## CASE REPORT

# **Pilomatrixoma in Periocular Tissues: A Case Report from Bangladesh**

Md. Nazmul Islam<sup>1</sup>, Dipankar Lodh<sup>2</sup>, Mohammad Mamun Siddiqui<sup>3</sup>, Mahedi Mannan<sup>4</sup>, AF Mohiuddin Khan<sup>5</sup>

# Abstract

Benign calcifying epithelioma of Malherbe or pilomatrixoma or pilomatricoma is less common lesion of the periocular tissues arising from the matrix cells at the base of the hair. In the periocular area, it usually arises from the lids and eyebrows. Pilomatrixoma has certain characteristic clinical and histopathological features, but since it is not commonly suspected preoperatively, certain distinctive clinical features of tumour should suggest clinical diagnosis followed by histopathological confirmation. [Journal of Science Foundation, 2015;13(1):21-24]

Keywords: Malherbe's epithelioma; pilomatricoma; pilomatrixoma

### Introduction

Benign calcifying epithelioma of Malherbe or pilomatrixoma or pilomatricoma is less common lesion of the periocular tissues, arising from the matrix cells at the base of the hair (Forbis and Helwig 1961). In the periocular area, it usually arises from the lids and eyebrows. Since it is not commonly suspected preoperatively, certain distinctive clinical features of tumour should suggest clinical diagnosis followed by histopathological confirmation (Malherbe and Chenantais 1880).

## **Case Report**

A 22-years young lady presented with a small nodule in the left sides of auricle (Helix) for last 3 years. The nodular lesion measuring about  $8\times6$  mm was present near the helix of the left auricle, subcutaneous in location, well-circumscribed, non-tender, mobile and firm with a gritty surface on palpation. Since the patient was keen on surgical removal, an excision biopsy was performed through a small incision given just over the lesion. Gross examination showed a well circumscribed single grayish white mass measuring  $7\times5\times5$  mm. Cut section showed grayish areas, firm and gritty. Microscopic examination showed numerous lobules with basophilic cells in the periphery and of ghost like squamous cells toward the center with a few lanucleated cells (Figure 1). Various stages of maturation of the basaloid cells into shadow cells could be seen (Figure 2). These islands are surrounded by foreign body giant cells with a

<sup>4</sup>Honorary Medical Officer, Department of Otolaryngology and HNS, Dhaka Medical College Hospital, Dhaka, Bangladesh

<sup>&</sup>lt;sup>1</sup>Associate Professor, Department of Otolaryngology and HNS, Dhaka Medical College Hospital, Dhaka, Bangladesh

<sup>&</sup>lt;sup>2</sup>Assistant Professor, Department of Otolaryngology and HNS, Dhaka Medical College, Dhaka, Bangladesh

<sup>&</sup>lt;sup>3</sup>Indoor Medical Officer, Department of Otolaryngology and HNS, Dhaka Medical College Hospital, Dhaka, Bangladesh

<sup>&</sup>lt;sup>5</sup>Professor and Head, Department of Otolaryngology and HNS, Dhaka Medical College Hospital, Dhaka, Bangladesh

**Correspondence:** Dr. Md. Nazmul Islam, Associate Professor, Department of Otolaryngology and HNS, Dhaka Medical College Hospital, Dhaka, Bangladesh; Email: <u>nimunna@gmail.com</u>

few lymphoplasmocytic infiltrates (Figure 3). There were numerous foci of calcification more in the necrotic areas and in the periphery of cellular islands (Figure 3). The histopathological features were consistent with a diagnosis of pilomatrixoma (benign calcifying epithelioma of Malherbe).

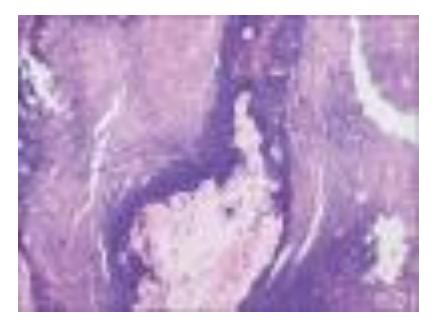


Figure 1: Photomicrograph showing central area of calcification surrounded by shadow cells. Basaloid cells can be seen in the periphery (H and E,  $\times 100$ )



Figure 2: Various stages of maturation of basaloid cells into shadow cells seen (H and E, ×400)

