Case Report

New Indian Journal of Surgery
Volume 12 Number 2 / April – June 2021
DOI: https://dx.doi.org/10.21088/nijs.0976.4747.12221.8

Perforating Pilomatrixoma of The Male Breast, A Rare Lesion Simulating Breast Cancer: A Case Report

Bhavuray Teli[1] , Dr Ashok Mallapur[2], Dr Yashaswini M[3], Dr Vishwanath G[4], Dr Sandeep Chinnapur[5], Dr Geetanjali M Katageri[6]

Author's Affiliation: [1,5]Associate Professor, [2]Professor and Dean, [3]Post Graduate Student, [4]Professor, Department of General Surgery, [6]Professor, Department of Obstetrics and Gynecology, S Nijalingappa Medical College & Hanagal Shree Kumareshwar Hospital & Research Centre, Bagalkot 587103, Karnataka, India.

How to cite this article:

Bhavuray Teli , Dr Ashok Mallapur, Dr Yashaswini M et al./Perforating Pilomatrixoma of The Male Breast, A Rare Lesion Simulating Breast Cancer: A Case Report/New Indian J Surg. 2021;12(2):51–54.

Abstract

Pilomatrixoma is a rare benign skin appendage tumor of hair matrix origin. It is commonly seen in the first two decades of life, most often presenting on the face, neck or upper extremities. Pilomatrixoma of the breast is uncommon, and can mimic breast malignancy. Male breast Pilomatrixomas an extremely rare entity with only 9 cases reported in English literature. We report a rare presentation of perforating Pilomatrixoma of the breast in a male patient aged 35 years.

Keywords: Pilomatrixoma; Male breast; Calcifying epithelioma; Pilomatricoma.

Introduction

Presentation of a male patient with a breast lump opens the possibility of a variety of differential diagnoses, ranging from relatively innocuous conditions like gynecomastia to malignancy.[1] In particular, the finding of a hard lump in the breast is very likely to lead the clinician to suspect a malignant tumour of the breast. However, there could be both non-neoplastic (fat necrosis) and benign neoplastic conditions, also presenting with a hard breast lump, presenting a diagnostic dilemma [2]. Ulceration associated with the lump, in particular, is all the more suggestive of a malignant neoplasm. Clinical features and thorough evaluation usually help in arriving at the correct diagnosis. We present this case of pilomatrixoma of the breast in a 35 year old male patient who presented with a hard lump in the left breast with ulceration. This case is presented due to the rarity of site and manner of presentation.

Corresponding Author: Dr Yashaswini M, Post Graduate Student, Department of General Surgery, S Nijalingappa Medical College & Hanagal Shree Kumareshwar Hospital & Research Centre, Bagalkot 587103, Karnataka, India.

Email: yashaswinimadhusudan@gmail.com

Case report

A 35-year old male presented with a painless lump in the left breast since 4years and increase in size over the last 6 months. There was a history of ulceration of skin over the lump since 2 months. There was no family history of breast cancer. On examination there was 6x5 cm lump in the outer quadrants of the left breast. There was bluish-red discolouration of the skin with a 2 cm x 1.5 cm ulcer over the summit [FIG 1].

There was whitish, hard material on the floor of the ulcer with indurated base. The lump was hard and mobile over the underlying muscles. There was no axillary lymph node enlargement. Ultrasonogram (USG) of the breast revealed epidermal cyst with calcification. Fine Needle Aspiration Cytology (FNAC) showed features suggestive of benign skin adnexal tumor.

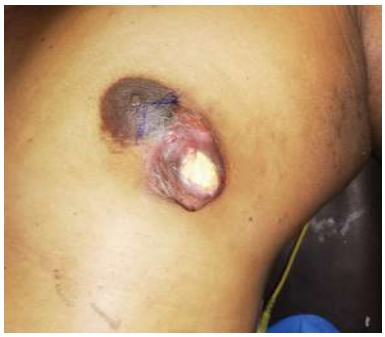

Fig. 1: Photograph showing bluish discolored swelling with ulceration in the left breast.

In view of hard lump, wide excision with primary closure was done. Post-operative period was uneventful. The histo-pathological examination revealed pilomatrixoma (Calcifying epithelioma of Malherbe) with the surgical margin free of tumour. On immunohistochemistry, p63 was found to be positive in basaloid cells.

Discussion

Pilomatrixoma, also known as pilomatricoma is a benign tumor of hair matrix differentiation. The first case was reported in 1880, by Malherbe and Chenantais[3] as a calcified tumour originating from the sebaceous glands, and was initially named 'calcifying epithelioma of Malherbe''. In 1961 the name was changed to 'pilomatrixoma' by Forbis and Helwig who established that the origin was from the outer sheath cell of the hair follicle root.[3]

