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Review Paper

Dermatomyositis Revealing Mucinous Breast Cancer: Report of a Clinical Case and Review of the Literature

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ABSTRACT:
Dermatomyositis is a rare inflammatory disease caused by skin and muscle damage. It may be associated with underlying neoplasia. the appearance of the cancer is before, simultaneously, or after the diagnosis of dermatopolymyositis. We report the case of a 58-year-old patient with dermatomyositis revealing mucinous breast cancer below with review of the literature.

Keywords: Mucinous cancer, breast, dermatomyositis, mammography, ultrasound, biopsy.

INTRODUCTION:

Dermatomyositis is a rare inflammatory disease, mainly affecting the skeletal muscles and skin. It can be associated with autoimmune diseases, infections, or neoplasia. It is associated in 18-32% of cases with an underlying neoplasia appearing before, simultaneously, or after the diagnosis of cancer. Breast cancer is the most common cancer found in women with dermatomyositis, with an incidence of 11 to 29%. Mucinous breast cancer is a rare form of breast cancer, accounting for 1-4% of all cases. It is characterized by the presence of mucus in tumor cells, which can be pure or mixed, depending on whether or not it is associated with an infiltrating carcinomatous component. Pure mucinous cancer has a favorable prognosis, while mixed mucinous cancer is more aggressive and metastatic.

Observation:

58-year-old woman, diabetic and hypertensive under medication, with no personal or family history of breast cancer in her family. She consulted the dermatology department following the appearance of erythematous, papular lesions affecting the entire body, as well as ulcerations on the right arm and the right breast. The patient reported the appearance of arthralgia, myalgia and a motor deficit in the right upper limb. The clinical examination revealed purplish erythema and telangiectasias on the shoulders, on the extended surface of the upper limbs and on the breast with localized skin necrosis. The neurological examination revealed a muscular deficit predominantly in the right upper limb. The biological examinations show an elevation of muscle enzymes (CPK and LDH), the patient underwent a mammogram which found an overall asymmetry of density at the expense of the right breast and also the presence of extensive bilateral vascular calcifications as well as diffuse rod-shaped calcifications, fine and regular bilateral retro-areolar site (Figure 1). Breast ultrasound revealed a lobulated mass, with circumscribed contours, hypoechoic, homogeneous, located retro-areolar measuring 17mm, without axillary lymphadenopathy (Figure 2). The radiological file was classified BI-RADS 3 by the ACR.

Breast skin biopsy was negative (absence of lesions suggestive of Paget's disease or granulomatous mastitis). The diagnosis of dermatomyositis was made following a muscle biopsy. the patient was put on treatment with corticosteroids.

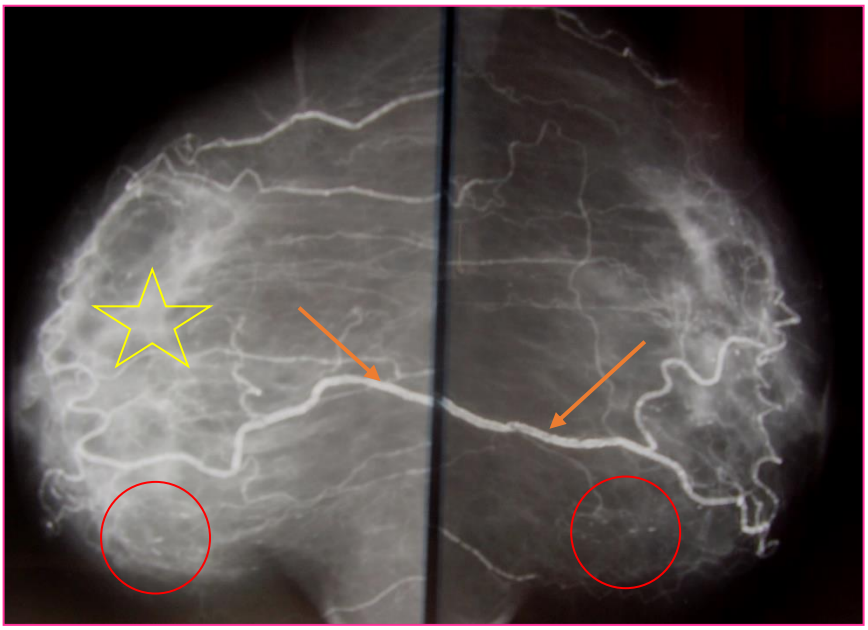

Figure 1. Bilateral mammography, frontal views. Overall density asymmetry at the expense of the right breast (star), bilateral vascular calcifications (arrow), stick-shaped calcifications (circle).

