

PILOMATRICOMA: AN UNUSUAL BENIGN DERMATOLOGICAL NEOPLASMDigvijay Singh Tanwar¹ and Ravi Verma^{2*}¹M.S Gen. Surgery, CH Kandaghat, Solan (H.P).²M.D Paediatrics, CH Kandaghat, Solan (H.P).

*Corresponding Author: Dr. Ravi Verma

M.D Paediatrics, CH Kandaghat, Solan (H.P).

Article Received on 02/07/2021

Article Revised on 23/07/2021

Article Accepted on 12/08/2021

ABSTRACT

Pilomatricoma is an uncommon, benign tumour with differentiation towards both the hair matrix and cells arising in the cortex, most frequently appearing in the first or second decade of life. The diagnosis is often made only after histopathology. Pilomatricoma is not rare in occurrence but rarely diagnosed and it is imperative that it should be kept in the differential diagnoses of all superficial skin tumours by dermatologists and surgeons. The present case report is of a 55-year-old male who presented with a swelling in the neck, which was diagnosed as pilomatricoma only after excision.

KEYWORDS: Pilomatricoma, histopathology, excision.**INTRODUCTION**

Pilomatricoma, or pilomatrixoma, or calcifying epithelioma of Malherbe is a benign skin neoplasia originating from hair follicle matrix cells.^[1] It usually develops slowly and is known as a single or sometimes multiple benign solid lesions lying just under or in the skin.^[2,3] Pilomatricomas represent 0.12% of all skin tumors.^[4] Pilomatrixoma is classified within the family of skin adnexal tumors.^[5] These tumours most frequently appear in the first or second decade of life. Pilomatrixoma occurs as a solitary lesion; however, multiple lesions and familial patterns have been reported. Hereditary types have been associated with myotonic dystrophy.^[6] The lesions are typically painless, slow-growing, deep-seated hard subcutaneous nodules arising most commonly within the face and upper extremities.^[7] Calcium deposits are present in over half the lesions identified. Thus, the skin lesion is also described as a calcifying epithelioma. Some debate exists regarding accurate preoperative diagnosis of pilomatricomas. Histologically, pilomatricomas present as a well demarcated lesion, stemming from dermis and extending into the subcutaneous fat. They classically consist of islands of epithelial cells comprised of both basophilic cells with meagre cytoplasm and ghost cells that have a central unstained area indicative of a lost nucleus.

Pilomatricomas are generally asymptomatic and found in the head and neck area and, upper extremities, but rarely

identified on the chest, trunk, or lower extremities. They are reported to occur in people of all ages; however, the distribution favours both children and the elderly.

Here, we report a case of pilomatricoma diagnosed after surgical excision with the help of histopathology.

CASE REPORT

A 55-year-old male presented to the dermatology department with complaint of a swelling over the back of neck on left lateral side for 2 years. The swelling had a gradual onset. Initially the size of the swelling was very small (pea-sized) but slowly over last 2 years it had grown to the size of a small lemon. It was painless with an occasional dull ache in the swelling. There was no associated fever or malaise. There was no history of trauma prior to the onset of swelling. Family history and past history of the patient was unremarkable.

