

BRIEF REPORT

Myofibroblastoma of the breast: 3 case reports and review of literature



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KEYWORDS

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Abstract Myofibroblastoma (MFB) is a rare, benign mesenchymal tumor of the breast. Three cases of breast MFB diagnosed in our clinical institution are presented, aiming to describe its clinical and radiologic characteristics, with a short literature review.

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PALABRAS CLAVE

Miofibroblastoma de mama;
Lesión mesenquimal;
Lesión hiperecoica;
Lesión benigna de la mama

Miofibroblastoma de mama: 3 casos clínicos y revisión de la literatura médica

Resumen El miofibroblastoma (MFB) es un tumor mesenquimal benigno y raro de la mama. Presentamos tres casos de MFB de mama diagnosticados en nuestra institución clínica, con el objetivo de describir sus características clínicas y radiológicas, con una breve revisión de la literatura médica.

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Introduction

Myofibroblastoma (MFB) is a benign mesenchymal tumour composed of myofibroblasts and fibroblasts. It accounts for less than 1% of breast tumours, but its exact prevalence is not known with certainty. MFB is predominantly diag-

nosed in postmenopausal women and elderly men, with no predominance of any race.¹ It frequently manifests as a slow-growing, painless and mobile unilateral palpable mammary gland mass, and it can also appear in extra-mammary locations, along the milk line.² MFBs can be difficult to differentiate radiologically from other mesenchymal tumours such as hamartomas, fibroadenomas or lipomas, since the findings of diagnostic imaging tests tend to be nonspecific.^{2,3} Under histological examination, most MFBs exhibit uniform spindle cells arranged in fascicles

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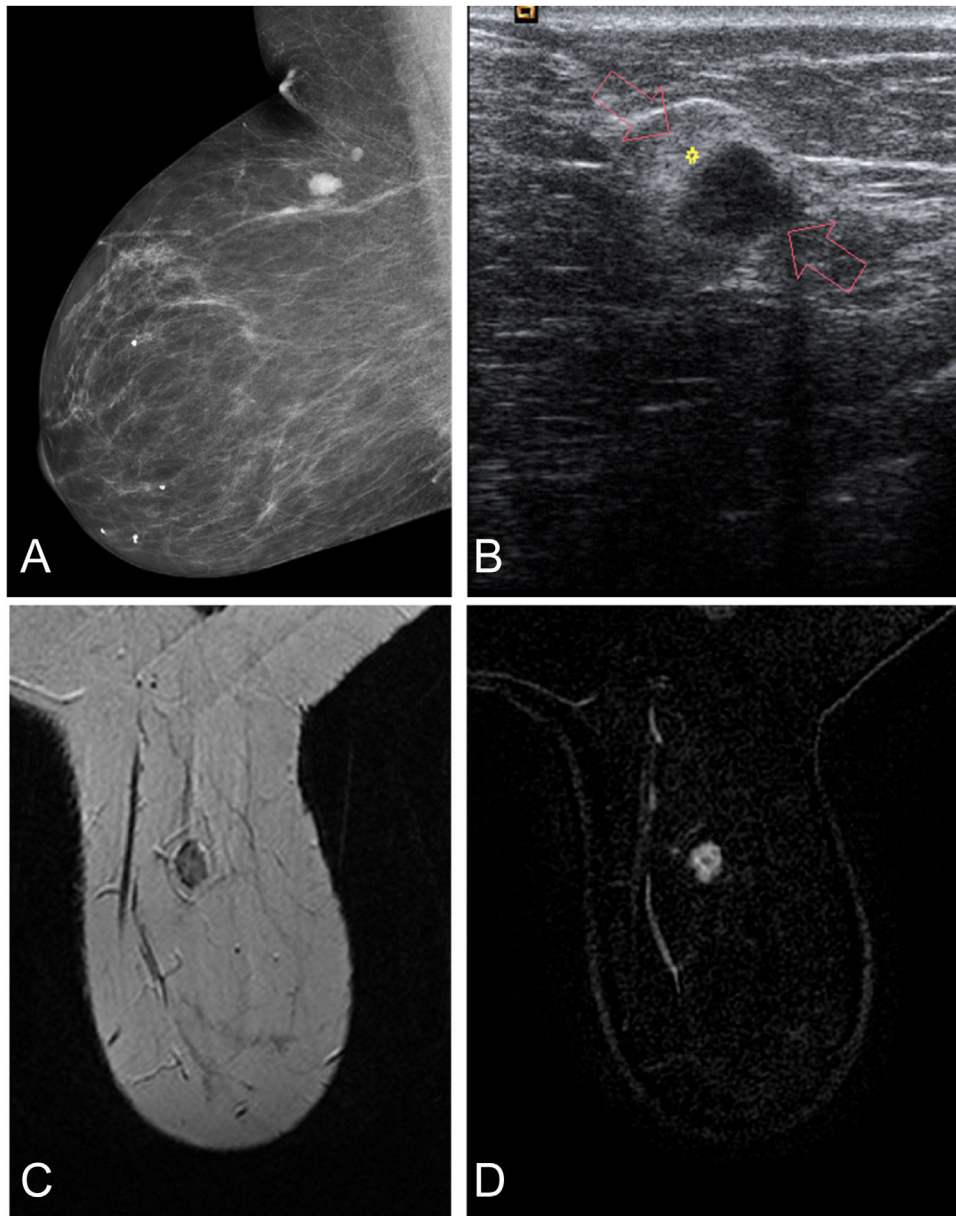


