Original Research Paper

Volume-9 | Issue-6 | June-2019 | PRINT ISSN No. 2249 - 555X

Otorhinolaryngology

PILOMATRICOMA PRESENTING AS AN ANTERIOR NECK MASS – A CASE REPORT

Dr. Abhilasha Goswami — Post-graduate trainee, Dept. of Otorhinolaryngology, Gauhati Medical College and Hospital

Dr. Hironya Borah* — Associate Professor, Dept. of Otorhinolaryngology, Gauhati Medical College and Hospital *Corresponding Author

ABSTRACT Pilomatricoma is a benign skin neoplasm originating from hair follicle matrix cells, which is not commonly encountered in general practice. Histopathological analysis is usually needed to make the diagnosis. Here, we present a case of a 35 year old male who presented with an anterior neck mass, which was diagnosed to be a case of pilomatricoma only after excision. We are reporting this case to highlight the importance of histopathological analysis after excision of any swelling and the need to keep pilomatricoma in the differential diagnosis of all superficial skin tumours.

KEYWORDS : Pilomatricoma, Skin Neoplasm, Histopathology

INTRODUCTION :

Pilomatrixoma or Pilomatricoma, also known as calcifying epithelioma of Malherbe, is a benign calcified tumour originating from the outer sheath cell of the hair follicle root, due to uncontrolled proliferation of hair matrix cells. They typically present as a slowly enlarging, solitary mass, typically found in the head and neck region, but also occur in the upper extremities and are rarely reported in other sites. Pilomatricomas represent 0.12% of all skin tumours.[1]

CASE REPORT :

A 35 year old male presented to the Otorhinolaryngology out-patient department of Gauhati Medical College complaining of a swelling in the front of the neck for the past 5 months. The swelling was insidious in onset and gradually progressed from the size of a pea to its current size of an almond. The swelling was not associated with any pain or elevation in body temperature. The patient did not give any history of trauma prior to the onset. He is neither a smoker, nor does he consume alcohol. His medical history included gastritis. Family history was unremarkable. His vitals and general physical examination were normal. On local examination, there was a swelling on the anterior aspect of the neck, measuring 3.0x2.0 cm in size and with a reddish discolouration of the overlying skin. On palpation, the swelling was smooth, moderately firm to hard in consistency, non-tender, adherent to the overlying skin with no attachment to the underlying tissues and did not move with swallowing or on protrusion of the tongue. There was no associated scar, sinus, local rise of temperature or any other remarkable feature. A provisional diagnosis of epidermoid cyst was made. After doing the necessary blood investigations, surgical excision was planned. The swelling was excised under local anaesthesia. On gross examination, the excised mass appeared to be calcified. The mass was sent for histopathological examination, where the features were suggestive of pilomatricoma. Post-operative recovery period was uneventful.

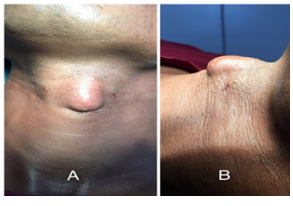

Fig 1: [A] anterior view and [B] lateral view of the patient's neck swelling

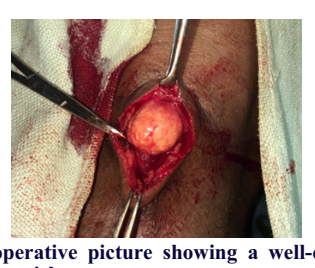

Fig 2 : Intra-operative picture showing a well-circumscribed, ovoid, solitary, greyish mass

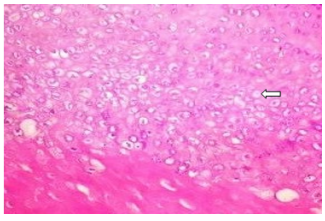

Fig 3 : Histopathological picture of pilomatricoma showing the basaloid-looking cells and the ghost cells(arrow)

