


Pilomatrixoma of the breast, a rare lesion simulating breast cancer: a case report

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ABSTRACT

Pilomatrixoma or calcifying epithelioma of Malherbe is a rare, benign, skin tumour originating from piliferous follicles; breast localization is considered to be very rare. These lesions can originate from the peri-areolar piliferous bulbs and, due to the clinical and imaging features, be easily misdiagnosed as a breast neoplasm. We present a case of pilomatrixoma of the left breast in a woman of 43 years appearing as a firm, deep nodule in the external quadrants. The lesion had mammographic and sonographic malignant features, but histological analysis on core-needle biopsy and surgical specimens revealed this unusual benign lesion.

CASE REPORT

CASE REPORT

A 43-year old woman came to our Senology Department to be investigated for a centimetre sized, non-tender breast lump that had a hard consistency and was firm on the deep layers of the breast. The nodule was localized in the peri-areolar area at the confluence of the outer quadrants of the left breast. Physical examination of the axillae was negative.

IMAGING FINDINGS

Mammography in two standard projections, cranio-caudal and medial-lateral-oblique (Fig. 1), showed, in a context of a dense breast, a round nodule containing a cluster of pleomorphic irregular microcalcifications that were classified as ACR BI-RADS IV-V. The size of the nodule was 12x11 mm.

The lesion could not be seen on the previous mammograms performed 12 months earlier (Fig. 2). The ultrasound examination (Fig. 3A) showed a hypoechoic nodule of 13x14x10 mm, with irregular peripheral margins and hyperechoic spots compatible with microcalcifications. There were no signs of intralesional vascularization at the Doppler analysis and no locoregional lymphadenopathy was found.

In consideration of the imaging features and the absence of any abnormality in the previous mammograms, the lesion was highly suspicious for malignancy

MANAGEMENT

A core-needle biopsy (CNB) was performed using a 14-gauge cutting needle with a 22-mm throw (Precisa HS Hospital Service, Rome) sampling three cores of tissue (Fig. 3B). The micro-histological diagnosis was of a pilomatrixoma.

The breast surgical team, considering the imaging findings, decided to perform the excision of the lesion as a precaution. The post-operative histological diagnosis of the surgical specimen confirmed the diagnosis of pilomatrixoma (Fig. 3C).

FOLLOW-UP

No recurrence was seen after 24 months (Fig.4). Patient was followed-up every year with mammography and ultrasound according to the Italian guidelines and to the American College of Radiology recommendations.

DISCUSSION

ETIOLOGY AND DEMOGRAPHICS

Pilomatrixoma is a benign epithelial tumor of the skin originating from piliferous follicles [1]. Originally named as "calcific epithelioma of Malherbe", it was described for the first time in 1880 by Dr. Chenantals Malherbe who considered it as a calcific epithelioma of the sebaceous glands [2]. Pilomatrixomas usually develop in the subcutaneous tissue from the cellular matrix of hair follicles.

They are usually found in 1st and 2nd decades although they have been described, with a lower incidence, in the 5th and 6th decades.

The etiology has not been established yet but it seems that repeated skin traumas could be the main causes. These stimuli induce an inflammatory response that leads to an overgrowth of hair matrix. Another hypothesis, formulated by Forbis and Helwig, is that pilomatrixomas are hamartomas [3]. Breast localization is very rare (1:100000 people), however, these lesions can originate from the peri-areolar piliferous bulbs and mimic a breast malignancy [2;5-15].

Solitary and multifocal diseases have been reported; in addition these lesions could be sporadic or associated to syndromes such as Gardner syndrome and myotonic dystrophy [4]. Histologically pilomatrixomas are made of epithelial cells organized in nodular aggregates into a connective matrix with scattered inflammatory-like elements. Each nodule is characterized by two types of epithelial cells with different organization: on the peripheral part there are densely packed basophilic cells producing keratin while the center of the nodule is occupied by eosinophilic cells known as "ghost" or "mummified" cells. Moreover, hair, calcifications, foci of necrosis and multinucleated giant cells can be often found [5].