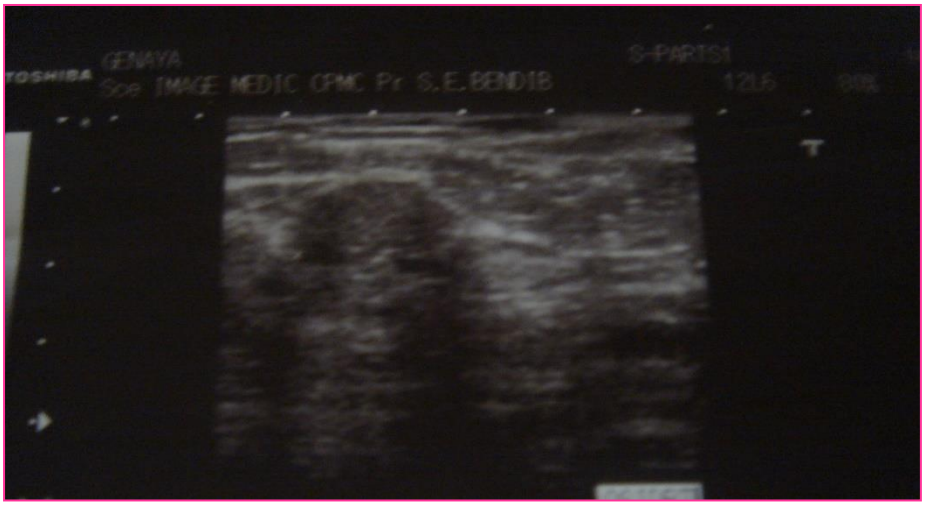

Figure 2. Breast ultrasound. Mass with a lobulated shape, with circumscribed contours, hypoechoic, homogeneous.

After 10 months of treatment, the patient consulted again for the appearance of a 4cm right breast mass and breast induration (figure 2).

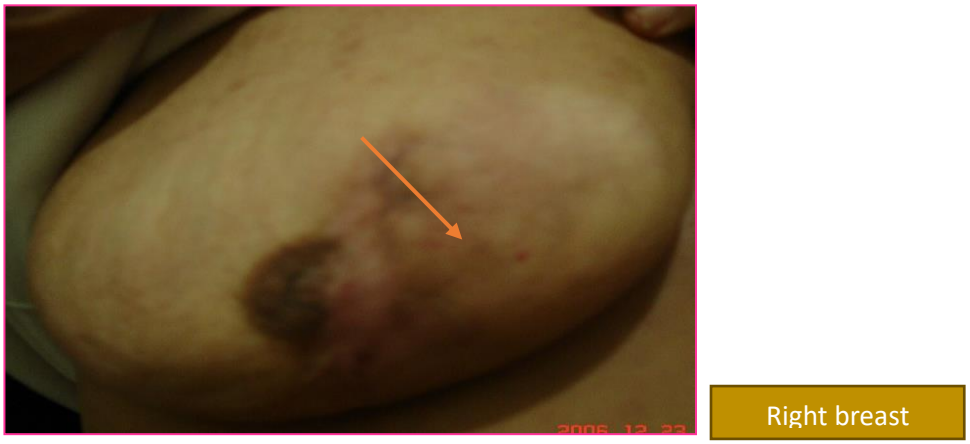

Figure 3. Breast arch associated with ulcerations and skin erythema.

Another mammographic and ultrasound assessment carried out again, revealed on mammography, a right retroarolar overdensity, an accentuation of vascular and rod calcifications.

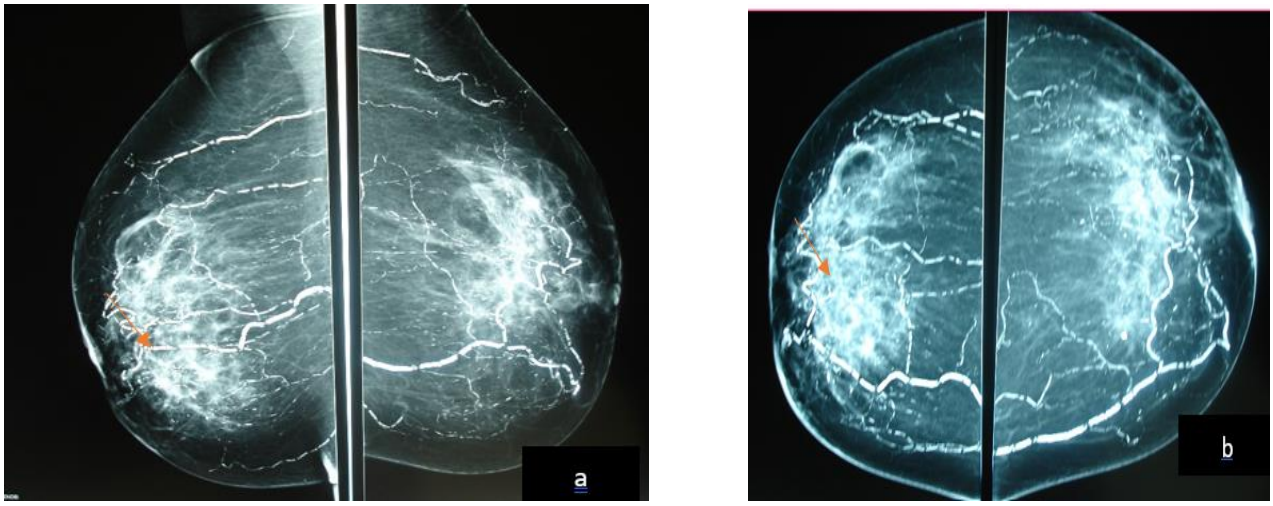

Figure 4. Right retroarolar overdensity, accentuation of linear, rod-shaped calcifications.

