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Breast Pilomatrixoma in a 47-Year-Old Woman: Pathological and Radiological Diagnosis

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ABSTRACT

Background: Pilomatrixoma is a benign adnexal tumor with differentiation towards the matrix of the hair follicle and occurs mostly in the head, neck, and upper extremities of the child. Pilomatrixoma of the breast is considered a very rare tumor.

Case presentation: Herein, we present a 47-year-old woman who was referred to the surgery ward for a palpable right breast mass. Imaging studies revealed a well-circumscribed lesion categorized as BIRADs3. Because clinically the lesion was large and had a stony hard consistency, the patient underwent surgery and intraoperative consultation was negative, revealing a well-defined lesion composed of basaloid cells, multinucleated giant cells admixed with bone trabeculae, and nests of ghost cells. Immunohistochemical (IHC) studies were done and the cells were positive for AE1/AE3, Beta-catenin, and BCL2 and negative for Mart1 and CD117. Based on H&E and IHC studies, pilomatrixoma of the breast tissue was confirmed.

Conclusion: Breast pilomatrixoma is a very rare benign neoplasm that could clinically mimic malignant neoplasm and should be considered in the differential diagnosis of breast masses.

INTRODUCTION

Pilomatrixoma (calcifying epithelioma of Malherbe) was first described by Malherbe and Chenatais in 1880 as a sebaceous gland origin, but in 1961, Forbis and Helwig proved that it originates from the hair matrix and was considered as a hamartoma.[1-11] Pilomatrixoma is a benign adnexal tumor of hair matrix cells.[1-17] The exact prevalence is unknown, but it accounts for nearly less than 1% of all benign skin tumors.[13] The most common sites are the head, neck, and upper extremities. It is less common in the trunk and lower extremities and very rarely reported in the breast. It can occur at any age but is more common in children and young adults, and again after the age of 60, with a higher incidence in women.[1-15] The etiology of pilomatrixoma is unclear, but it may be related to the inflammatory process as a result of repeated traumas.[1,3,8,10] To the best of our knowledge, there are only a few reported cases in the literature for breast pilomatrixoma.[1-17] Herein, we report the case of a 47-year-old woman presenting with a right breast mass diagnosed as breast pilomatrixoma, followed by a brief discussion of its histopathological and imaging characteristic.

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CASE PRESENTATION

A 47-year-old woman was referred to the surgery ward with a complaint of a palpable mass in the right breast from 1 year ago. On physical examination,

there was a painless and firm lump in the right breast. No skin abnormalities were observed. Mammography revealed benign calcification in the right breast (BIRADs=0) and ultrasound studies showed a lobulated hypoechoic area measuring 37x8 mm at 12 o'clock and 14.7x5 mm at 10 o'clock in the left breast and 10.9x6.8 mm at 10 o'clock in the right breast, which looked like several lumps in the vicinity of each other, primarily indicative of benign breast lesions (BIRADs3). There was a well-defined hypoechoic area measuring 21x15 mm in the right axillary area (Figure 1).

The patient underwent surgery with informed consent and the sample was sent for a frozen section. The received specimen consisted of yellow-colored tissue measuring 8x5x2cm with the overlying skin tissue that measured 6x3cm. On the section, there was a well-circumscribed gray-colored lesion measuring 2.5cm in diameter with bony consistency. Microscopic examination revealed a tumoral lesion composed of nests of dark round cells, ghost cells admixed with multinucleated giant cells, and bone trabeculae (Figures 2a, 2b, 2c,2d).