Figure 1 (A) Mammogram (MLO projection) shows a dense, oval mass with microlobulated margins at the junction of the upper quadrants of the right breast. (B) Echography image shows a parallel, oval, heterogeneous nodule with a hypoechoic centre and a peripheral hyperechoic border (*). (C) Axial T2-weighted image shows a well-defined mass with a predominantly hypointense heterogeneous centre. (D) Dynamic T1-weighted axial subtraction image shows heterogeneous enhancement of the lesion.

mixed with bands of hyalinised collagen, frequently surrounded by pseudocapsules.^{3,4} Three cases of MFB of the breast diagnosed in our hospital and their characteristics on diagnostic imaging tests are described below.

Case series

Case 1

A 68-year-old asymptomatic woman attended a routine screening mammogram. She had no family history of breast cancer or other types of malignant neoplasms. The mammogram revealed an oval mass with microlobulated margins in the right breast (Fig. 1A). An echography showed a mass with

circumscribed margins, with a central hypoechogenic area and a peripheral echogenic border (Fig. 1B). Magnetic resonance imaging (MRI) showed a heterogeneous oval mass with early uptake with washout and diffusion restriction (Figs. 1C and D). An echography-guided core needle biopsy revealed spindle cell proliferation consistent with MFB. The patient underwent a lumpectomy and histological findings in the surgical specimen confirmed a MFB.

Case 2

A 62-year-old man came to our hospital with a palpable mass in the right breast. He had no family history of breast cancer, ovarian cancer or other types of neoplasms. The sub-