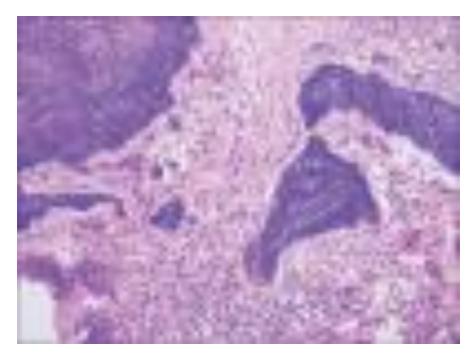


Figure 3: Photomicrograph showing numerous islands of basaloid cells and few foreign body giant cells (H and E,  $\times 100$ )

# Discussion

Pilomatrixoma is less common lesion that arises from the matrix cells. The hair **matrix** produces the actual hair shaft as well as the inner and outer root sheaths at the base of the hair. It was first described by Malherbe as benign calcifying epithelioma (Lever and Griesemer 1949). Subsequently numerous ultrastructural and electron microscopic studies (Mc Gavran 1965) provided strong evidence of its origin from the matrix cells and the term pilomatrixoma was then coined by Forbis and Helwig keeping the histogenesis into consideration (Mencia-Gutierrez et al., 2002).

Pilomatrixoma is usually a solitary lesion affecting young individuals. Forty percent of them develop in the first decade of life and another 20% in the second decade (Yap et al., 1999). They most commonly involve the head and neck region followed by upper extremities, trunk, and lower extremities. Periocular tissues are involved in 10–17% of cases and 5% of cases are multifocal (Boniuk and Zimmerman 1963; Orlando et al., 1983).

Pilomatrixoma generally presents with subcutaneous red to blue mass that is fairly well circumscribed, mobile and firm to gritty on palpation. Should arise clinical suspicion and they include onset in childhood or early adulthood, average size of 10 mm or less, consistency ranging from firm to cystic, moderate pattern of growth, pink to purple hue with subepithelial yellowish tinge, and intact overlying skin with telangiectatic vessels (Kang and Kang 2000).

Pilomatrixoma sometimes misdiagnosed with epidermoid cysts, dermoid cyst, sebaceous adenoma or carcinoma, juvenile xanthogranuloma, capillary hemangioma, chalazion, and rhabdomyosarcoma. Although they grow very slowly, they occasionally demonstrate rapid growth and may resemble keratoacanthoma (Martelli and Giardini 1994). They can rarely undergo malignant transformation into pilomatrix carcinoma.

Management includes a complete excision biopsy if there is any clinical suspicion of pilomatrixoma based on the clinical features described since this tumour is often confined to the soft tissues. Various aesthetic approaches can be used for excision.

### Conclusion

The diagnosis of pilomatrixoma will be made when the mass is adherent to the skin but loosely connected with the underlying tissue. It is difficult to distinguish between benign and malignant tumours by imaging methods alone, so the recommended treatment must be complete excision including adherent skin.

### References

Forbis R, Jr, Helwig EB. Pilomatrixoma (Calcifying epithelioma) Arch Dermatol. 1961;83:606–18

Malherbe A, Chenantais JE. Note sur l'epitheliome calcifie des glandes sebacees. Progres Med Par. 1880;8:826-8

Lever WF, Griesemer RD. Calcifying epithelioma of Malherbe: report of fifteen cases with comments on its differentiation from calicified epithelial cyst and on its histogenesis. Arch Derm Syphilol. 1949;59:506–18

Mc Gavran MH. Ultra structure of pilomatrixoma (calcifying epithelioma) Cancer. 1965;18:1445–56

Mencia-Gutierrez E, Gutierrez-Diaz E, Garcia-Suarez E, Ricoy JR. Eyelid pilomatricomas in young adults: A report of 8 cases. Cutis. 2002;69:23-6

Yap EY, Hohberger GG, Bartley GB. Pilomatrixoma of the eyelids and eyebrows in children and adolescents. Ophthal Plast Reconstr Surg. 1999;15:185–9

Boniuk M, Zimmerman LE. Pilomatrixoma (benign calcifying epithelioma) of the eyelid and eyebrow. Arch Ophthalmol. 1963;70:399–406

Orlando RG, Rogers GL, Bremer DL. Pilomatricoma in a pediatric hospital. Arch Ophthalmol.1983;101:1209–10

Kang HY, Kang WH. Guess what! Perforating pilomatricoma resembling keratoacanthoma. Eur J Dermatol. 2000;10:63-4

Martelli G, Giardini R. Pilomatrix carcinoma: A case report and review of the literature. Eur J Surg Oncol. 1994;20:703-4