The ultrasound revealed an increase in the size of the mass measuring 58mm, retroarolar location, lobulated shape, with microlobulated contours in places and irregular in other places, hypoechoic echostructure, heterogeneous by the presence of echogenic spots (figure 3). The lesion was classified BI.RADS 4 by the ACR. The microbiopsy of the lesion came back in favor of a mucinous carcinoma expressing hormonal receptors (ER 70%, RP 60%) and negative for HER2. aassociated with a small classic ductal component of grade I of SBR (without vascular emboli). The extension assessment carried out by a thoracoabdominal CT scan did not find any distant metastases the patient.

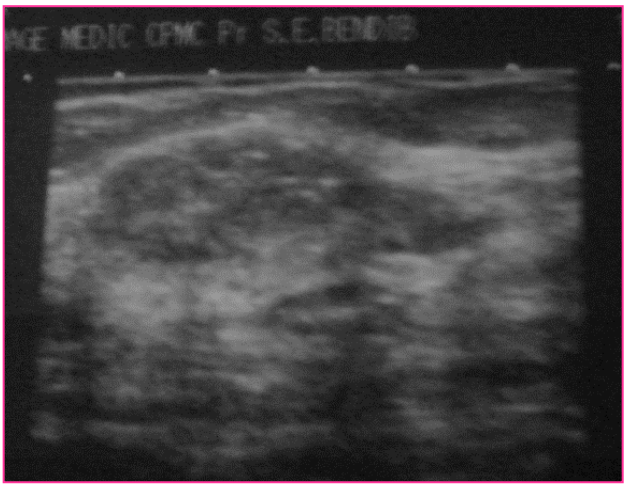

Figure 5. Breast ultrasound: Hypoechoic, heterogeneous, poorly circumscribed mass measuring 38 × 36 mm with posterior attenuation in places.

DISCUSSION:

Dermatomyositis is a disease mainly affecting the skeletal muscles and the skin. It is a rare inflammatory disease. It can be associated with autoimmune diseases or neoplasia (1). The risk of cancer in patients with dermatomyositis is estimated at 15 to 32%, which is 3 to 5 times higher than in the general population (2). Breast cancer is the most common cancer found in women with dermatomyositis, with an incidence of 11 to 29% (3,4). The average time between the diagnosis of dermatomyositis and that of breast cancer is 6 to 12 months, but it can vary from a few weeks to several years (5,6).

Mucinous breast cancer is a rare histological entity, which is distinguished by the production of mucus by tumor cells. There are two subtypes (7): pure mucinous cancer (8), and mixed mucinous cancer mixed with a carcinomatous component (9).

Mucinous cancer represents approximately 1 to 6% of breast cancers and preferentially affects older women (10). It most often manifests itself as a palpable, painless, mobile mass with regular contours. It presents as a palpable, often painful, fixed mass with irregular contours. It may be associated with inflammatory signs, nipple discharge, or skin ulceration.

In imaging, the mammographic appearance in mixed forms is generally that of a lobulated mass with irregular contours, sometimes spiculated if there is an infiltrating ductal association (11,12). Pure mucoid forms have a lobulated morphology with circumscribed contours, calcifications are absent except in mixed types and this is the case in our case. Ultrasound reveals a hypoechoic mass, well limited, without vascularization, and without lymphadenopathy in pure colloid carcinomas and in the mixed type it takes the appearance of a hypoechoic, heterogeneous mass, of irregular shape with posterior acoustic attenuation, in relation with the infiltrative nature of the tumor (13). In elderly women, magnetic resonance imaging is of great interest in distinguishing a pure colloid carcinoma from a fibroadenoma [14]. The definitive diagnosis is based on biopsy, which makes it possible to differentiate the two subtypes and specify the histological grade, the status of hormonal receptors, HER2 and Ki-67. Pure mucinous cancer is usually grade 1 or 2, hormone dependent, negative for HER2 and Ki-67. Mixed mucinous cancer is more often grade 2 or 3 hormone resistant, positive for HER2 and Ki-67.

The treatment of mucinous breast cancer is based on surgery [15], which can be conservative or radical, depending on the size and number of lesions, radiotherapy [16] and chemotherapy.

CONCLUSION:

The association of dermatomyositis cancer remains rare, and sometimes revealing. Its diagnosis is based on clinical, radiological, biological and histological data. Its treatment is based on corticosteroid therapy and treatment of the underlying cancer.

Conflicts of Interest: The authors declare that they have no conflicts of interest.

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