



**Ministério da Saúde
Instituto Nacional de Câncer
Coordenação de Ensino
Aperfeiçoamento nos Moldes Fellow em Radiologia Mamária**

ANA LÉA BURNIER GANIMI COSTA

**DERMATOFIBROSARCOMA PROTUBERANS SIMULATING
PRIMARY BREAST TUMOR**

Rio de Janeiro

2024

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Trabalho de Conclusão de Curso
apresentado ao Instituto Nacional de
Câncer como requisito parcial para a
conclusão do curso de Aperfeiçoamento
nos moldes Fellow em Radiologia
Mamária.

Orientadora: Dra. Marcele França Barreto Côrtes

Revisora: Profª Dra. Shirley Burburan

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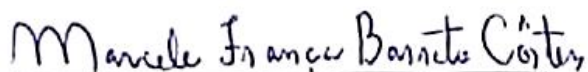
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Dermatofibrosarcoma protuberans simulating primary breast tumor

Trabalho de Conclusão de Curso apresentado ao Instituto Nacional de Câncer como requisito parcial para a conclusão do curso de Aperfeiçoamento nos moldes Fellow em Radiologia Mamária.

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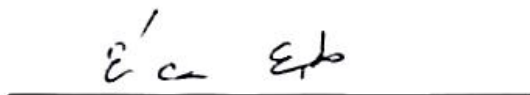
Banca examinadora:



Dra. Marcelle França Barreto Côrtes



Dra. Paula Medina Gomes Maciel Curi Bonotto



Dra. Érica Endo

Rio de Janeiro

2024

*Dedico este trabalho à minha filha
Helena, minha maior motivação, aos
meus pais, Léa e Sebastião, que tanto me
apoiaram durante este ano, e ao meu
marido Bruno, pelo incentivo.*

*Somos aquilo que repetidamente fazemos, portanto, a excelência não é um
feito, mas um hábito.*

Aristóteles

RESUMO

COSTA, Ana Léa Burnier Ganimi. **Dermatofibrossarcoma protuberans simulando tumor primário de mama.** Trabalho de Conclusão de Curso (Aperfeiçoamento nos Moldes Fellow em Radiologia Mamária) — Instituto Nacional de Câncer (INCA), Rio de Janeiro, 2024.

Dermatofibrossarcoma protuberans é uma neoplasia mesenquimal rara, de crescimento lento, da derme e tecidos subcutâneos e tende a ser localmente agressiva. É caracterizada como uma neoplasia de grau baixo a intermediário, com alta taxa de recorrência, mas raramente cursa com metástases. Comumente envolve o tronco e as extremidades, e muito raramente a pele da mama, mimetizando um tumor primário de mama com poucos casos relatados na literatura. Relatamos uma paciente de 18 anos que foi encaminhada ao nosso serviço, com uma massa palpável crescendo na mama esquerda. A mamografia revelou um nódulo isodenso, irregular e indistinto, na união dos quadrantes inferiores na mama esquerda. Na ultrassonografia, a lesão se apresenta como um nódulo complexo sólido cístico, com vascularização periférica ao Doppler, sendo categorizado como ACR BI-RADS 4. A paciente foi submetida à biópsia da lesão na mama esquerda e o diagnóstico histopatológico foi dermatofibrossarcoma protuberans. Uma excisão local em um segundo momento confirmou o diagnóstico histopatológico.

Palavras-chave: dermatofibrossarcoma; neoplasia mamária; tumores da mama; neoplasia cutânea; tumores cutâneos.

ABSTRACT

COSTA, Ana Léa Burnier Ganimi. **Dermatofibrossarcoma protuberans simulating primary breast tumor.** Final paper (Fellowship in Breast Radiology) — Brazilian National Cancer Institute (INCA), Rio de Janeiro, 2024.

Dermatofibrosarcoma protuberans is a rare, slow-growing mesenchymal neoplasm of the dermis and subcutaneous tissues and tends to be locally aggressive. It is characterized as low to intermediate grade neoplasm with high recurrence rate but rarely metastasises. Commonly involves the trunk and extremities, and very rarely the breast skin, mimicking a primary breast tumor with few reported cases in the literature. We report an 18-year old female patient who was referred to our service, with a palpable mass growing in left breast. Assessment of the breasts with mammography revealed an irregular, indistinct, equal density mass in union of the lower quadrants in left breast. On ultrasound, the lesion presents as oval, indistinct, complex cystic and solid mass, with vessels in rim. It was categorized as breast imaging reporting and data system (BIRADS) 4. The patient underwent core biopsy of the lesion in left breast and the histopathological diagnosis was dermatofibrosarcoma protuberans. A local excision in a second time, confirmed the histopathological diagnosis.

Keywords: dermatofibrosarcoma; breast neoplasm; breast tumors; skin neoplasms; skin tumors.