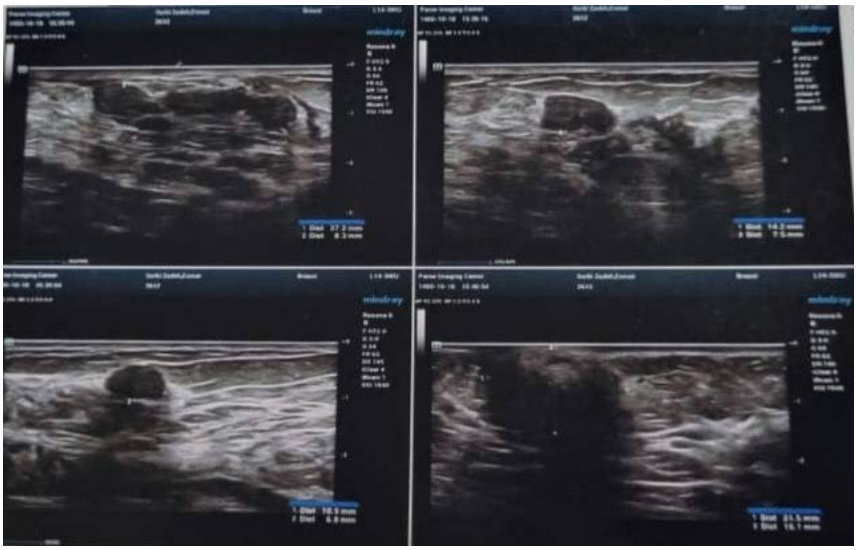
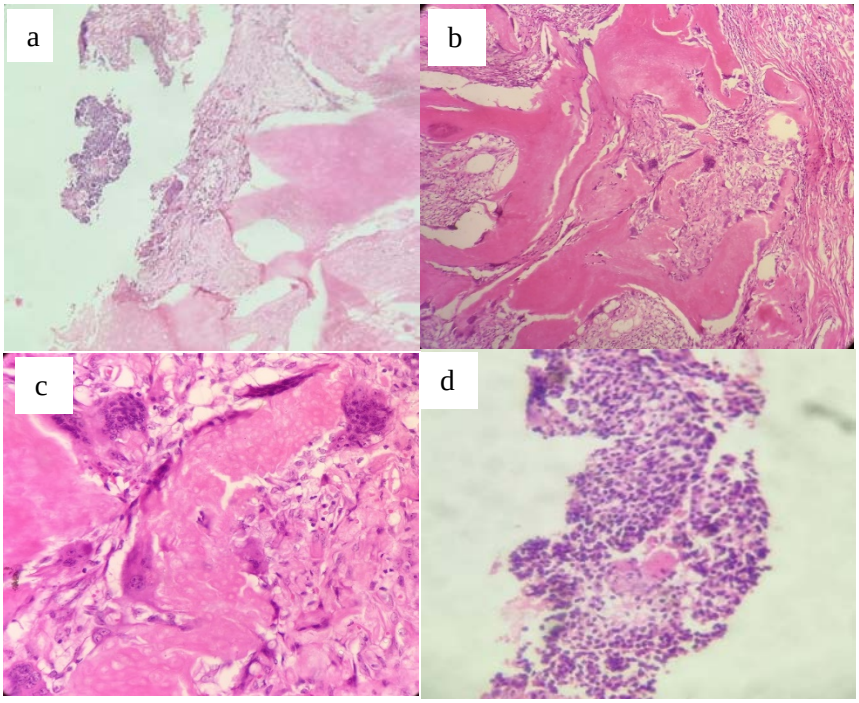
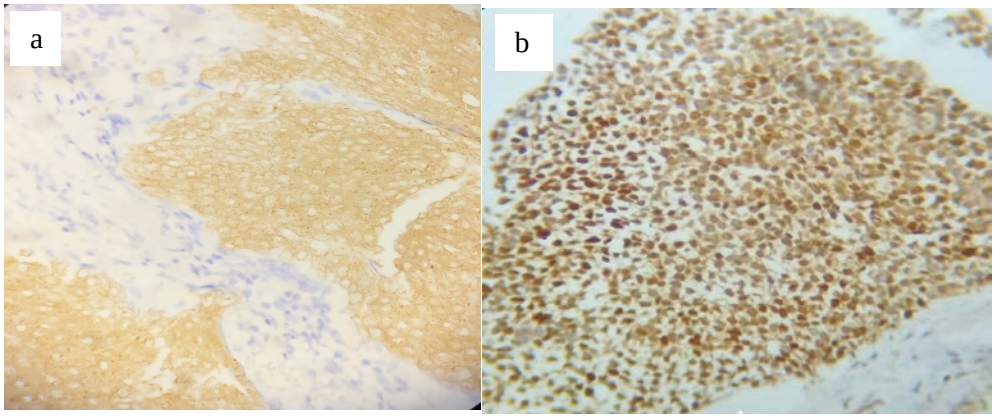


Figure 1. Sonography revealed a lobulated hypoechoic area measuring 10.9 x 6.8 mm in 10 o'clock of right breast, which looks like several lumps in the vicinity of each other (BIRADs 3).



Figures 2. (a): A tumoral lesion composed of basaloid cells admixed with ghost cells with bone trabeculae (hematoxylin & eosin staining, 40x). (b): Ghost cells and multinucleated giant cells with bone trabeculae (hematoxylin & eosin staining, 100x). (c): Ghost cells and multinucleated giant cells (hematoxylin & eosin staining, 400x). (d): Basaloid cells (hematoxylin & eosin staining, 400x).



Figures 3. (a): Positive reaction for AE1/AE3 (immunohistochemical staining, 400 x magnification). (b): Positive reaction for Beta catenin (immunohistochemical staining, 400 x magnification)

The permanent section showed a sclerotic nodule. In terms of histology, the tumor was completely compatible with pilomatrixoma and only immunohistochemistry (IHC) was used to confirm the tumor and to rule out breast metaplastic carcinoma as well as round cell neoplasms. The basaloid cells were positive for AE1/AE3, Beta-catenin, and BCL2 and negative for CD45, Mart1 and CD117 (Figure 3a, 3b). The surgeon also dissected the right axillary lymph nodes suspicious of malignancy, which were negative for malignancy and showed reactive lymphoid follicular hyperplasia. Based on the H&E and IHC studies, pilomatrixoma of the breast tissue was confirmed.