CLINICAL AND IMAGING FINDINGS

Pilomatrixoma is most commonly diagnosed in the head (peri-auricular and juxta-parotid areas), neck, upper and lower extremities and more rarely on the trunk [6]. Breast localization is very rare and only a very few cases have been reported so far [2;5-15]. While lesions in the skin are often easily diagnosed, breast pilomatrixomas require several diagnostic tests due to the difficulty of distinguishing these lesions from breast cancer.

Clinically, breast pilomatrixomas can range in size from 3 to 30 mm. On physical examination they appear as hard nodules with lobulated margins. In addition there may be other signs such as rubor, calor, dolor due to inflammatory phenomena. The skin can be intact, purple or rarely ulcerated [8,16]. In the latter case the differential diagnosis with malignant skin lesions can be difficult. Malignant pilomatrixomas are extremely rare; they have been described the first time in 1980 by Lopansri et al [17] and only a few articles are available reporting this type of disease. [18,19, 20].

The diagnostic management of breast pilomatrixomas should include clinical examination, mammography, ultrasound and ultrasound-guided CNB.

On mammograms, breast pilomatrixomas appear as nodular opacities with pleomorphic coarse irregular calcifications (ACR BI-RADS IV-V) whose number can increase gradually, simulating the microcalcifications often associated with breast cancer. Sonographically, these lesions appear as hypoechoic nodules with irregular margins, hyperechoic spots and a posterior acoustic shadowing (Tab 1).

TREATMENT AND PROGNOSIS

Imaging findings overlap often those that are characteristic of breast neoplasms. This necessarily leads to an ultrasound-guided CNB which usually proves the biology of the nodule.

The breast pilomatrixoma that came to our observation is unusual and rarely seen. Classic forms of intramammary pilomatrixomas show generally coarse calcification [11], while the lesion we report had granular polymorphic microcalcifications that were considered BI-RADS IV-V and that were not present in the previous examinations.

Despite the percutaneous biopsy, we recommended the surgical excision of the lesion that confirmed the diagnosis of the core-needle biopsy.

Considering the benignity of this lesion, the expected prognosis is excellent.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis of breast pilomatrixoma with both benign and malignant breast lesions can be very challenging. It includes skin calcifying lesions (seborrheic keratosis and inclusion cysts), fibrocystic changes (usual ductal hyperplasia, adenosis, apocrine metaplasia), lobular neoplasias, papillomas, calcified fibroadenomas, fat necrosis and invasive ductal carcinoma as all can appear as nodular opacities with pleomorphic calcifications on mammography (Tab.2).

Differential diagnosis with ultrasound is also difficult as pilomatrixomas mimic both benign nodules (fibroadenomas, papillomas) and invasive ductal carcinoma (Tab. 2).

MRI features of those more common lesions that can be simulated by a breast pilomatrixoma are well-known, no data are currently available regarding the MRI appearance of this rare disease (Tab.2).

In addition, despite the presence of cells considered typical of pilomatrixomas (ghost cells and basaloid cells), differential diagnosis with breast cancer by cytology only can be challenging in most cases with a high rate of false positives (75%) [21-24]. On the other hand, core-needle biopsy has proved to be an effective and safe technique for diagnosing breast nodules [25] and should be performed to obtain a pre-surgical diagnosis. In conclusion, breast pilomatrixomas,

despite their rarity, should be considered in the differential diagnosis of calcified nodules visible on mammograms and an excisional biopsy for a definitive diagnosis might be advisable.

TEACHING POINT

Pilomatrixomas can have an intramammary localization and, although they are very rare, should be considered in the differential diagnosis of breast nodules that show calcifications on mammograms. Standard examination with mammography and ultrasound should be followed by core-needle biopsy in order to have a precise pre-operative diagnosis.