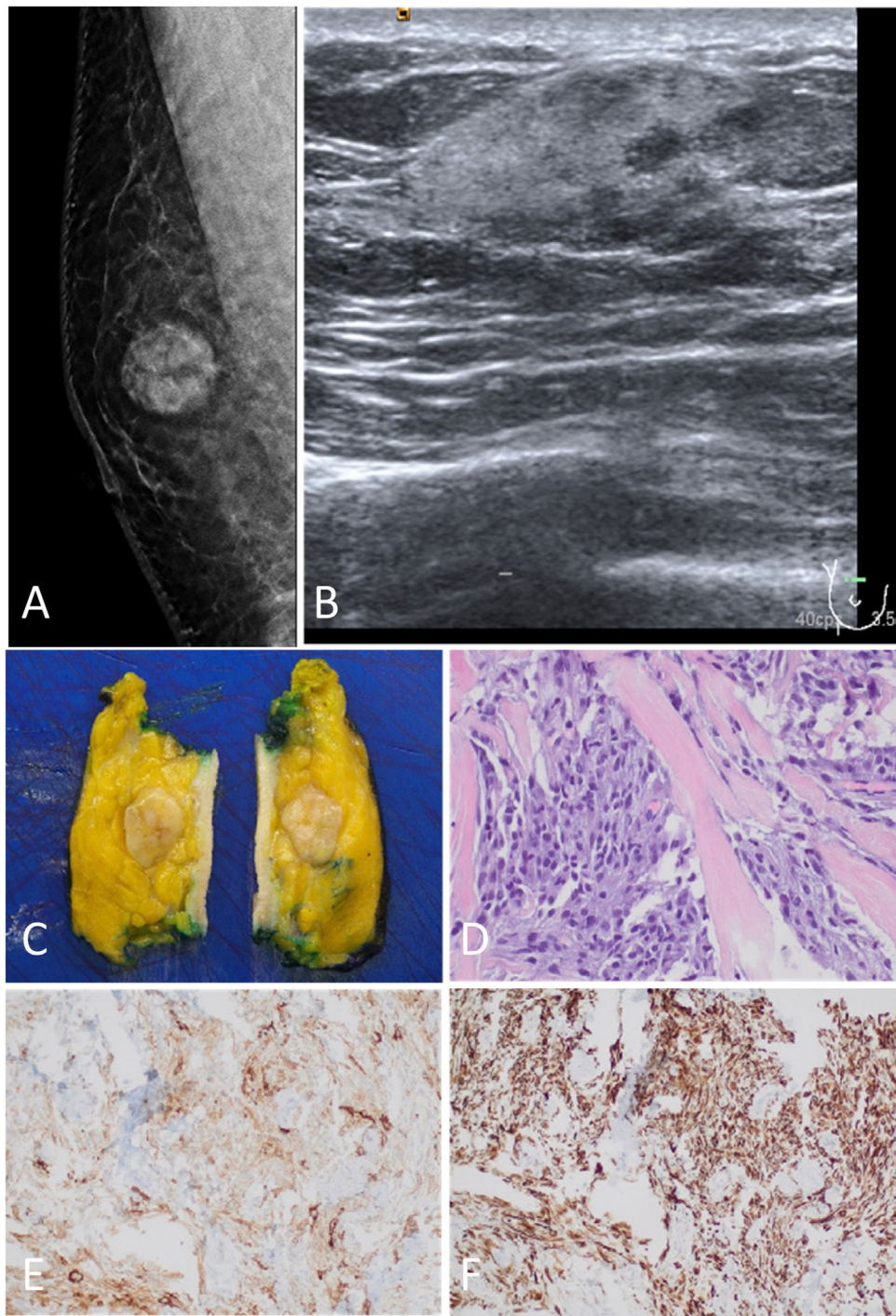


Figure 2 (A) DBT shows a fat-containing 25-mm nodule with circumscribed margins. (B) Ultrasound image shows an oval hyper-echoic heterogeneous nodule. (C) Macroscopically, the mastectomy revealed a firm, rubbery, yellowish, well-defined mass without infiltration into the surrounding tissue. (D) Photomicrograph (original magnification, $\times 40$; H&E stain) shows fascicles of spindle cells interspersed with bands of hyalinised collagen. (E, F) Moderate immunoreactivity for CD10 and desmin $\times 20$.

ject reported a slow-growing, painless mass located in the right retroareolar region that was firm and mobile on clinical examination, without nipple discharge or associated skin changes.

