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 origin 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

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 organised 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).
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.

REFERENCES


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

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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: Hypoechogenic 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.
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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.

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

Table 1: Summary table for breast pilomatrixoma
<table>
<thead>
<tr>
<th>Lesion</th>
<th>Mammography</th>
<th>Imaging modality</th>
<th>MRI</th>
</tr>
</thead>
<tbody>
<tr>
<td>Skin calcifying lesions (seborrheic keratosis, inclusion cysts)</td>
<td>Well circumscribed superficial mass.</td>
<td>• Inclusion cysts have a solid or hypoechoic appearance with or without hypoechoic spots due to calcifications.</td>
<td>• T1WI: hypointense cystic component.</td>
</tr>
<tr>
<td></td>
<td>• Heterogenous calcifications.</td>
<td>• Surrounding vascularity may be seen if inflammation is present.</td>
<td>• T2WI FS: hyperintense cystic component.</td>
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<td></td>
<td></td>
<td></td>
<td>• T1 C.E.: rim enhancement when inflammation is present.</td>
</tr>
<tr>
<td>Fibrocystic changes (usual ductal hyperplasia, adenosis, apocrine metaplasia)</td>
<td>Circumscribed lobulated low density mass with pleomorphic calcifications.</td>
<td>• Simple, complicated, clustered cysts or complex cystic and solid masses showing scattered echogenic foci due to microcalcifications and fibrosis.</td>
<td>• T1WI: cysts typically isointense to parenchyma.</td>
</tr>
<tr>
<td></td>
<td>• Clustered pleomorphic, punctate or amorphous calcifications without mass.</td>
<td></td>
<td>• Variable signal when cysts contain proteinaceous or hemorrhagic fluid.</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>• T2WI FS: hypointense to hyperintense signal due to the content of the cysts.</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>• T1.C.E. FS: possible smooth rim enhancement of cysts when inflamed.</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>• Scattered and diffuse foci (&lt; 5 mm) of enhancement.</td>
</tr>
<tr>
<td>Lobular neoplasia</td>
<td>Amorphous calcifications.</td>
<td>• Irregular hypo-to anechoic mass with or without hypoechoic spots due to calcifications.</td>
<td>• T1 C.E. FS: non-mass ductal enhancement or irregular enhancing mass.</td>
</tr>
<tr>
<td></td>
<td>• Spiculated mass or architectural distortion.</td>
<td></td>
<td></td>
</tr>
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<td></td>
<td>• Multifocal/multicentric lesions.</td>
<td></td>
<td></td>
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<tr>
<td>Papilloma</td>
<td>Oval or round mass centrally or peripherally located with/without clustered pleomorphic calcifications.</td>
<td>• Intraductal mass near nipple with lobulated margins.</td>
<td>• T2WI: hyperintense duct with intraductal hypointense mass.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Dilated ducts around the lesion.</td>
<td>• T1.C.E. FS: variable weak enhancement to malignant enhancement profile.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Internal vascularity on Doppler analysis.</td>
<td></td>
</tr>
<tr>
<td>Fibroadenoma</td>
<td>Oval or lobulated mass with/without coarse calcifications (clustered or &quot;pop-corn&quot; shaped).</td>
<td>• Hypoechoic circumscribed oval mass with/without internal hypoechoic spots.</td>
<td>• T2WI FS: isointense with parenchyma or moderately hyperintense when myxoid.</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>• T1 C.E. FS: oval or lobulated mass showing generally rapid, homogeneous intense enhancement.</td>
</tr>
<tr>
<td>Fat necrosis</td>
<td>Round, oval or lobulated radiolucent mass.</td>
<td>• Anechoic/irregular hypoechoic/complex cystic/solid mass.</td>
<td>• T1WI: high signal.</td>
</tr>
<tr>
<td></td>
<td>• Pleomorphic calcifications.</td>
<td>• Posterior shadowing once fibrosis and calcifications develop.</td>
<td>• T2WI FS: low signal with fat suppression.</td>
</tr>
<tr>
<td></td>
<td>• Spiculated or irregular mass.</td>
<td>• Internal flow may be seen up to 6 months after surgery due to granulation tissue.</td>
<td>• T1 C.E. FS: thin rim of peripheral enhancement may persist up to 18 months post-surgery.</td>
</tr>
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<td>Rarely contrast enhancement may persist for years.</td>
</tr>
<tr>
<td>Invasive ductal carcinoma (IDC)</td>
<td>Dense mass with spiculated/lobulated margins.</td>
<td>• Irregular hypoechoic mass with posterior shadowing.</td>
<td>• T2WI FS: usually hypointense focal mass if visible. Central necrosis can be hyperintense.</td>
</tr>
<tr>
<td></td>
<td>• Focal asymmetric mass/distortion.</td>
<td>• &quot;Taller than wide&quot;.</td>
<td>• T1 C.E. FS: usually rapid and intense enhancement.</td>
</tr>
<tr>
<td></td>
<td>• Clustered irregular, branched, pleomorphic calcifications.</td>
<td>• Architectural distortion with/without hypoechoic halo</td>
<td>• May show rim enhancement, internal enhancing septations and rapid washout.</td>
</tr>
<tr>
<td>Pilomatrixoma</td>
<td>Round, lobulated nodule with pleomorphic, irregular microcalcifications.</td>
<td>• Hypoechoic nodule with lobulated/irregular margins and hypoechoic internal spots.</td>
<td>• No data available.</td>
</tr>
</tbody>
</table>

Table 2: Differential diagnosis of breast pilomatrixoma
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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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