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FIGURES

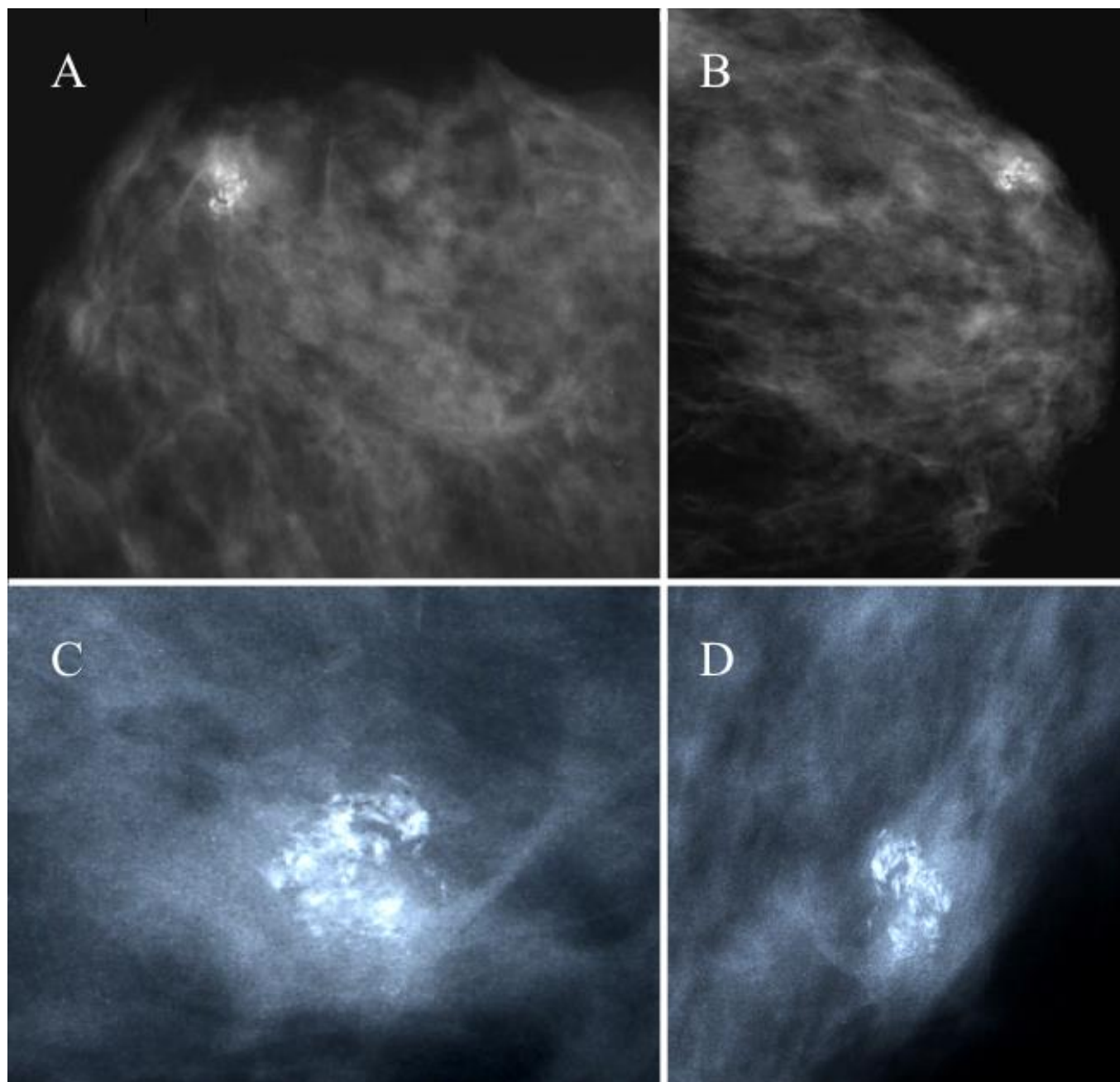


Figure 1: 43 year old female with a hard, firm nodule at the confluence of the external quadrants of the left breast diagnosed as a breast pilomatrixoma after percutaneous and excisional biopsy.

