neck with overlying skin ulcerations.

A contrast enhanced computed tomography (CECT) neck scan was performed, which revealed a well-defined oval-shaped heterogeneously enhancing soft tissue density in the right posterior part of the neck in the subcutaneous plane, with mild surrounding fat stranding and abutting the underlying muscle (Figure 2). Based on these findings, the plan of surgical excision was made. Complete surgical excision of the mass was done under general anaesthesia (Figure 3).

Histopathological examination (HPE) report stated that the tumour was composed of a solid nest of basaloid cells and ghost cells with underlying stroma showing foreign body giant cells and calcification. The features were suggestive of pilomatrixoma (Figures 4 and 5).

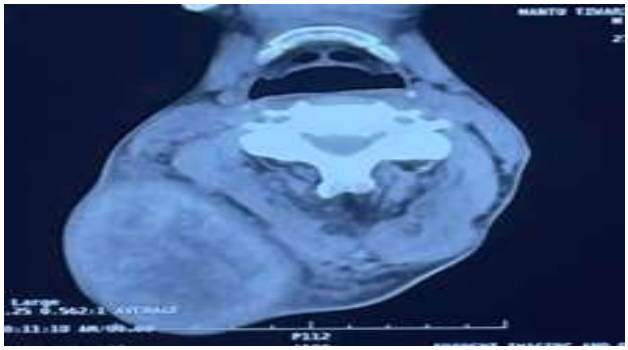

Figure 2: Axial CECT scan showed a well-demarcated soft tissue mass in the posterior neck.

Figure 3: Intra-operative picture of complete surgical excision.

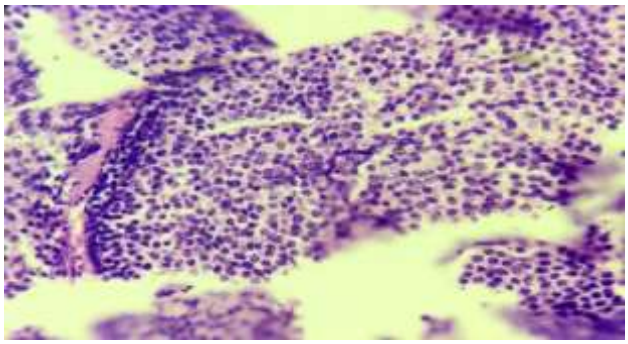

Figure 4: High power view showing basaloid cells with a round hyperchromatic nucleus and a scant cytoplasm.

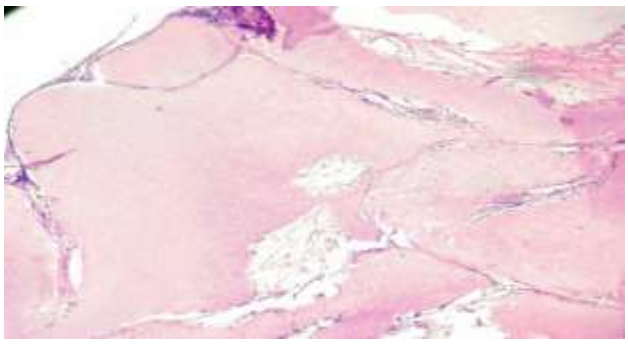

Figure 5: HPE image shows a large area consisting of ghost cells.

DISCUSSION

Pilomatrixomas are benign tumours usually present as small subcutaneous swellings, less than 3 cm.[5] These are slow-growing and seldom undergo malignant transformation. We present a challenging case in terms of diagnosis, both clinically and radiologically. The large size of the tumour, extensive inflammatory skin changes, and location were unlike most of the cases of pilomatrixoma reported in the literature and posed a diagnostic dilemma.

The clinical features of the tumour were ambiguous and suggestive of malignancy. Radiological aids like CECT were also non-conclusive and representative of other neoplasms like Hodgkin's lymphoma. The treatment done in almost all of these cases are surgical and is most definitive. Lesions that present over different regions have distinct reconstruction techniques. In our case, surgical excision led to a complete cure and relief from all symptoms. HPE remain the cornerstone of the final diagnosis and guide the treatment required after that. Through our case, we are highlighting the need to consider another differential diagnosis in obscure circumstances. We also reiterate the need for close follow up of small lesions due to the small but grave risk of malignant transformation and recurrence.

CONCLUSION

Pilomatrixomas are an unusual adnexal tumour derived from hair follicles that are vastly misdiagnosed. We present a unique and peculiar case of a slow-growing biopsy-proven pilomatrixoma in a 35-year-old man with ambiguous clinical features and conflicting radio-pathological pre-operative reports. This case report can help ease diagnostic dilemmas and push pilomatrixoma as an important differential diagnosis in head-neck tumours, significantly if slow-growing with varied presentation.

ACKNOWLEDGEMENTS

Authors would like to thank Dr. N. R. Biswas, Director and Dr. Manish Mandal for providing hospital material to perform this case and administrative support for the preparation of the manuscript.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Jha S, Singh J, Kumar A, Singh RK. Giant pilomatrixoma on the back of neck: a unique case. Int J Otorhinolaryngol Head Neck Surg 2022;8:409-11.