

Tulachan B
Parajuli R

Ganesh Man Singh Memorial Academy of ENT and Head & Neck Studies, Tribhuvan University Teaching Hospital Institute of Medicine, Kathmandu, Nepal.

Correspondence to:

Dr B Tulachan
Ganesh Man Singh Memorial Academy of ENT and Head & Neck Studies, Institute of Medicine, Maharajgunj, Kathmandu, Nepal
E-mail: tulachanbishow@hotmail.com

PILOMATRIXOMA IN HEAD AND NECK

This is a case report of 16 years old male postoperatively diagnosed to be pilomatrixoma of right parotid. He underwent total conservative parotidectomy with Delto-Pectoral flap reconstruction.

Introduction

Pilomatrixoma was first reported in 1880 by Malherbe and Chenantais. They described a calcifying epithelioma, believing it to be a sebaceous gland tumor. Since 1905, this uncommon neoplasia has been called Malherbe's calcifying epithelioma. Numerous studies suggest that this tumor stems from the external sheath of hair follicles. In 1961, Forbis and Helwig proposed the term pilomatrixoma to avoid a connotation of malignancy. It is a benign skin neoplasia, originating from hair follicle matrix cells, representing 0.12% of skin tumours. Pilomatrixoma most commonly occurs in the head and neck. These are frequently misdiagnosed or missed from the differential diagnosis or both.

Keywords: Pilomatrixoma, Head and Neck.

CASE REPORT:

A 16 years old male from Solukhumbu presented to us with the swelling in the right parotid area for 1 year (fig. 1). It was insidious in onset, gradually progressive, painless without aggravating and relieving factors. On examination, there was a 6x5cm diffuse, firm, multi nodular erythematous swelling, fixed to skin, extending superiorly upto the inferior limit of the right ear lobule, inferiorly upto the inferior border of angle of mandible, laterally upto the posterior border of ascending ramus of mandible and medially approximately 3 cm away from the angle of mouth. USG of right parotid gland showed multiple nodular lesion clumped together within the right parotid parenchyma. FNAC showed atypical small to intermediate cells with round to oval hyperchromatic nuclei with small nucleoli in some cells arranged in clusters, loose aggregates and few singly scattered, occasional macrophages and giant cells, showing compatibility with malignant tumour with differential diagnoses of adenoid cystic carcinoma, solid variant and basaloid adenocarcinoma. He underwent total conservative parotidectomy with DP flap reconstruction (fig 2). Per operatively there was 8x7cm bosselated surface firm tumour in superficial lobe of parotid gland attached with skin. HPE showed focally ulcerated keratinized stratified squamous epithelium with numerous solid nests of small basaloid cells, ghost cells, giant cells and chronic inflammatory cells along with focal areas of calcification in subepithelial stroma suggestive of pilomatrixoma (calcifying epithelioma of Malherbe) (Fig 3).

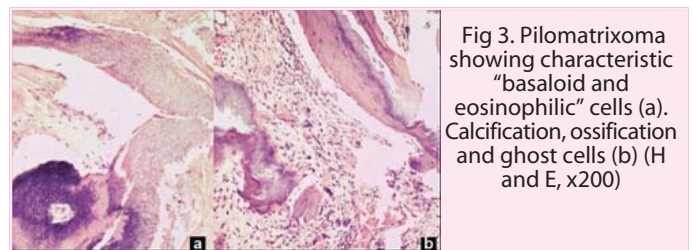


Fig 3. Pilomatrixoma showing characteristic "basaloid and eosinophilic" cells (a). Calcification, ossification and ghost cells (b) (H and E, x200)

DISCUSSION:

The pilomatrixoma is a relatively rare skin neoplasia. It may affect individuals at any age, with peaks incidence on the first and sixth decades of life, and it is more common in women (1.5 to 2.5:1). Among young people, 40% happen before 10 years of age and 60% before 20.¹ Growth is usually slow and may occur over a period of months to years.⁹ It most commonly occurs in the head and neck, with the upper extremity a distant second, followed by the trunk and lower extremity. In the head and neck, the cervical, temporal, eyelid, and preauricular regions are most frequently reported locations. None has been reported on the palms or soles, perhaps because of the lack of hair-bearing skin in these areas.³ Clinically it manifests as a subcutaneous or intradermal, hard and slow growing tumor.⁴ The criteria with which we may be able to distinguish a common benign mass as opposed to pilomatrixoma are the following: age of the patient, size of the mass, appearance of the overlying skin and features on palpation. It usually appears as an asymptomatic (it is associated with pain only in cases of inflammation and ulceration) deeply seated, firm, non-tender, subcutaneous mass and adherent to skin but not fixed to underlying tissue. Stretching the skin over the tumour may show the tent sign, with multiple facets and angles. A characteristic feature is the blue-red discoloration of the skin which allows us to exclude the possibility of the existence of epidermal inclusion and dermoid cysts.⁶ The pre-surgical diagnosis is almost always difficult, especially when the elderly are concerned. The finding of a keratinous material in the FNA can be misinterpreted as carcinoma.⁵ Histological characteristics include ghost cells in the center with basophilic

nucleated cells in the periphery. Calcification is present in 70-95%.⁴ The presence of nuclear pleomorphism, atypical mitosis, central necrosis, skin and adjacent tissue infiltration, besides ulceration, is suggestive of malignancy.⁷ The CT scan has little value to approach pilomatrixomas. It is mainly used to differentiate pre-auricular from parotid tumors and to assess large and aggressive tumors. The characteristic is a well outlined and calcified subcutaneous lesion.⁴ The treatment of choice and standard therapy for benign pilomatrixoma is complete surgical excision.² It is recommended to remove the tumor with safety margins in order to minimize the risk of recurrence of the malignant variants. Adjacent skin must be occasionally resected when adhered to the dermis.¹ The incidence of recurrences after surgery has been reported to be between 0% and 6%.⁶

REFERENCES:

1. José Arruda Mendes Neto, Rafael Mônaco Raposo, Danilo Kanashiro Segalla, Fernando Danelon Leonhardt. Pilomatrixoma in head and neck. *Braz J Otorhinolaryngol.* 2009;75(4):618
2. Angélique Danielson-Cohen, Samuel J. Lin, C. Anthony Hughes, Young H. An, John Maddalozzo. Head and Neck Pilomatrixoma in Children, *Arch Otolaryngol Head Neck Surg.* 2001;127:1481-1483
3. Gülden Avci, M. Akan and T. Aköz. Simultaneous Multiple Pilomatrixomas. *Pediatric Dermatology.* Vol. 23 No. 2 157-162, 2006
4. Lan MY, Lan MC, Ho CY, Li WY, Lin CZ. Pilomatrixoma of the head and neck: a retrospective review of 179 cases. *Arch Otolaryngol Head Neck Surg.* 2003;129(12):1327-30
5. Saussez S, Mahillon V, Blaivie C, Haller A, Chantrain G, Thill MP. Aggressive pilomatrixoma of the infra-auricular area: a case report. *Auris Nasus Larynx.* 2005;32(4):407-10.
6. and , et al. , 2005, , Pages 673-677
7. Phyu KK, Bradley PJ. Pilomatrixoma in the parotid region. *J Laryngol Otol.* 2001;115(12):1026-8.