A digital breast tomosynthesis (DBT) was performed, which revealed a 25-mm round mass with circumscribed margins, composed of internal densities of both fatty and

soft tissue, located in the right breast (Fig. 2A). In an echography, the mass was ovoid, parallel and markedly heterogeneous, with predominantly hyperechoic and circumscribed margins (Fig. 2B). Taking these findings into account, it was thought to be a hamartoma. However, due to the increase in size reported by the subject, it was decided to perform an echography-guided core needle biopsy.

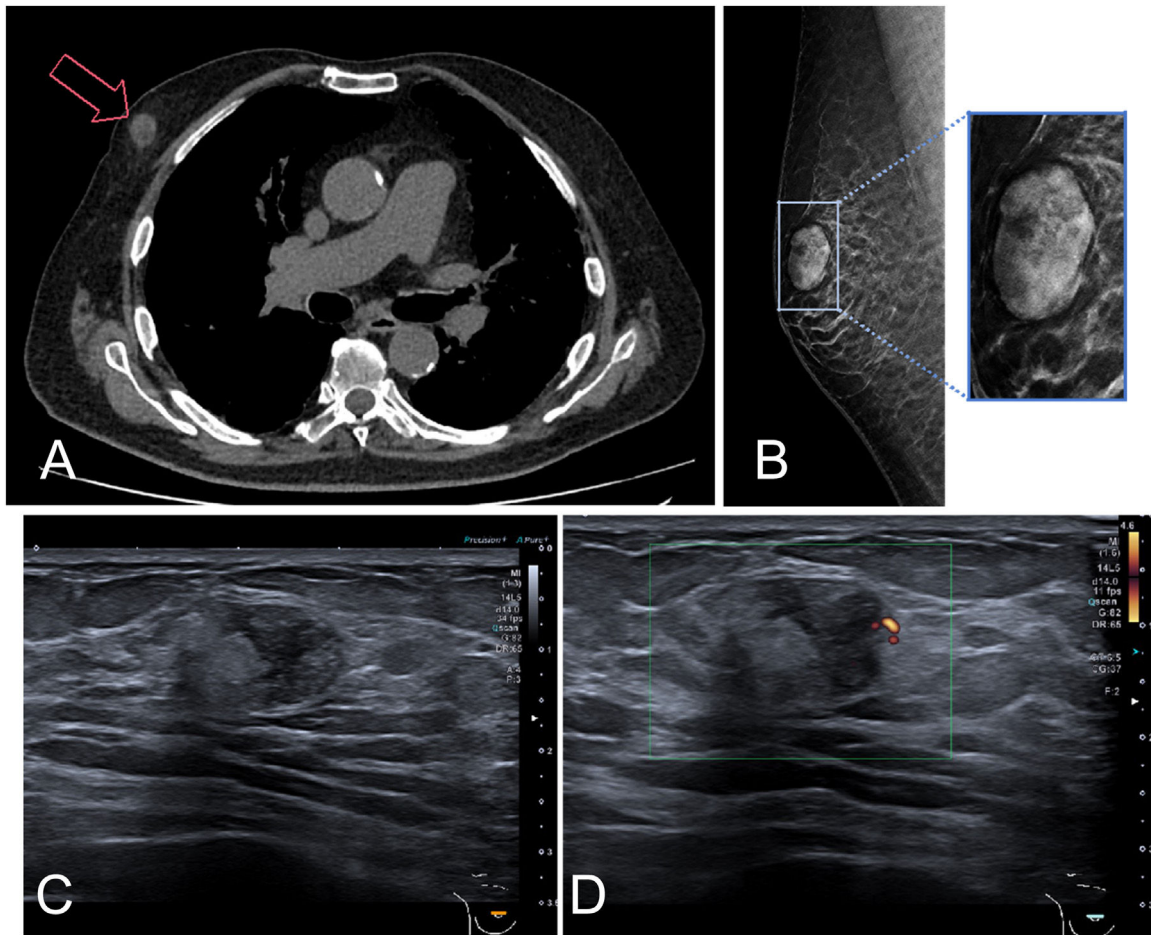


Figure 3 (A) Chest CT shows a 16-mm oval mass in the right breast (arrow). (B) Mediolateral oblique DBT of the right breast shows an oval nodule with circumscribed margins at the 12 o'clock position. (C,D) Echography shows a heterogeneous oval nodule with circumscribed margins with peripheral vascularisation on Doppler evaluation.