Pilomatrixoma mainly occurs in the first two decades of life, though some authors have described a bimodal age distribution with a second smaller peak of occurrence in in the fifth and sixth decades. Most authors have reported a female preponderance of up to 2 : 1[4] with some stating that it occurs equally in both genders.[5] Common sites are the head (peri-auricular and juxta-parotid areas), neck or upper extremities. Occurrence over the breast is very rare (1:100000 individuals). These lesions can originate from the peri-areolar piliferous bulbs and mimic a breast malignancy. It presents clinically as a slow-growing, painless mass in the deep dermis or subcutaneous tissue. The skin over the swelling may appear normal, or have a bluish-red discoloration, and it may occasionally ulcerate. Such a tumour with ulcerated overlying skin has been referred to as a perforating pilomatrixoma.[6] On palpation it is found to be superficial, solitary, firm to hard and mobile.[3, 7] It's occurrence in the male breast is extremely rare, with only 9 cases reported in English literature according to Clark A et al.[1] These tumors can easily be misdiagnosed because of clinical and radiological resemblance to breast malignancy.[1,8,9]

The exact cause for the pilomatrixoma has not been identified though several theories have been proposed. A familial predisposition has been noted, as a genetic component has been postulated in the occurrence. Some external factors have also been considered in the etiology including trauma, insect bites and surgery.[10] Pilomatrixomas exhibit mutations of CTNNB1, which is the gene encoding β-catenin, a signaling protein involved in the development of the pilous follicles.[11] Active mutations of the beta-catenin gene have been reported in 75% of human pilomatrixomas.[12] Another hypothesis by Forbis and Helwig is that, pilomatrixomas are hamartomas.[13]

Typical cases of pilomatrixoma show normal overlying skin colour and are hard in consistency and small nodules. They typically measure between 3-30 mm in diameter.[14] Tumours more than 5 cm in size are generally regarded as giant.[15] They are usually solitary. Multiple lesions are associated with Gardner Syndrome, Turner Syndrome, myotonic dystrophy and sarcoidosis and are reported in 2-10% of cases. Atypical variants described in literature include lymphangiectatic, anetodermic and ulcerated.[14]

The diagnosis of breast neoplasms can be made by clinical examination, mammography, USG, USG-guided fine needle aspiration cytology/core needle biopsy. On mammogram, pilomatrixomas exhibit nodular opacities with pleomorphic coarse irregular calcifications. On USG, the lesions appear as hypoechoic nodules with irregular margins and hyperechoic spots (calcification). Imaging findings often overlap with characteristic findings of malignant breast neoplasms. In such cases an ultrasound-guided core needle biopsy is helpful in establishing the diagnosis.[8, 16]

On histology, pilomatrixomas characteristically show epithelial cells organized in nodular aggregates in a connective matrix with scattered inflammatory-like elements. The tumour is characterized by the presence of both basophilic cells derived from hair matrix cells, and shadow cells which are equivalent to the hair cortex.[14] Each nodular aggregate has two types of epithelial cells with different organization: densely packed basophilic cells in the peripheral part and eosinophilic cells known as "ghost" or "mummified" cells in the centre of the nodule. Hair, calcifications, foci of necrosis and multinucleated giant cells are rarely seen.[16] On immunohistochemistry, p63 was found to be

positive in basaloid cells [Fig 2, 3].

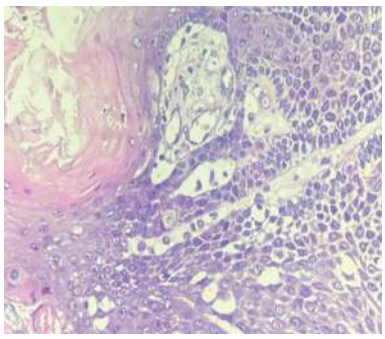

Fig. 2: Tumour shows sheets and strands of basaloid cells with peripheral palisading of nuclei.

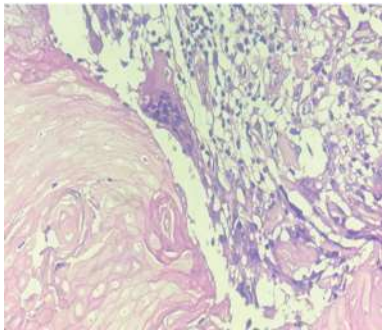

Fig. 3: Tumoue shows shadow cells in sheets and stroma shows foreign body granulomatous reaction.

The differential diagnosis includes sebaceous cyst, calcified giant hemangioma, giant dermatofibroma, atypical mycobacterial infection, ossifying/ calcified hematoma, giant cell tumor, foreign body reaction, cutaneous lymphoma, sarcoma, cutaneous metastasis, basal cell carcinoma, benign and malignant breast diseases.[3, 7, 17]

The treatment of benign Pilomatrixoma involves complete surgical excision of the lesion or wide excision when diagnosis is uncertain. Spontaneous regression has never been reported. Considering the benign nature of this lesion, the prognosis is excellent.[18]

The recurrence rate is around 4%, and could be related to incomplete excisions. There have also been speculations whether these tumours with recurrence were the rarer malignant pilomatrixomas which were misdiagnosed to be benign on initial evaluation. If there is recurrence, wider excision with detailed histopathologic examination is necessary.[17]

Malignant pilomatrixomas are extremely rare and were first described in 1980 by Lopansri et al.,.[19] Pilomatrix carcinoma (malignant pilomatricoma', 'trichomatrical carcinoma' or 'calcifying epitheliocarcinoma of Malherbe') is a very rare tumor and commonly seen in the head and neck region in adults. Typically it is larger than Pilomatrixoma and more common in older males (in contrast to pilomatrixoma, which shows a female predominance). Metastases are less frequent, but this condition has a high potential for recurrence.[19,20]

Conclusion

Pilomatrixoma is a rare, benign hair matrix tumor and rarely diagnosed in the male breast. Clinical and radiological findings of Pilomatrixoma overlap with characteristic findings of breast malignancy. It is therefore difficult to distinguish between Pilomatrixoma and other benign/malignant breast disease without complete evaluation including histopathology. Treatment involves complete surgical excision of the lesion, with recurrence of benign Pilomatrixoma being rare.