DERMATOFIBROSARCOMA PROTUBERANS SIMULATING PRIMARY BREAST TUMOR

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INTRODUCTION

- ❑ Dermatofibrosarcoma protuberans (DFSP) is a rare, slow-growing mesenchymal neoplasm of the dermis and subcutaneous tissues and tends to be locally aggressive.
- ❑ DFSP is characterized as low to intermediate grade neoplasm with high recurrence rate but rarely metastasises.

INTRODUCTION

- ▣ DFSP commonly involves the trunk and extremities, and very rarely the breast skin, mimicking a primary breast tumor with few reported cases in the literature.
- ▣ DFSP of the breast presents with a slow growing pattern, initially in the dermis and then invading the subcutaneous tissue.

DESCRIPTION

- ▣ We report an 18-year old female patient who was referred to our service, with a palpable mass growing in left breast.
- ▣ Assessment of the breasts with mammography revealed an irregular, indistinct, equal density mass in union of lower quadrants in left breast.
- ▣ On ultrasound, the lesion presents as oval, indistinct, complex cystic and solid mass, with vessels in rim. It was categorised as breast imaging reporting and data system (BIRADS) 4.

MAMMOGRAPHY

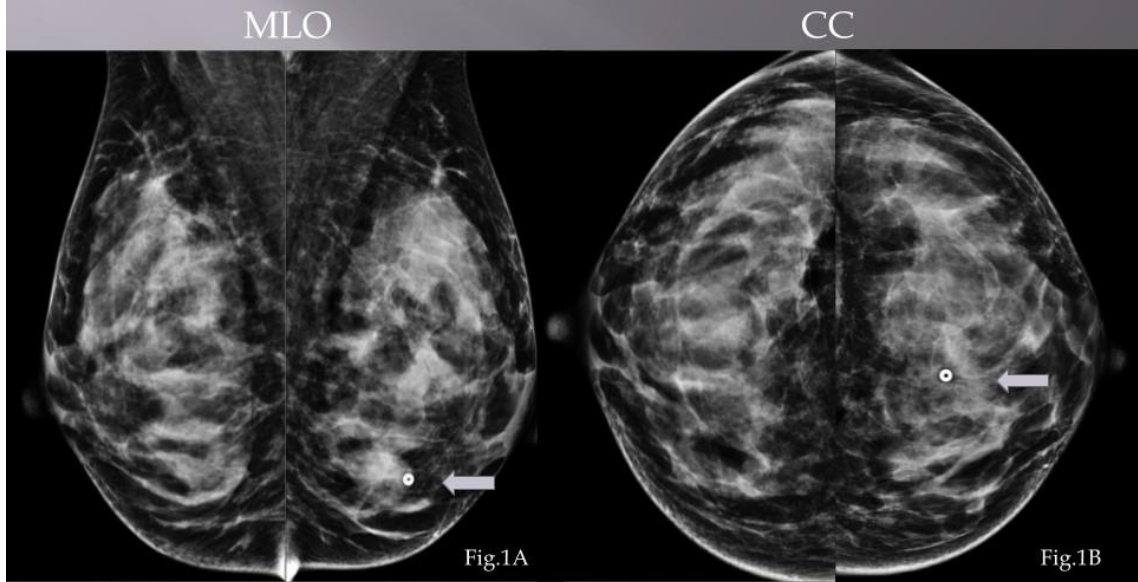


Fig.1A and 1B: Mediolateral oblique and craniocaudal mammogram respectively, shows an irregular, indistinct, equal density mass in the union of lower quadrants in left breast, marked by metallic BB marker on skin (arrow).

ULTRASOUND

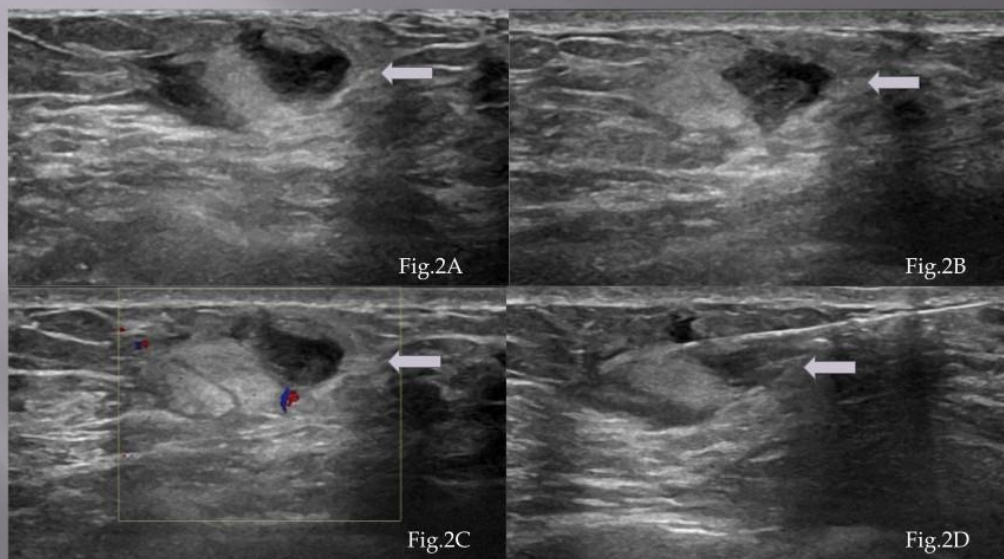


Fig. 2A and 2B: Ultrasound shows an oval, indistinct, complex cystic and solid mass (arrow).
Fig. 2C: Colour Doppler shows vessels in rim of the lesion (arrow).
Fig. 2D: Core biopsy of the lesion (arrow).