Under histological examination, it exhibited cellular proliferation characterised by oval nuclei, arranged in fascicles in different directions surrounded by myxoid stroma that encompassed dense eosinophilic bands of collagen and adipose tissue. There were no signs of necrosis or cytological atypia (Fig. 2D). An immunohistochemical study showed positivity for tumour cells - they were positive for vimentin and smooth muscle actin, CD10 and androgen and oestrogen receptors - and negativity for CK7, EMA, CD34, S-100 protein and STAT-6 (Fig. 2E and F) in neoplastic cells. All these findings pointed to a MFB. The subject underwent a right mastectomy, and histopathological examination confirmed the mass with free resection margins.

Case 3

A 77-year-old man came to our hospital with an oval mass in the right breast, detected during a chest CT (Fig. 3A) that had been performed to evaluate a worsening of his chronic obstructive pulmonary disease. He had no personal or family history of breast cancer. CT showed an oval mass with soft tissue density and well-defined margins in the right breast. Clinically, a firm, mobile mass was palpated in the right breast. A diagnostic DBT revealed a 23 mm oval, solid, fat-

containing circumscribed mass in the right breast at the 12 o'clock position (Fig. 3B). An echography of the right breast revealed an oval, parallel, circumscribed, hypoechoic mass with a pseudocapsule (Fig. 3C and D). No axillary adenopathy was identified. Subsequent echography-guided core needle biopsy revealed histological findings consistent with a MFB. A few months after diagnosis, the subject died due to pancreatic cancer.

Discussion

MFB is classified as a myofibroblastic lesion included within mesenchymal tumours, according to the WHO classification of tumours of the breast.⁵ It was first described in 1981 as a benign spindle cell tumour, spindle cell lipoma, or fibroma, and Wargotz et al. recognised it as a distinct pathological entity in 1987.⁴

MFB was initially described as a tumour affecting the breast of adult males. Since then, multiple cases have been described in postmenopausal women. It is typically characterised by a slow-growing, well-defined, mobile and firm unilateral palpable mass. However, non-palpable lesions that are detected in screening mammograms and by

other imaging techniques (CT and PET-CT) have also been described, as in cases 1 and 3.^{1,3,4}

The onset of MFB is believed to be sporadic. However, several cases have been described in the context of gynecomastia, which points to a possible pathogenic role of steroid hormones, as well as in the context of malignant neoplasms such as pancreatic, kidney, prostatic and urothelial carcinomas.² The subject in case 3 was diagnosed with pancreatic cancer shortly after being diagnosed with MFB.

Our case studies examine the spectrum of diagnostic imaging findings in three patients diagnosed with MFB of the breast at our hospital. Although MFB findings on imaging tests are nonspecific, some common features have been identified. In mammograms and DBT, our subjects were revealed to have a round to oval mass, with circumscribed or microlobulated margins, and high density or with fatty content, which is consistent with the characteristics of MFB previously described in the medical literature. The DBT procedure allows for more precise analysis of shape, margins and internal composition than conventional digital mammography, making it a useful technique for detecting fat in MFBs. No calcifications or architectural distortion were observed, nor have they been previously described.^{1,3,6–11} In echographies, all of our patients and most of the cases described in the medical literature^{1,3,6–12} were revealed to have a heterogeneous oval mass with markedly hyperechoic areas. Although this echography finding has already been mentioned, we would like to highlight its importance when considering MFB diagnosis. On the MRI we describe, the mass showed heterogeneous T2-weighted signal with few internal septa and heterogeneous enhancement on dynamic T1-weighted subtraction images, which was consistent with the findings previously described by Santamaría et al.⁷ MRIs are usually not performed, so there are few cases in the medical literature detailing MRI findings. However, other MR imaging findings described include homogeneous high signal intensity on T2-weighted images and homogeneous or heterogeneous enhancement on dynamic T1-weighted images.^{6,7}

