Pilomatrixoma is a benign uncommon neoplasm of hair follicles that was first described by Malherbe and Chenantois in 1880. It was in 1961 that Forbes and Helwig suggested the name pilomatrixoma.1,5 The majority of these tumours arise in the first two decades of life, a second peak can occur in older patients. The tumour is usually deep seated, solitary firm nodule with overlying normal epidermis. The tumour may be solid or partly...
solid and partly cystic and calcium deposits are also seen, that originates from the pleuropotential precursors of the hair matrix cells, hence also called ‘Calcifying epithelioma of Malherbe’.

Skin overlying may turn purplish in colour and in some cases may present with unusual haemorrhagic mass. Usually solitary, multiple tumours are also described. The majority of tumours are benign, rarely malignant changes are also mentioned in the literature.

**Case Report**

A seventy-six year old Caucasian female presented in the ENT department of Dalhousie St-Joseph Community Hospital, in Dalhousie, New Brunswick, Canada with swelling over the left preauricular region that has been present for the past three months. The swelling appeared as a small nodule the size of a pea and increased in size rapidly. There was associated pain and blood stained discharge from the swelling. Examination of the swelling showed firm, partly solid and partly cystic swelling, that was mobile and 2.2cm in diameter. The skin over the swelling was haemorrhagic with a purplish hue surrounding it (Figure 1). FNAC performed showed haemorrhagic and was inconclusive. A CT scan of the neck confirmed the swelling was partly cystic, partly solid and was superficial to the parotid gland (Figure 2). Total excision of the swelling was performed under local anaesthesia. The skin over the swelling adhered to swelling was excised and sent for histopathology, and was reported as benign Pilomatrixoma (Figure 3). The skin excised was free from

**Figure 1:** Haemorrhagic nodular growth in the left preauricular region with a purplish halo.

**Figure 2:** Axial CT scan showing the left preauricular nodular swelling, partly solid, partly cystic, and attached to the skin over it.

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**Key Point**

Pilomatrixoma is a rare, slow growing, benign skin tumour derived from the hair matrix cells that mainly occurs in the head and neck.
the Pilomatrixoma. The patient was seen after a month for follow up and there was good cosmetic result (Figure 4).

**Discussion**

Pilomatrixoma or calcifying epithelioma of Malherbe is a rare benign tumour of the hair matrix that usually arises in the head and neck region mostly occurring in children or in the first two decades of life.\(^3\)\(^5\) Such histopathological analysis matrix as basaloid hair cells and shadow or ghost cells which represent central unstained cells show lost nucleus.\(^5\)\(^8\) Intracellular and stromal calcification is reported in 70% of cases. It has been suggested that osteopontin (a protein marker associated with bone production) may be produced by macrophages and play a role in the deposition of calcium phosphate in the shadow nest cells.\(^1\)

Clinical presentation usually asymptomatic,\(^1\) may present with superficial, solitary firm mass associated with reddish–blue discoloration overlying the skin.\(^5\) Usually these are slow growing,\(^5\) but our case was relatively fast growing. The diagnosis should be suspected when the mass is nodular, subcutaneous or intradermal and adherent to skin but not fixed to the underlying tissue.\(^3\) The majority of cases are in the head and neck. In the head predominantly in the cheek followed by around the eyes, scalp and least of all in the preauricular region.\(^8\)

**Key Point**

Very few cases of Pilomatrixoma have been reported in young adults or the middle aged group, as they occur predominantly in the first two decades of life.

**Figure 3:** Photomicrograph of Pilomatrixoma: Low power view showing an intradermal cystic like tumour composed of benign Basaloid cells, Shadow cells and Ghost cells.

**Figure 4:** Healthy skin over left preauricular region.
confirmed. Though these tumours are commonly benign, rarely malignant changes are described due to mutation changes in beta-catenin genes (CTNNB1) which were detected in 75% of cases. The exact role of mutation remains to be elucidated.

The differential diagnosis includes sebaceous cyst, ossifying haematoma, keratoacanthoma, pyogenic granuloma, fat necrosis, giant cell tumour, chondroma, dermoid cyst, foreign body reaction, degenerating fibroxanthoma, metastatic bone formation, and osteoma cutis.

Malignant pilomatrixoma can occur rarely, differential diagnosis would be basal cell carcinoma and metastatic skin cancer. In some cases of Parotid nodule when adherent to the skin can be difficult to differentiate from Pilomatrixoma.

CT Imaging findings show solid or partly cystic, non-infiltrating subcutaneous nodule that shows calcification. It is difficult to differentiate benign and malignant tumour by CT scan alone. Ultrasoundography showed heterogenous hyperechoic rim with internal echogenic in the subcutaneous

Key Points
We are presenting a case of Pilomatrixoma in a seventy-six year old lady with swelling and an unusual clinical presentation, which has not been reported before in the literature.

Clinical Presentation of Pilomatrixoma

Common Locations of Pilomatrixoma

- Scalp
- Periorbital
- Cheek
- Preauricular
- Neck

Differential Diagnosis of Pilomatrixoma
- Sebaceous Cyst
- Ossifying Haematoma
- Keratoacanthoma
- Pyogenic Granuloma
- Fat Necrosis
- Giant Cell Tumour
- Chondroma
- Dermoid Cyst
- Foreign Body Reaction
- Degenerating Fibroxanthoma
- Metastatic Bone Formation
- Osteoma Cutis

Differential Diagnosis of Malignant Pilomatrixoma
- Basal Cell Carcinoma
- Metastatic Skin Cancer
A Rare Case of Pilomatrixoma

SUMMARY OF KEY POINTS

Pilomatrixoma is a rare, slow growing, benign skin tumour derived from the hair matrix cells that mainly occurs in the head and neck.

Very few cases of Pilomatrixoma have been reported in young adults or the middle aged group, as they occur predominantly in the first two decades of life.

We are presenting a case of Pilomatrixoma in a seventy-six year old lady with swelling and an unusual clinical presentation, which has not been reported before in the literature.

Complete excision of the swelling after CT scan of the neck has given good cosmetic result.

CLINICAL PEARLS

We have presented here a seventy-six year old lady with haemorrhagic swelling and surrounding purplish skin in the left preauricular region. The swelling appeared the size of a pea and was growing rapidly. A CT scan of the neck was performed to localise the extension and a complete excision of the swelling was performed under local anaesthesia. Histopathology showed proximal portion of the hair follicle, darkly stained ‘basophilic cell’ and ‘shadow cells’, there was no calcification seen in the histopathology. Review of the literature has not shown similar presentation in the past.

Conclusion

Pilomatrixoma is a rare benign appendageal tumour of the hair, commonly seen in children and the first two decades of life. It is usually an asymptomatic solitary firm nodule. The image of non-infiltrating containing calcification within the subcutaneous tissue of the head and neck should raise the possibility of Pilomatrixoma. Diagnosis is confirmed after FNAC and excision biopsy. We have presented a case in a seventy-six year old lady with a

layer. Fine needle aspiration produces clustered or isolated basaloid cells with variable size nuclei, but is not always conclusive.

Complete excision of the tumour is the treatment of choice. Recurrence is seen in incomplete excision. However occasionally the tumour if not removed can grow to a giant size. If the tumour recurs then the possibility of malignancy should be considered. Inverted malignant Pilomatrixoma is also reported in the literature.

Key Points

Complete excision of swelling after CT scan of the neck has given good cosmetic result.
rare clinical manifestation and complete excision was performed. The skin over the swelling and adhered to the swelling was removed. There was no recurrence of the tumour after excision to this day.

Dr. P.K Shenoy takes responsibility for the integrity of the content of the paper.
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