

Case Report

DOI: <http://dx.doi.org/10.18203/2349-2902.ijssurgery20182792>

Benign proliferative pilomatrixoma over sternum: a rare case

Muvva Sri Harsha^{1*}, Rajesh Goud E.², Srikanth Jakkula¹

¹Department of General Surgery, ²Department of Surgical Oncology, SVS Medical College, KNR University of Health Sciences, India

Received: 04 May 2018

Accepted: 29 May 2018

***Correspondence:**

Dr. Muvva Sri Harsha,

E-mail: dr.muvvaharsha@gmail.com

Copyright: © the author(s), publisher and licensee Medip Academy. This is an open-access article distributed under the terms of the Creative Commons Attribution Non-Commercial License, which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

ABSTRACT

Pilomatrixoma is a benign, uncommon, subcutaneous tumor arising from the outer root sheath cell or hair matrix cells. Pilomatrixoma, benign calcifying epithelioma of Malherbe, trichomatricoma are synonyms for pilomatrixoma. They occur mostly on head, neck and upper limb extremities. Histologically they classically consist of peripheral basilar cells and central enucleated ghost or shadow cells. Malignant changes are rare in pilomatrixomas. However, the proliferative variety is aggressive and considered to be precursor of malignancy. Here we present a case of benign proliferative pilomatrixoma over sternum for which the patient underwent wide local excision, diagnosis was later confirmed by histopathological examination. We have not come across any cases of pilomatrixoma occurring over sternum in literature.

Keywords: Benign proliferative, Pilomatrixoma, Sternum, Trichomatricoma

INTRODUCTION

Malherbe and Chenantais were first to describe pilomatrixoma in 1880.¹ Pilomatrixoma is defined as a benign tumor which is a hamartoma of hair matrix and consists of cells which are similar to hair matrix, cortex and inner sheath of root. They are also called as Benign calcifying epithelioma of Malherbe or Trichomatricoma or pilomatrixoma. Proliferating pilomatrixoma is a rare variant of pilomatrixoma, first described by Kaddu S. et al in 1997.² They are more common in younger age group i.e. before 20 years of age.³ They occur more commonly in head and neck (frontal, temporal, periorbital and preauricular areas) followed by upper limb extremities. Incidence among female is more than male counterparts. The cells usually undergo mummification. Histologically pilomatrixoma shows sharply demarcated dermal nodules surrounded by compressed fibrous tissue and is located in reticular dermis extending upto subcutaneous fat. Each nodule consists of multiple islands and each island consists of nucleated basilar cells towards periphery and

enucleated ghost cells in centre with in between transition zone. Calcification can be seen in ghost cells.

Here we present a case of Benign proliferative pilomatrixoma over sternum in a 45-year-old male patient. Till date, not even a single case of pilomatrixoma over sternum has been described in literature.

CASE REPORT

A 45-year-old man presented with complaints of a painless swelling over sternum since one year which rapidly increased in size since last 2 months. No history of trauma, discharge associated with the swelling. On examination, a 4x5 cm solitary swelling is seen over mid sternum area which is fixed to skin, has restricted mobility and not fixed to the sternum (Figure 1). It is firm in consistency. Skin over the swelling is erythematous and is pinkish red in colour. Surrounding skin is normal and there are no palpable lymph nodes. Fine needle aspiration cytology was done (FNAC) which suggested a

diagnosis of Pilomatrixoma with a suspicion of malignancy.



Figure 1: Pre-operative picture showing erythematous swelling over mid-sternum.

Wide excision with two (2) centimetres margin was done and the specimen was sent for Histopathological examination (Figure 2, 3).



Figure 2: Specimen obtained after wide excision with 2 cms margin.



Figure 3: Gross cut-section of the specimen.

arranged in a circular configuration and each island consisting of basloid cells in periphery along with enucleated 'Ghost' or 'Shadow' Cells in the centre which confirmed the diagnosis of Benign proliferative pilomatrixoma (Figure 4). Patient was kept on regular follow-up.

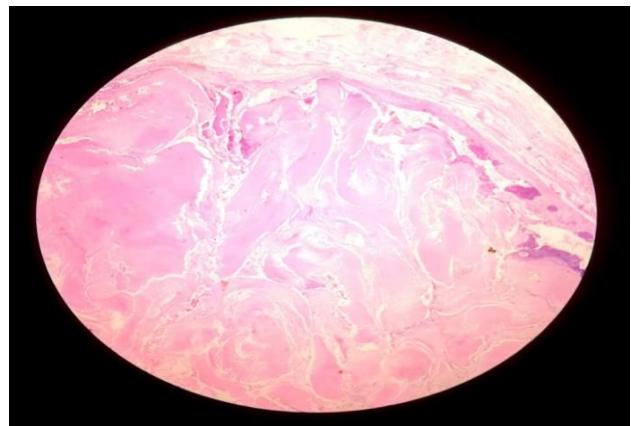


Figure 4: Microscopic picture showing peripheral basloid cells and central enucleated Ghost cells.