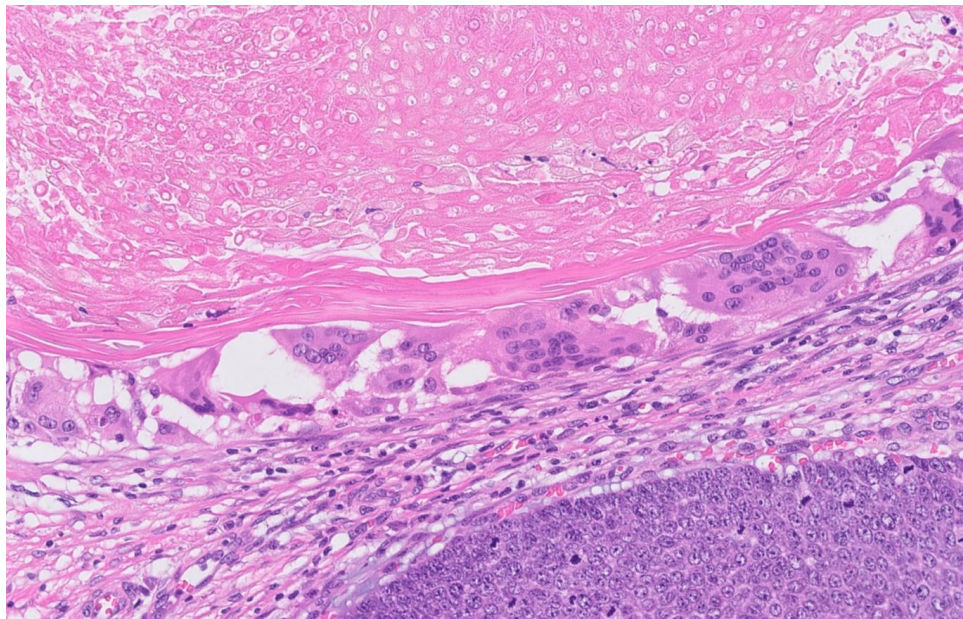
On examination there was a round swelling in the back of neck of size 3.5 × 3.0 cm. Its surface was variegated on palpation, moderately firm to hard in consistency and slightly tender to touch. It could be moved laterally with a little restriction in craniocaudal direction. There was fixity with the skin with no attachment to the underlying tissues. There was no rise of local temperature, scar, sinus, or any other remarkable feature. General physical and systemic examination was found normal.



A provisional diagnosis of a calcified sebaceous cyst was made. The case was referred to General Surgery where surgical excision was planned after blood investigations. The swelling was excised under local anaesthesia and sent for histopathological examination.

On gross examination, the excised mass appeared to be calcified during excision. A gritty sensation was felt while cutting the specimen. The cut surface of the nodule was variegated in appearance. Histopathologically, the

haematoxylin and eosin stained sections from the specimen showed a tumour composed of tissue fragment lined by benign, keratinizing, stratified, squamous epithelium revealing variable acanthosis and mild subepithelial, perivascular, mononuclear, inflammatory cell infiltrate and vascular congestion. Deep in the dermis were present islands of ghost cells surrounded by viable basaloid and squamous cells with foci of dystrophic calcification and foreign body histiocytic giant cell reaction.



These findings were consistent with the histopathology of Pilomatricoma.

DISCUSSION

Pilomatricoma is an asymptomatic slowly growing benign cutaneous tumour, differentiating towards the hair matrix of the hair follicle. It is covered by normal or hyperaemic skin, and usually varies in size from 0.5 to 3 cm. It is found particularly on the head and neck region (over 50% of cases) with a female predominance.^[8] Other locations include the upper extremity, trunk and lower extremity in decreasing order of frequency. No cases have been reported on the palms, soles or genital

region.^[9] Though pilomatricoma can develop at any age, it demonstrates bimodal peak presentation during the first and sixth decades of life, however, 40% of cases occur in patients younger than 10 years of age and 60% of cases occur within the first two decades of life.^[10]

Pilomatricomas usually are asymptomatic (pain appears only with associated inflammation and ulceration); deeply seated, firm, nontender subcutaneous masses adherent to the skin but not fixed to the underlying tissue. Stretching of the skin over the tumour shows the "tent sign" with multiple facets and angles, a pathognomonic sign for pilomatricoma.^[7] In addition,

pressing on one edge of the lesion causes the opposite edge to protrude from the skin like a "teeter- totter". Both these "tent sign" and "teeter- totter sign" are the most helpful clinical clues to the diagnosis of pilomatricoma. Another characteristic feature of Pilomatricoma is the blue red discoloration of the overlying skin which definitely excludes the possibility of epidermal inclusion or dermoid cyst.

