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Breast location of Darier Ferrand dermatofibrosarcoma

Sara Yacoubi Khebiza , Kenza Benchaaboune , Yassine Belhaj∗, ∗, Sofia Jayi∗, Fatima Zahra Fdili Alaoui∗ , Hekmat Chaara and MyAbdeillah Melhouf

∗ Department of Obstetrics and Gynecology II CHU Hassan II of Fez.

Abstract : *Dermatofibrosarcoma of Darrier and Ferrand or dermatofibrosarcoma protuberans (DFSP) is a rare tumor of the skin and soft tissues usually appearing on the trunk and extremities. The occurrence of DFSP in the breast is extremely rare. This is a slow-growing lesion that can go unnoticed – and affects more frequently women aged 20 to 40 years. It is classified as a low-grade malignant tumor but can be locally aggressive with a high rate of recidivism; especially if the resection margins are not healthy. We report here the case of a young 30-year-old patient, followed for a fibrocystic dystrophy, which caused a skin lesion to appear on the breast, including the diagnosis after histology came back in favor of dermatofibrosarcoma protuberans. Through this case, we reviewed the literature in order to discuss results of ultrasound, magnetic* ***resonance****, histology and surgical treatment of dermato fibrosarcoma of the breast.*

Keywords: Dermato fibrosarcoma protuberans, breast, MRI, ultrasound, histology

1- INTRODUCTION

Dermato fibrosarcoma protuberans (DFSP) is a rare tumor of the skin and subcutaneous tissue [1]. This tumor involves commonly the trunk, extremities and neck [1]. This is a slow-growing lesion that can go unnoticed, and affects more frequently women aged 20 to 40 years [2].

It is classified as a low-grade malignancy but can be locally aggressive with a high rate of recidivism; especially if the resection margins are not healthy[3] The breast is a rare location of DFSP [2].

The radiological findings of this lesion are often misleading, suggesting benignity. This is a case of a woman of 30 years old with a 5-year history of a skin lesion on the left breast which turned out to be DFSP on the pathological result.

In this article, we report a case of DFS breast cancer and review the literature.

2- CLINICAL CASE

This is a 30-year-old female patient, presenting with ATCD of centrifugal annular erythema on clinical and histological grounds, followed for 5 years for cystic dystrophy of both breasts based on an old breast ultrasound associated with a lesion skin on the left breast On clinical examination: indurated sclerotic pigmented plaques in number of 3, slowly growing not sensitive above the nipple opposite the junction of the upper quadrants.

On breast ultrasound: fibro dystrophy cysts of both breasts with the presence next to the skin lesions of the right breast, small lesional skin formations an echogenic ones that do not take color Doppler encoding, of millimeter size.

On breast MRI: Enhancement without a mass at the level of the QSI of the right breast near the nipple, The lesion showed significant enhancement measuring post-contrast.

The patient also underwent a skin biopsy in the upper inner quadrant of the left breast. The histological examination was performed.

reveals .es mitoses are uncommon spindle cell agencies in sheets and flows with diffuse immuno-reactivity and intense CD34 and anti P53 heterogeneous nuclear expression were negative for markers anti actin smooth muscle, anti PS100 and anti CD163 which is in favor of a dermato fibrosarcoma of Darier Ferrand.

The patient underwent a mastectomy with latissimus dorsi flap A follow-up of 6 months after surgery was without recurrence.