FINDINGS: nodular opacity in the external quadrants of the left breast of 12x11 mm, showing a cluster of pleomorphic irregular microcalcifications (ACR BI-RADS IV-V).

TECHNIQUE: Analogic mammography (28 kV, 100 mAs), cranio-caudal (A) and medio-lateral oblique projections (B) of the left breast. Magnification views of cranio-caudal (C) and medio-lateral oblique (D) projections showing the lesion.

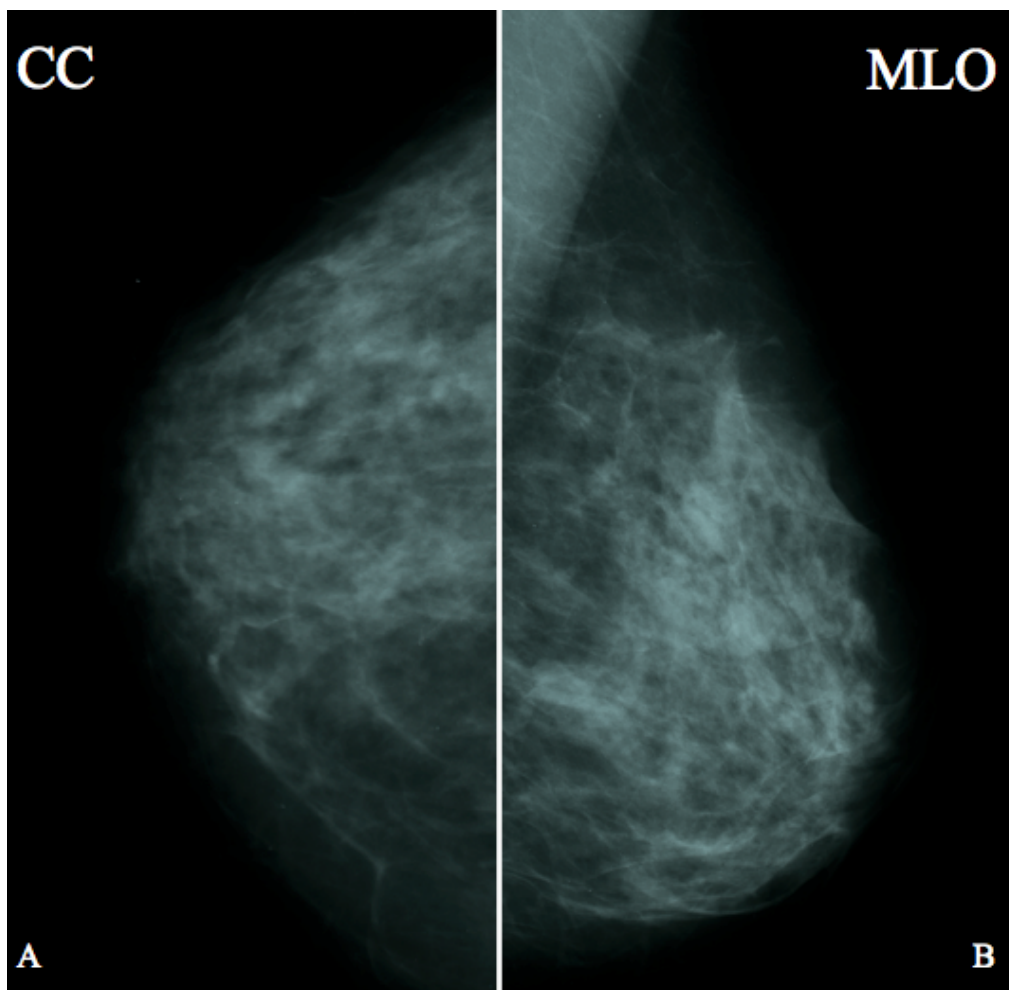


Figure 2: 43 year old female one year before developing a breast pilomatrixoma at the confluence of the external quadrants of the left breast.