Macroscopically, the cut surface reveals a well-defined, round, pale pink or reddish-brown mass with a smooth or lobed surface. No cystic degeneration, necrosis or haemorrhage were observed. Microscopically, a proliferation of spindle cells with rounded margins (myofibroblasts) is observed, arranged in short fascicles that randomly intersect with hyalinised collagen bundles. A minority of these lesions contain variable amounts of adipose tissue. These tumours are well circumscribed but do not have a true capsule. However, they do have a pseudocapsule formed by the surrounding compressed breast tissue. There are no breast ducts or lobules. In the immunohistochemical analysis, MFBs stain positively for vimentin, desmin and CD34, and often for alpha-SMA (smooth muscle actin), bcl-2 and CD10, while they stain negatively for cytokeratin, EMA, S-100, HMB-45 and c-Kit (CD117). These results indicate that myofibroblasts are cells of myogenic origin, not neurogenic. Their size varies between 0.9 and 4 cm, but the average diameter is about 2 cm. Larger tumours and some cases of giant masses (15 cm) are also reported in the literature.^{1,6,8–10} Various histological variants of MFB have been identified in addition to the classic type: lipomatous, epithelioid, decid-

uoid, palisaded/schwannian-like and MFB with extensive myxoedematous stromal changes.¹¹

The nonspecific findings of MFB on diagnostic imaging tests could be attributed to these different histological variants. However, as mentioned above, MFB usually presents as a heterogeneous lesion on mammogram, echography and MRI. As indicated by Santamaría et al. and Porter et al.,¹³ the heterogeneity of the lesion may be due to the multiple tissues present in the tumour; high density, hypoechogenicity and low signal could be related to hyalinised collagen, while low density, hyperechogenicity and high signal, to fat and loose myxoid stroma.

The differential diagnosis of MFB should consider heterogeneous and hyperechoic lesions, including benign neoplasms and conditions such as chronic haematoma and fat necrosis, hamartoma, phyllodes, fibromatosis, lipoma, pseudoangiomatous stromal hyperplasia, angiolipoma and haemangioma. Malignant lesions can also present as hyperechoic masses and, although very rare, should also be taken into account, such as invasive lobular carcinoma, breast lymphoma, angiosarcoma and liposarcoma.¹⁴ Clinical correlation is essential to establishing the correct diagnosis and a biopsy should always be performed for histological confirmation.

Lumpectomy is the treatment of choice. Mastectomy may also be necessary in large tumours or in men with coexisting gynecomastia. Wide local excision is considered curative, as less than 1.5% local recurrence has been reported after 20 years of follow-up. No distant metastases have been reported in the medical literature.

In conclusion, MFB should be considered when a circumscribed heterogeneous mass with fatty content is seen on a mammogram or DBT, with a heterogeneous hyperechoic appearance on echography and heterogeneous on T2-weighted MRI. Above all, it should be taken into consideration whenever it reveals to be a slowly-growing breast mass in men or postmenopausal women. A biopsy is necessary to confirm the diagnosis, and the treatment of choice is surgical removal.

Authorship

- 1 Responsible for the integrity of the study: CS, XB, BU, SG, ES.
- 2 Study concept: CS, XB.
- 3 Study design: CS, XB.
- 4 Data collection: CS.
- 5 Data analysis and interpretation: none.
- 6 Statistical processing: N/A.
- 7 Literature search: CS.
- 8 Drafting of the article: CS, ES.
- 9 Critical review of the article with intellectually significant contributions: CS, XB, BU, SG, MM, ES.
- 10 Approval of the final version: CS, ES.

Conflicts of interest

There are no conflicts of interest pertaining to the published article.

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