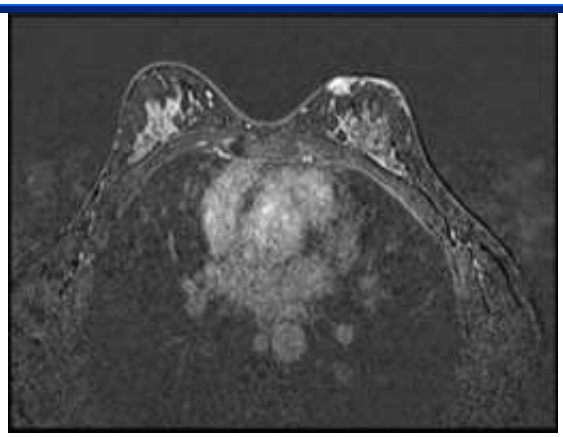

Figure 1 : Breast MRI showing in sequence injected a lesion which enhances after

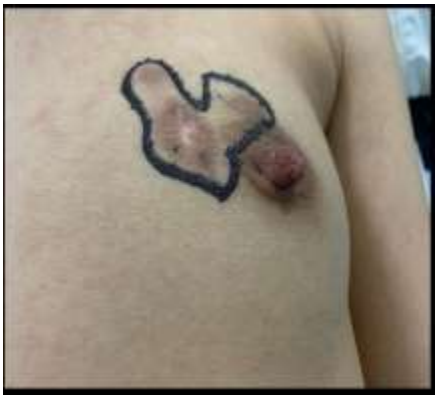

Figure2: pigmented lesion of the upper inner quadrant of the right breast above the nipple

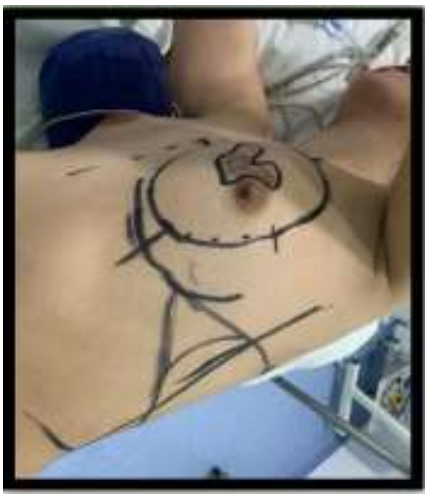

Figure3: wide excision limits

Figure4: Immediate post-operative mastectomy with major flaps dorsal with installation of 2 drains of the recipient site and the donor site

3- DISCUSSION

Due to the rarity of dermato fibrosarcoma of the breast, knowledge has been limited almost exclusively to case reports.

DFSP is a rare mesenchymal tumor whose incidence is estimated at 0.5 cases per million per year. It was first described once by Darier and Ferrand in 1924, but the term was then coined by Hoffmann in 1925 [4].

The preferred site for the DFS is the trunk 50 to 60. The breast is a rare location for mesenchymal tumors and even rarer for DFSP[3], which presents generally as an indolent skin lesion with progressive growth which may or may not be associated with a breast mass [3]

It rarely presents only as an intramammary lesion [6]. DFS is generally ignored by patients in due to its slow growth [3]. The skin lesion is most often slightly hyperpigmented, reddish to bluish in color and nodular in appearance [7]. Young pre-menopausal women are frequently affected.

The lesion in our case was brownish, sclerosing and hard, in the form of plaques without nodules.

Typical ultrasound findings reported from DFS are a hypoechoic or heterogeneous oval lesion, parallel to the skin, taking Doppler encoding[3]. In our case, the ultrasound showed small rounded lesions not dopplerized.

MRI is used for clear demarcation of the involved region, an accurate assessment of the size and to measure its distance to the pectoral muscle.

Most cases have been reported to be iso to hypointense on T1 and T2 weighted images with intense contrast enhancement and Washout curve.[3, 7, 8] . DFS can rarely metastasize, however, local recurrence can occur in up to 60 years.

Surgery is the standard treatment. The treatment is based on a wide surgical excision up to the aponeurosis, with side margins of 3 cm. Margins could be reduced if the Mohs technique is used. Given the high risk of recurrence, rigorous clinical monitoring every 6 months is necessary with re-biopsy of suspicious regions.

At our patient, we did not detect any local recurrence after 15 months.

4- Conclusion:

In summary, SSM of the breast can mimic a primary malignant tumor of the breast. Clinical diagnosis alone is difficult, especially

If there is no clinically visible skin component. The results of ultrasound and MRI are useful additions. It is a differential diagnosis of broad-based subcutaneous lesions with high vascularity.

It is a cancer with a good prognosis, because despite local recurrences, the Vital prognosis is only exceptionally poor.

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