FINDINGS: No significant abnormality to report.

TECHNIQUE: Analogic mammography (28 kV, 100 mAs), cranio-caudal (A) and medio-lateral oblique projections of the left breast.

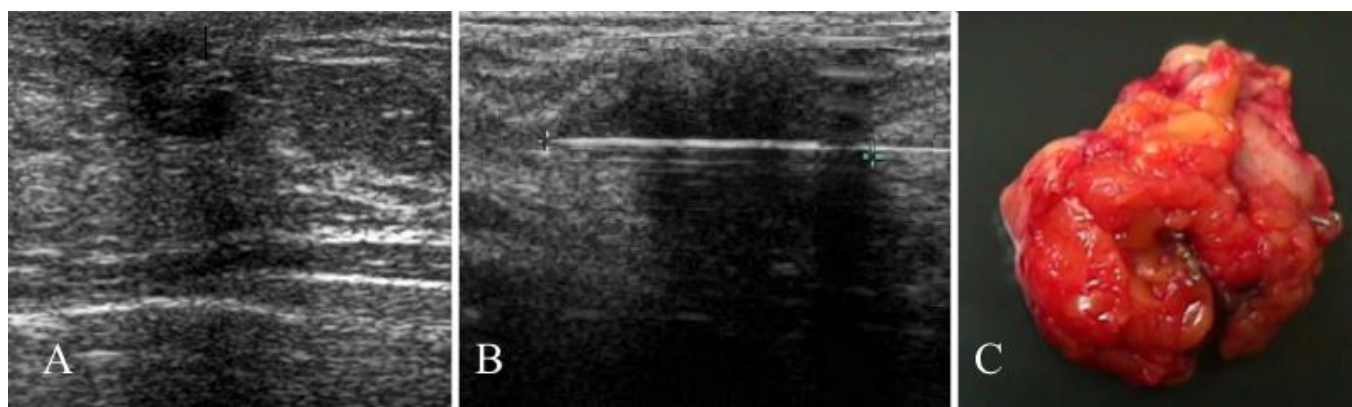


Figure 3: 43 year old female with a hard, firm nodule at the confluence of the external quadrants of the left breast diagnosed as a breast pilomatrixoma after percutaneous and excisional biopsy.

FINDINGS: hypoechoic nodule (13x14x10 mm) showing irregular margins, hyperechoic spots due to microcalcifications and posterior acoustic shadowing (A). Ultrasound-guided 14-G core-needle biopsy of the lesion (B). Surgical specimen (C).

TECHNIQUE: ultrasound examination using a broadband 10-13 MHz linear transducer.