DISCUSSION

Pilomatrixoma is a superficial benign tumour with origin from outer root sheath cell or the hair matrix cells. Malherbe and chenantais were first to describe pilomatrixoma, they described it as a benign subcutaneous tumour arising from the sebaceous glands in 1880. Subsequently Forbis and Helwig proposed the term "pilomatrixoma" denoting its origin from the hair matrix cells.⁴ Synonyms for pilomatrixoma are pilomatrixoma, trichomatricoma and benign calcifying tumour of Malherbe. The common sites involved are head (frontal, temporal, periorbital and preauricular areas) and neck followed upper extremities, trunk and lower extremities.^{5,6}

It may occur at any age, however they are mostly seen either in children under 10 years of age or within the second decade of life.⁷ There is slight female preponderance. CTNNB1 gene have been demonstrated in pilomatrixoma indicating a betacatenin misregulation. BCL2 which is a proto-oncogene is also seen in pilomatrixomas. Familial association with conditions like myotonic dystrophy, turner syndrome, rubinstein-taybi syndrome, gardner syndrome and steiner disease is seen.⁸

Clinically, pilomatrixoma is usually a solitary, 0.5 to 3 cm sized, slow growing painless dermal swelling which is firm to hard in consistency. In most of the cases, skin over the swelling is normal and it is not fixed to underlying structures. Rarely it can also present as a either a granulomatous or ulcerated lesions. Diagnosis of pilomatrixoma can be considered if a subcutaneous nodule feels hard. Differential diagnosis include epidermal cyst, dermoid cyst, sebaceous cyst, sebaceous adenoma or

carcinoma, capillary hemangioma, rhabdomyosarcoma, basal cell tumor, squamous cell tumor etc.^{9,10}

Histologically, pilomatrixoma is composed of well circumscribed round islands giving a lobulated shape. The cells are broadly organised into two types, the outer and inner cells. The outer cells are small with rounded nuclei which makes this region deeply basophilic. The inner layer has enucleated cells with abundant cytoplasm i.e. "Shadow cells" or "Ghost cells". Thus, on HPE, pilomatrixoma usually shows lobules of basilar cells intermixed with pale pink areas containing ghost cells.

According to Kaddu et al, pilomatrixoma exhibits a distinct histomorphologic spectrum of four stages consisting of early, fully developed, early regressive and late regressive stages. Of the different histological varieties of pilomatrixoma, proliferative pilomatrixoma is a distinctive entity as mimics malignancy due to numerous mitosis and atypia.¹¹ The predominant clusters of basilar cells with prominent nucleoli, bare nuclei, numerous mitosis, atypia and necrotic debris can be mistaken as malignancy. But the presence of ghost cells, calcification and giant cells favours the diagnosis of pilomatrixoma.¹²

The lesion is usually benign but the tendency local recurrence is high, therefore it is important to excise the tumour completely. Malignancy is recorded in few cases but it is very rare and is usually in large long standing pilomatrixomas. Pilomatrical carcinoma is the malignant counterpart of pilomatrixoma. It is very rare and are seen in large, long standing and ulcerative type of pilomatrixomas. They are locally aggressive with extremely high tendency of recurrence. Distant metastasis is rare.

There is no role of medical management in pilomatrixoma and the tumour doesn't regress spontaneously. Treatment includes local excision and a wide excision with one to two centimetres margin can be done if malignancy is suspected. Local recurrence in non malignant conditions is rare provided the tumour is excised completely. The patient should be kept on regular follow-up in case of malignancy suspicion.

CONCLUSION

Benign proliferative pilomatrixoma over the sternum is a rare clinical entity and can easily be misdiagnosed as any other subcutaneous lesion. Hence, it is important to go for appropriate investigations in cases of ambiguous diagnosis so that a proper decision regarding the management can be made. To our knowledge, we haven't

come across any case of pilomatrixoma occurring over the sternum in the literature.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: The study was approved by the Institutional Ethics Committee

REFERENCES

1. Malherbe A, Chenanatis J. Note sur l'epithelioma calcifie des glandes sebacees. Prog Med. 1880;8:826-37.
2. Kaddu S, Soyer HP, Wolf IH, Kerl H. Proliferating pilomatrixoma: A histopathologic simulator of matrical carcinoma. J Cutan Pathol. 1997;24:228-34.
3. Moehlenbeck FW. Pilomatrixoma (calcifying epithelioma). A statistical study. Arch Dermatol. 1973;108:532-4.
4. Forbis R Jr, Helwig EB. Pilomatrixoma (calcifying epithelioma). Arch Dermatol. 1961;83:606-17.
5. Yap EY, Hohberger GG, Bartley GB. Pilomatrixoma of the eyelids and eyebrows in children and adolescents. Ophthal Plast Reconstr Surg. 1999;15:185-9.
6. Cypel TK, Vijayasekaran V, Somers GR, Zuker RM. Pilomatrixoma: Experience of the hospital for sick children. Can J Plast Surg. 2007;15:159-61.
7. Bansal C, Handa U, Mohan H. Fine needle aspiration cytology of pilomatrixoma. J Cytol 2011;28:1-6.
8. Barberio E, Nino M, Dente V, Delfino M. Guess what! Multiple pilomatrixomas and Steiner disease. Eur J Dermatol 2002;12:293-4.
9. Bansal C, Handa U, Mohan H. Fine needle aspiration cytology of pilomatrixoma. J Cytol 2011;28:1-6.
10. Ali MJ, Honavar SG, Naik MN, Vemuganti GK. Malherbe's calcifying epithelioma (Pilomatrixoma): An uncommon periocular tumor. Int J Trichol. 2011;3:31-3.
11. Satoh M, Ookouchi M, Yamamoto T. Proliferating pilomatrixoma with no recurrence during a 3-year follow-up. J Dermatologic Case Reports. 2012;6(4):127.
12. Sassemannshausen J, Chaffins M. Pilomatrix carcinoma: A report of a case arising from a previously excised pilomatrixoma and a review of the literature. J Am Acad Dermatol. 2001;44(2 Suppl): 358-61.

Cite this article as: Harsha MS, Goud RE, Jakkula S. Benign proliferative pilomatrixoma over sternum: a rare case. Int Surg J 2018;5:2657-9.