DISCUSSION

Pilomatrixoma of the breast is very rare, originating from the hair matrix cells, and comprises 1% of all benign skin tumors.[1-17] It could be misdiagnosed as a malignant lesion clinically, so radiological imaging and histopathological findings should be evaluated together.[1] They are usually asymptomatic, have a firm consistency, and are revealed as solitary, slow-growing, non-tender lesions measuring 3mm to 30mm in diameter. Sasaki *et al.* reported a parotid pilomatrixoma with dimensions of 8x5cm.[18] The overlying skin can be normal or inflamed.[1-6,8] A pathogonomic sign of pilomatrixoma is the stretching of the skin over the tumor. It is called a "tent sign" and another characteristic feature is the blue-red discoloration of the overlying skin.[1,3]

Tokur *et al.* reported a breast pilomatrixoma in a 42-year-old male that was palpable firm and painless mass in the retroareolar area with no skin abnormalities and axillary lymphadenopathy.[1] Our case was a 47-year-old woman, who was presented with a palpable, painless, and firm mass without skin abnormality and with axillary lymphadenopathy. Clinical types of pilomatrixoma include ulcerative lesions, multiple eruptive and familial cases, superficial pilomatrixoma presenting as a cutaneous horn or bullous lesion, and giant pilomatrixoma. Multiple lesions account for 2-10% of cases and are associated with Gardner syndrome, Turner syndrome, Myotonic dystrophy, and sarcoidosis.[2,3,15] Lesions larger than 5 cm are named "giant" pilomatrixoma and can cause pain and tenderness. The tumor may be misdiagnosed clinically with hemangiomas or neurofibromas.[2] Imaging findings of the breast pilomatrixoma are similar to malignant lesions, so the diagnosis is confirmed by histopathology. On mammography, they are presented as amorphous and well-defined lesions with irregular coarse calcifications that can increase in size over time. They are usually diagnosed as BIRADs III to IV. In sonography, they are well-defined, hyperecho or isoecho lesions with a hypoechoic rim and posterior acoustic shadowing.[1,2,5] There are many differential diagnoses for breast pilomatrixoma and it is difficult to distinguish them from each other, which include seborrheic keratosis, inclusion cyst, epidermoid cyst, calcified giant hemangioma, foreign body reaction, sarcoma, squamous and basal cell carcinoma, calcified fibroadenoma, fibrocystic changes, fat necrosis, ductal hyperplasia, adenosis, apocrine metaplasia, papillomas, lobular neoplasia, metaplastic carcinoma and invasive ductal carcinoma.[1-3,7,8,10,12,13,15] The basaloid cells of pilomatrixoma have a positive reaction for pancytokeratin, Beta catenin, BCL2, CDX2, ER, PR and a negative reaction for Mart1, and SOX10 in immunohistochemical studies. Complications of pilomatrixoma are rare. However, sometimes they can get too big, and very rarely become malignant. Malignant transformation of pilomatrixoma should be distinguished from proliferating pilomatrixoma which is a benign tumor with lobular proliferation of

basaloid cells and shadow cells with nuclear atypia and variable mitosis.[1, 3, 8] Pai *et al.* reported a malignant pilomatrixoma in a 49-year-old woman who presented with a palpable mass measuring 15 cm involving all the quadrants of the left breast.[4] Pilomatrixoma of the breast was reported mainly in males and it is very rare in females.[1] Spontaneous regression of the breast pilomatrixoma is rare and surgical excision is the treatment of choice, and in cases where mammographic or ultrasound findings are benign, conservative treatment is suggested.[1, 3, 14, 15] In our case, surgical excision was performed. Recurrence after surgical treatment is rare, with a rate of 2-6%, and is usually the result of incomplete excision. Recurrence should make the surgeon suspicious of malignant pilomatrixoma.[2, 9, 15]

CONCLUSION

Pilomatrixoma of the breast is a very rare breast lesion and can be misdiagnosed as a breast neoplasm. It should be considered in the differential diagnosis of the breast masses. Breast pilomatrixoma cannot be reliably diagnosed by mammographic and sonographic features alone and clinical examination, radiological features, and histopathological findings should be evaluated together to avoid more invasive surgeries and unnecessary removal of axillary lymph nodes.

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ETHICAL CONSIDERATIONS

Not applicable.

CONFLICT OF INTERESTS

The authors declare no conflict of interest.

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