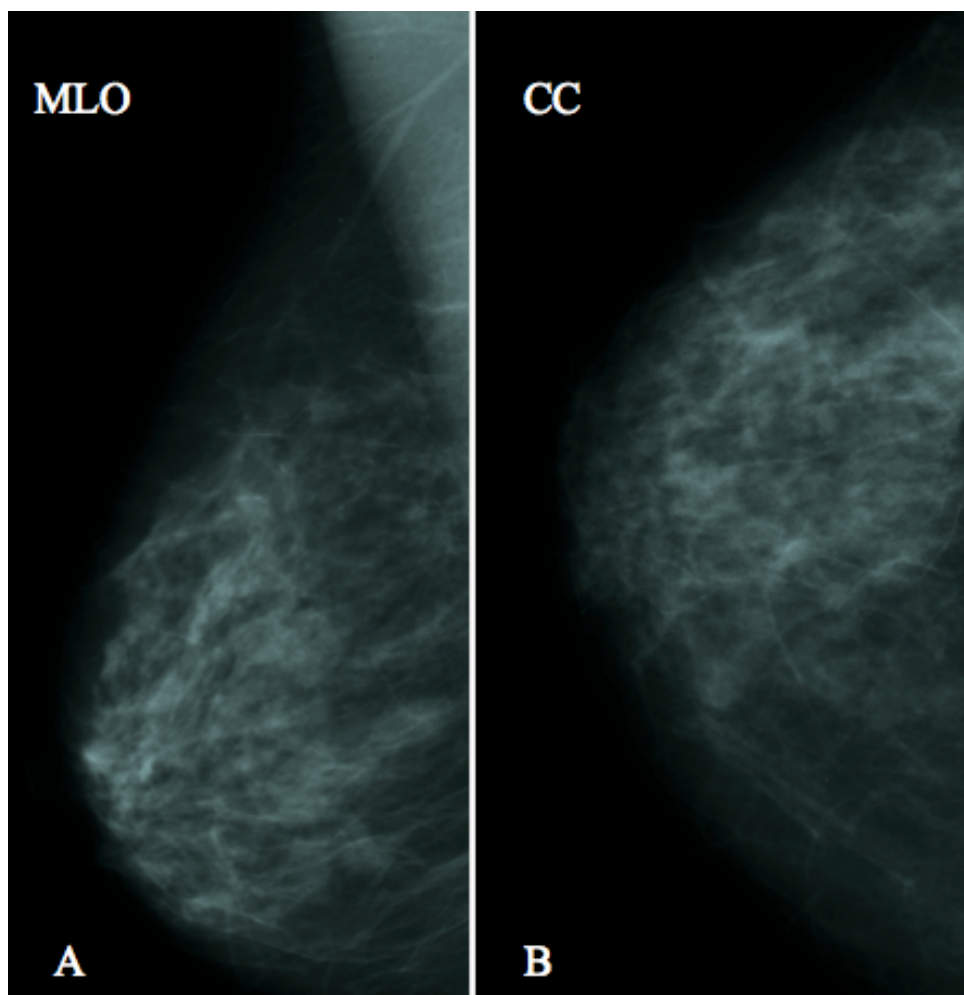


Figure 4: 43 year old female two years after the excision of a breast pilomatrixoma at the confluence of the external quadrants of the left breast.

FINDINGS: No significant abnormality to report.

TECHNIQUE: Analogic mammography, cranio-caudal (A) and medio-lateral oblique projections of the left breast.

Etiology	Unknown. Repeated traumas on skin may lead to an overgrowth of hair matrix.
Incidence	Very rare (1:100000 people)
Gender Ratio	More frequent on women
Age Predilection	1st and 2nd decades
Risk Factors	Unknown
Treatment	Surgical excision
Prognosis	Excellent
Findings on Imaging	Mammographic findings: nodular opacity with pleomorphic heterogeneous microcalcifications (ACR BI-RADS IV-V). Ultrasonographic findings: hypoechoic nodule with irregular margins, posterior acoustic shadowing and hyperechoic spots due to microcalcifications.