DESCRIPTION

- The patient underwent core biopsy of the lesion in left breast and the histopathological diagnosis was dermatofibrosarcoma protuberans.
- A local excision in a second time, confirmed the histopathological diagnosis.

DISCUSSION

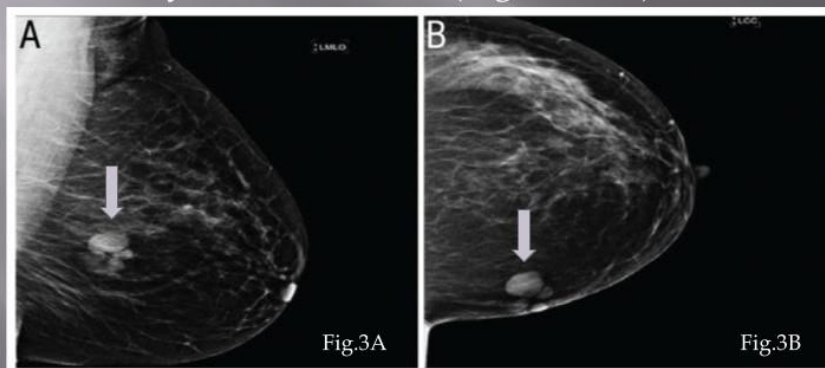
- DFSP is a rare sarcoma of the dermis and subcutaneous tissue that has an estimated incidence of 0.8-5 cases per one million per year and accounts for less than 6% of all sarcomas.
- It commonly occurs on the trunk (50-60%), followed by the limbs (25%) and the head and neck area (10-15%). Involvement of the breast by DFSP is extremely rare, with only a few cases reported in the literature

DISCUSSION

- It usually affects patients aged between 20–50 years.
- DFSP has a high tendency of local recurrence and a low propensity for metastasis.
- Although rare, distant metastasis, especially in the lungs and lymph nodes, has been reported.

DISCUSSION

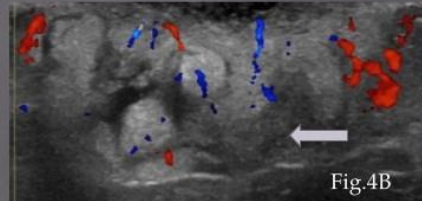
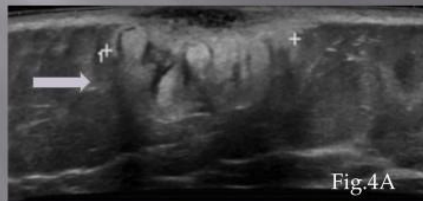
- Mammography: breast DFSP appears as a skin-based or intra-mammary located well-circumscribed oval-shaped lesion with no microcalcification nor speculation. These findings are not specific and hence, breast DFSP can mimic a benign breast lesion such as fibroadenoma, sebaceous cyst or an abscess. (Fig.3A e 3B)



Al-Farsi, Abeer et al., Dermatofibrosarcoma Protuberans Mimicking Primary Breast Neoplasm: A case report and literature review. (Accessed on 06 Jul 2023) DOI: 10.18295/squmj.2020.20.03.019

DISCUSSION

- Ultrasonographically: breast DFSP appears as a heterogeneously hyperechoic solid mass centered in subcutaneous tissue, with overlying hypoechoic region in the dermis with mild lobulation and increased vascularity on colour Doppler. Increased vascularity can be used to differentiate DFSP from benign breast lesions that usually appear as well-circumscribed lesions such as sebaceous cysts and liver abscesses. (Fig.4A e 4B)



Gilcrease-Garcia B, Dermatofibrosarcoma protuberans. Case study, Radiopaedia.org (Accessed on 06 Jul 2023) <https://doi.org/10.53347/rID-64660>

DISCUSSION

- Breast skin DFSP is a rare entity that can mimic a primary breast tumour.
- Awareness of this rare lesion and its typical imaging features will help to avoid the misdiagnosis of DFSP with a benign breast lesion and suggest a pre-operative diagnosis.

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THANK YOU



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Declaramos que

*ANA LEA BURNIER GANIMI COSTA; CAROLINA FERNANDES RAMALHO;
GIOVANA ORTIZ D'ÁVILA; MARCELE FRANÇA BARRETO CÔRTEZ; THAIS
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