References

1. Clark A, Leddy R, Spruill L, Cluver A. Pilomatrixoma, a Rare Mimicker of Male Breast Cancer. J Clin Imaging Sci 2019;9:46.
2. Goldsmith LA, Katz SI, Gilchrist BA, Paller AS, Leffell DJ, Wolff K Ed. Fitzpatrick's Dermatology in General Medicine 8th Edition Ed. Chapter 19 Appendage tumours and Hamartomas of the skin. p1337. John Wiley and Sons Ltd, West Sussex, UK in 2016. Srivastava D, Taylor RS
3. Souto MP, Medeiros Matsushita M, Medeiros Matsushita G, Souto LR. An unusual presentation of giant pilomatrixoma in an adult patient. J Derm Case Rep. 2013; 7:56-59.
4. Saniasiaya J, Mohamad I, Kamaludin Z. Pilomatrixoma of the neck: A forgotten entity. Egyptian Journal of Ear, Nose, Throat and Allied Sciences 2017;18:311-12
5. Lan MY, Lan MC, Ho CY, Li WY, Lin CZ. Pilomatricoma of the head and neck: a retrospective review of 179 cases. Arch Otolaryngol Head Neck Surg. 2003 Dec;129(12):1327-30. doi: 10.1001/ archotol.129.12.1327. PMID: 14676160.
6. Thakur BK, Verma S, Mishra J. Perforating pilomatricoma in a 62-year-old female: A rare case report. Int J Trichol 2014;6:173-4
7. Sinhasan SP, Jadhav CR, Bhat RV, Amaranathan A. Pilomatrixoma – Presented as hypopigmented tender nodule: diagnosed by FNAC: A case report with review of literature. Indian J Derm. 2013; 58:405.
8. Hubeny CM, Sykes JB, O'Connell A, Dogra VS. Pilomatrixoma of the adult male breast: a

rare tumor with typical ultrasound features. J Clin Imaging Sci. 2011;1:12. [PMC free article] [PubMed]

9. Becker TS, Moreira MA, Lima LA, de Oliveira EL, Freitas-Júnior R. Pilomatrixoma mimicking breast cancer in man. Breast J. 2010 Jan-Feb;16(1):89–91. [PubMed]
10. Le C, Bedocs PM. Calcifying Epithelioma of Malherbe (Pilomatrixoma) [Updated 2020 Jun 25]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2020 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK493165/
11. Stefan Kraft and Scott R. Granter (2014) Molecular Pathology of Skin Neoplasms of the Head and Neck. Archives of Pathology & Laboratory Medicine: June 2014, Vol. 138, No. 6, pp. 759-787.
12. Chan EF, Gat U, McNiff JM, Fuchs E. A common human skin tumour is caused by activating mutations in beta-catenin. Nat Genet. 1999 Apr;21(4):410-3. doi: 10.1038/7747. PMID: 10192393.
13. Forbis R, Jr, Helwig Eb. Pilomatrixoma (calcifying epithelioma) Arch Dermatol. 1961 Apr;83:606–18.[PubMed]
14. Julian, C.G. and Bowers, P.W. A clinical review of 209 pilomatricomas. J Am Acad Dermatol. 1998; 39:191–195
15. Khammash, M.R., Todd, D.J., and Abalkhail, A. Concurrent pilomatrix carcinoma and giant pilomatrixoma. Australas J Dermatol. 2001; 42: 120–123
16. Pascual A, Casado I, Colmenero I, Pelayo A, Asenjo JA. Fine needle aspiration cytology of pilomatrixoma of the breast. Acta Cytol. 2000 Mar-Apr;44(2):274–6. [PubMed]
17. Whittemore K, Cohen M. Imaging and review of a large pre-auricular pilomatrixoma in a child. World J of Rad. 2012; 4:228-230.
18. Hamilton A, Young GI, Davis RI. Pilomatrixoma mimicking breast carcinoma. Br J Dermatol. 1987;116:585–586. [PubMed]
19. Lopansri S, Mihm MC., Jr Pilomatrix carcinoma or calcifying epitheliocarcinoma of Malherbe: A case report and review literature. Cancer. 1980;45:2368-73.
20. Lazar AJ, Calonje E, Grayson W, et al. Pilomatrix carcinomas contain mutations in CTNNB1, the gene encoding beta-catenin.J Cutan Pathol. 2005;32(2):148–157. [Crossref] [Medline] [Google Scholar]