Table 1: Summary table for breast pilomatrixoma

Lesion	Imaging modality		
	Mammography	Ultrasound	MRI
Skin calcifying lesions (seborrheic keratosis, inclusion cysts)	<ul style="list-style-type: none"> Well circumscribed superficial mass. Heterogenous calcifications. 	<ul style="list-style-type: none"> Inclusion cysts have a solid or hypoechoic appearance with or without hyperechoic spots due to calcifications. Surrounding vascularity may be seen if inflammation is present. 	<ul style="list-style-type: none"> T1WI: hypointense cystic component. T2WI FS: hyperintense cystic component. T1 C.E.: rim enhancement when inflammation is present.
Fibrocystic changes (usual ductal hyperplasia, adenosis, apocrine metaplasia)	<ul style="list-style-type: none"> Circumscribed lobulated low density mass with pleomorphic calcifications. Clustered pleomorphic, punctate or amorphous calcifications without mass. 	<ul style="list-style-type: none"> Simple, complicated, clustered cysts or complex cystic and solid masses showing scattered echogenic foci due to microcalcifications and fibrosis. 	<ul style="list-style-type: none"> T1WI: cysts typically isointense to parenchyma. Variable signal when cysts contain proteinaceous or hemorrhagic fluid. T2WI FS: hypointense to hyperintense signal due to the content of the cysts. T1C.E. FS: possible smooth rim enhancement of cysts when inflamed. Scattered and diffuse foci (< 5 mm) of enhancement.
Lobular neoplasia	<ul style="list-style-type: none"> Amorphous calcifications. Spiculated mass or architectural distortion. Multifocal/multicentric lesions. 	<ul style="list-style-type: none"> Irregular hypo- to anechoic mass with or without hyperechoic spots due to calcifications. 	<ul style="list-style-type: none"> T1 C.E. FS: non-mass ductal enhancement or irregular enhancing mass.
Papilloma	<ul style="list-style-type: none"> Oval or round mass centrally or peripherally located with/without clustered pleomorphic calcifications. 	<ul style="list-style-type: none"> Intraductal mass near nipple with lobulated margins. Dilated ducts around the lesion. Internal vascularity on Doppler analysis. 	<ul style="list-style-type: none"> T2WI: hyperintense duct with intraductal hypointense mass. T1C.E. FS: variable weak enhancement to malignant enhancement profile.
Fibroadenoma	<ul style="list-style-type: none"> Oval or lobulated mass with/without coarse calcifications (clustered or "pop-corn" shaped). 	<ul style="list-style-type: none"> Hypoechoic circumscribed oval mass with/without internal hyperechoic spots. 	<ul style="list-style-type: none"> T2WI FS: isointense with parenchyma or moderately hyperintense when myxoid. T1 C.E. FS: oval or lobulated mass showing generally rapid, homogeneous intense enhancement.
Fat necrosis	<ul style="list-style-type: none"> Round, oval or lobulated radiolucent mass. Pleomorphic calcifications. Spiculated or irregular mass. 	<ul style="list-style-type: none"> Anechoic/irregular hypoechoic/complex cystic/solid mass. Posterior shadowing once fibrosis and calcifications develop. Internal flow may be seen up to 6 months after surgery due to granulation tissue. 	<ul style="list-style-type: none"> T1WI: high signal. T2WI FS: low signal with fat suppression. T1 C.E. FS: thin rim of peripheral enhancement may persist up to 18 months post-surgery. Rarely contrast enhancement may persist for years.
Invasive ductal carcinoma (IDC)	<ul style="list-style-type: none"> Dense mass with spiculated/lobulated margins. Focal asymmetric mass/distortion. Clustered irregular, branched, pleomorphic calcifications. 	<ul style="list-style-type: none"> Irregular hypoechoic mass with posterior shadowing. "Taller than wide". Architectural distortion with/without hyperechoic halo Rarely lobulated, rounded hypoechoic nodule. 	<ul style="list-style-type: none"> T2WI FS: usually hypointense focal mass if visible. Central necrosis can be hyperintense. T1 C.E. FS: usually rapid and intense enhancement. May show rim enhancement, internal enhancing septations and rapid washout.
Pilomatrixoma	<ul style="list-style-type: none"> Round, lobulated nodule with pleomorphic, irregular microcalcifications. 	<ul style="list-style-type: none"> Hypoechoic nodule with lobulated/irregular margins and hyperechoic internal spots. 	<ul style="list-style-type: none"> No data available.

Table 2: Differential diagnosis of breast pilomatrixoma

ABBREVIATIONS

ACR BI-RADS = American College of Radiology, Breast Imaging Reporting and Data System
CNB = core needle biopsy
IDC = invasive ductal carcinoma
MRI = magnetic resonance imaging
T1/T2 WI = T1/T2-weighted images
T2 WI FS = T2-weighted images with fat suppression
T1 C.E FS = T1-weighted contrast-enhanced images with fat suppression

KEYWORDS

breast; breast pilomatrixoma; breast calcified nodules; core needle biopsy

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