Despite the well described features, pilomatricomas, till date, are frequently misdiagnosed. Literature survey shows that the accuracy rate of the preoperative diagnosis of pilomatricoma ranges from 0% to 30%.^[11] This may be attributable to the lack of familiarity with this tumour. Major factors contributing to misdiagnosis include: cystic lesions with varying consistency, punctum like appearance (due to skin tethering), atypical location and absence of clinically recognizable calcification. Another clinical dilemma encountered is the differentiation of this tumour from other benign masses, encountered in the clinical practice more frequently. These lesions include: epidermal inclusion cyst, dermoid cyst, brachial cleft remnants, preauricular sinuses, foreign body reaction, lipoma, degenerating fibroxanthoma, osteoma cutis, ossifying hematoma etc.^[8] Occasionally, there may be a history of previous trauma although this association is unusual. Finally, pilomatricoma can be associated with other diseases such as myotonic dystrophy, Gardner syndrome, Steinert's disease, Turner syndrome and sarcoidosis.^[12]

Radiologic imaging is of little diagnostic value for pilomatricoma. Fine needle aspiration cytology (FNAC) may be helpful. However, the results of FNAC can be misleading if there are no ghost cells present in the aspirate attributing to the misdiagnosis of many cases.^[13] Histopathologically, pilomatricoma consists of lobules and nests of epithelial cells composed of two major cell types: basophilic cells and eosinophilic shadow cells. Early lesions show a predominance of basophilic cells grouped in islands at the tumour periphery. With tumour maturation, the basophilic cells acquire more cytoplasm and gradually lose their nuclei to become eosinophilic shadow cells. These latter cells constitute the central portion of the tumour and frequently calcify. Gradually these calcified foci increase imparting the bony hard consistency to the lesion.

Since spontaneous regression is never observed and malignant transformation is rare, the standard treatment of pilomatricoma is complete surgical excision. Recurrence after surgery is rare, with an incidence of 0% to 3%.^[14]

CONCLUSION

Pilomatricoma is a benign neoplasm of the skin, which is not rare in occurrence but rarely diagnosed preoperatively and it is imperative that it be kept in the differential diagnoses of all benign skin tumours by dermatologists and surgeons.

REFERENCES

1. Fernandes R, Holmes J, Mullenix C. Giant pilomatricoma (epithelioma of Melherbe): Report of a case and review of literature. *J Oral Maxillofac Surg*, 2003; 61: 634-6.
2. Moehlenbeck FW. Pilomatricoma (calcifying epithelioma). A statistical study. *Arch Dermatol* 1973; 108:132-4.
3. Urvoy M, Legall F, Toulemont PJ, Chevrant-Breton J. [Multiple pilomatricoma. Apropos of a case]. *J Fr Ophthalmol*, 1996; 19: 464-6.
4. Greene RM, McGuff HS, Miller FR. Pilomatricoma of the face: A benign skin appendage mimicking squamous cell carcinoma. *Otolaryngol Head Neck Surg*, 2004; 130: 483-5.
5. Forbis R Jr, Helwig EB. Pilomatricoma (calcifying epithelioma). *Arch Dermatol*, 1961 Apr; 83(4): 606-18.
6. Berberian BJ, Colonna TM, Battaglia M, Sulica VI. Multiple pilomatricomas in association with myotonic dystrophy and a family history of melanoma. *J Am Acad Dermatol*, 1997 Aug; 37(2 Pt 1): 268-9.
7. Graham JL, Merwin CF. The tent sign of pilomatricoma. *Cutis*, 1978 Nov; 22(5): 577-80.
8. Yench MW. Head and neck pilomatricoma in the pediatric age group: a retrospective study and literature review. *Int J Pediatr Otorhinolaryngol*, 2001; 57: 123-8.
9. 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.
10. Moehlenbeck FW. Pilomatricoma (calcifying epithelioma). A statistical study. *Arch Dermatol*, 1973; 108: 532-4.
11. Julian CG, Bowers PW. A clinical review of 209 pilomatricomas. *J Am Acad Dermatol*, 1998; 39: 191-5.
12. Barberio E, Nino M, Dente V, Delfino M. Guess what! Multiple pilomatricomas and Steiner disease. *Eur J Dermatol*, 2002; 12: 293-4.
13. Agarwal RP, Handler SD, Matthews MR, Carpentieri D. Pilomatricoma of the head and neck in children. *Otolaryngol Head Neck Surg*, 2001; 125: 510-515.
14. Goufman DB, Murrell GL, Watkins DV. Pathology forum. Quiz case 2. Pilomatricoma (calcifying epithelioma of Malherbe). *Arch Otolaryngol Head Neck Surg*, 2001; 127: 218-20.