DISCUSSION

Pilomatricoma is a slowly growing, benign, calcifying, cutaneous tumour , differentiating towards the hair matrix of the hair follicles, which undergo mummification and may calcify. It was first described by Malherbe and Chenantais in 1880 as a calcifying "epithelioma", arising from the sebaceous glands.[2] Dubreuilh and Cazenave in 1922 described the unique histopathlogical features of this neoplasm, including the islands of epithelial cells and ghost cells. [3] In 1961, Forbes and Helwig discovered that this tumour originated from the outer sheath of the hair follicle root and proposed the term pilomatrixoma.[4] The term was changed to pilomatricoma in 1977 as it was believed to be a more appropriate term etymologically.[5] These tumours are often found in the head and neck region, but occurrence in other parts of the body such as the upper extremity, trunk and the lower extremity have also been reported, in decreasing order of frequency. So far, no cases have been reported in the palms, soles or genital region.[6] Pilomatricomas are found in both sexes, but often show a female preponderance.[7] They show a bimodal pattern of incidence – in the first and sixth decades of life.[8] Pilomatricomas usually present as a solitary, asymptomatic nodule. Multiple lesions were found in 3.5% of reported cases, and were usually associated with other diseases or syndromes such as Gardner syndrome, Turner syndrome, myotonic dystrophy, sarcoidosis and Steinert syndrome.[8] Pilomatricomas may undergo malignant transformation, and though such cases have been reported, they are rare.[9,10] Some characteristic features of pilomatricoma are the "tent sign" and the "teeter-totter sign". Tent sign refers to the various facets and angles shown by the tumour whenever its overlying skin is stretched.[11] This is pathognomic of pilomatricoma. Also, whenever one edge of the tumour is pressed, the opposite edge protrudes out like a teeter-totter. Another characteristic feature of pilomatricoma is the bluish or reddish tinge of the overlying skin.[12] All these three features are the most helpful clinical clues to the diagnosis of pilomatricomas. In our case, all these three features were found to be present. Gross examination of biopsied pilomatricomas usually reveal a well-circumscribed ovoid or spherical mass, which may or may not be encapsulated. Histological analysis reveals the typical ghost cells in the center with periphery showing basophilic nucleated cells. Calcification is seen in 70 to 95% of cases.[13] Surgical excision is the treatment of choice, but care must be taken to obtain a clear margin, so as to reduce chances of recurrence due to incomplete resection.[12]

CONCLUSION

Pilomatricoma, though not uncommon, is rarely diagnosed preoperatively and it is imperative that it be kept in the differential diagnosis of all benign skin tumours. In conclusion, we would like to raise awareness amongst clinicians about this often forgotten entity and illustrate the importance of histopathological examination in making a definitive diagnosis of any superficial skin tumours.

FOOTNOTES

Financial Support : NIL
Conflict of Interest : NIL

REFERENCES

1. Greene R, McGuff H, Miller F. Pilomatrixoma of the Face: A Benign Skin Appendage Mimicking Squamous Cell Carcinoma. Otolaryngology–Head and Neck Surgery. 2004;130(4):483-485.
2. Malherbe A, Chenanatis J. Note sur l'epithelioma calcifiedes glandes sebacees. Prog Med 1880;8:826-37.
3. Dubreuilh W, Cazenave E. De I' epithelioma calcifie: etude histolgique. Ann Dermatol Syphilol 1922;3:257-68.
4. Forbis R Jr, Helwig EB. Pilomatrixoma (calcifying epithelioma).Arch Dermatol 1961;83:606-17.
5. Arnold HL. Pilomatricoma. Arch Dermatol 1977;113:1303
6. Boyd AS, Martin RW 3rd. Pathologic quiz case 1. Pilomatricoma (calcified epithelioma of Malherbe) with secondary ossification. Arch Otolaryngol Head Neck Surg 1992;118:212-5.
7. Yencha MW. Head and neck pilomatricoma in the pediatric age group: a retrospective study and literature review. Int J Pediatr Otorhinolaryngol 2001;57:123-8.
8. Moehlenbeck FW. Pilomatrixoma (calcifying epithelioma). A statistical study. Arch Dermatol 1973;108:532-4.
9. Zaman S, Majeed S, Rehman F (2009) Pilomatricoma-study on 27 cases and review of literature. D:/Biomedica25: 69–72.
10. Chuang CC, Lin HC (2004) Pilomatrixoma of the head and neck.J Chin Med Assoc 67: 633-636.
11. Graham JL, Merwin CF. The tent sign of pilomatricoma. Cutis 1978;22:577-80.
12. Roche N, Monstrey S, Matton G. Pilomatricoma in Children: Common but often Misdiagnosed. Acta Chirurgica Belgica. 2010;110(2):250–4.
13. Lan M-Y, Lan M-C, Ho C-Y, Li W-Y, Lin C-Z. Pilomatricoma of the Head and Neck. Archives of Otolaryngology–Head & Neck Surgery. 2003;129(12):1327.