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DERMATOFIBROSARCOMA PROTRUBERANS OF THE BREAST SKIN SIMULATING MAMMARY CARCINOMA: A CASE REPORT

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ABSTRACT

Dermatofibrosarcoma protuberans (**DFSP**) is an uncommon slow growing neoplasm of the dermis with tendency to invade the subcutaneous tissues. Usually, it behaves as an intermediate grade malignancy but in rare instances, it can metastasize. It presents in the middle age of life and frequent occurrences have been observed in trunk and extremities but involvement of the breast has rarely been reported. Pre-operative diagnosis with mammography, ultrasonography and fine-needle aspiration cytology is challenging. Histomorphology and immuno- histochemistry are helpful in making a definitive diagnosis.

We report a case of a DFSP of the left breast in a middle aged lady. The diagnosis was posed by histological and immuno- histochemistry studies of a biopsy of the breast skin lesion (colored plaque); Tumor cells show strong reactivity for CD34 antibody. The patient is programmed for a wide local excision.

Key words: CD34 antibody; Dermatofibrosarcoma protuberans; Neoplasm of the dermis; Wide excision

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INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) of the breast is a rare neoplasm that can remain indolent and stable for years; however, it can be accompanied by rapid growth and fibrosarcomatous changes [1].It is known to be locally aggressive, though metastases are rare and when occur affect the lungs and the lymph nodes. It usually presents in the middle age between the fourth to sixth decade but can occur at all ages[1-2].It presents as an indurated plaque on which multiple reddish purple, firm nodules arise, sometimes in association with ulceration. It usually appears on trunk and extremities; however, involvement of the breast has been reported in rare instances [3].Differentiating pre-operatively from other benign and malignant lesions of the breast by routine investigation may not be possible. Computerised tomography (CT), magnetic resonance imaging (MRI) and magnetic resonance spectroscopy (MRS) have also been reported as an investigation that would aid in the diagnosis of DFSP. Histological examination and immunochemical stains are required to confirm the diagnosis [1].

CASE REPORT

Patient of 46 years old, with no previous medical or surgical history, 7th gesity, 5th pare with two miscarriages, menarche at 14 years, regular cycles , oral contraception for 5 years, she always has her menstruation; she was referred to the Breast Unit at the National Institute of oncology of Morocco for an infected ulcerative necrotic cutaneous lesion at the level of the superior-external quadrant of the left breast ; measuring 4 cm of major axis; which present since 3 months; the examination revealed a firm, plaque-like lesion of the skin surrounded by blue discoloration with absence of palpable breast nodule and absence of adenopathies in the two axillary regions (figure 1); Examination of the right breast was normal; The patient benefited from a mammogram that has not shown any abnormalities (figure 2); A breast ultrasound complement reveals the presence of a small subcutaneous cystic

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formation of the left SEQ measuring 32 mm of major axis with thickened wall taking color during color doppler (**Figure 3**); a biopsy of cutaneous lesion was performed; Histological examination showed a typical aspect of Dermatofibrosarcoma of Darrier and Ferrand with fusocellular tumor proliferation made of bundles of fibroblasts arranged according to the aspect of radiuses of wheel (**Figure 4a**: morphological aspect (Hematoxylin and Eosin stain (H&E stain) x10)); The tumor cells have elongated nuclei without frank atypia (**Figure 4b**: morphological aspect (H&E stain x40)); Immuno- histochemical (IHC) analysis of tumor cells showed strong immune reactivity to CD34 (**Figure 4c**). The patient is programmed for a wide local excision.

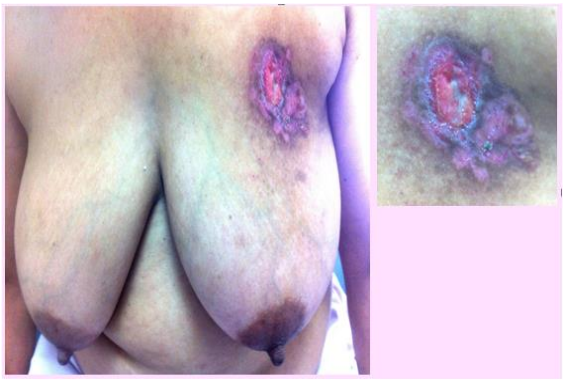

Figure 1: Infected ulcerative necrotic lesion of the skin at the superior-external quadrant of the left breast surrounded by blue discoloration .

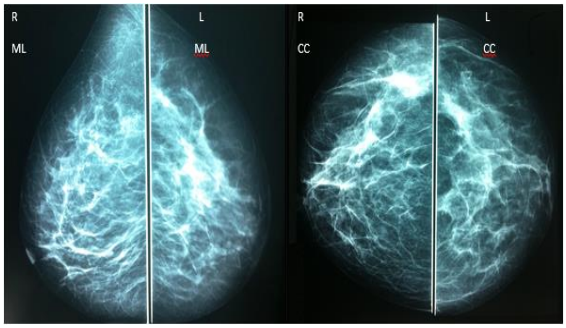

Figure 2: No abnormalities seen in mammography

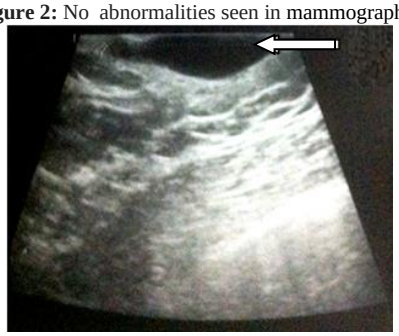

Figure 3: Ultrasound complement reveals the presence of a small subcutaneous cystic formation (arrow) of the left SEQ measuring 32 mm of major axis with thickened wall.

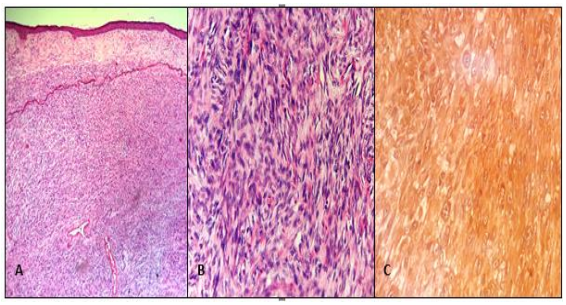

Figure 4: (a): morphological aspect (H&E stain x10); **A** fusocellular tumor proliferation made of bundles of fibroblasts arranged according to the aspect of radiuses of wheel. (**b**): morphological aspect (H&E stain x40); the tumor cells have elongated nuclei without frank atypical. (**c**)**:** Immunohistochemical (IHC) analysis of tumor cells showed strong immune reactivity to CD34**.**

DISCUSSION

Primary mesenchymal tumors of the breast are rare where sarcomas arising in the breast parenchyma account for 0.2 to 1.0% of all breast malignancies. The reported incidence of DFSP is approximately five cases per one million individuals per year. Dermatofibrosarcoma protuberans, first described by Darrier and Ferrand in 1924 and termed by Hoffmann in 1925, is an uncommon tumor originating in the skin and subcutaneous tissue [3].Being a rare dermal tumour with varied presentation, it can cause a diagnostic dilemma and needs differentiation from other lesion-like fibrous histiocytoma, phylloides tumour and other malignant breast tumours [1].DFSP arising in a breast is rare, and a comprehensive search on PubMed and Google Scholar displayed less than 90 such cases from 1990 to 2016.The clinical presentation is usually in the form of a firm, plaque-like lesion of the skin surrounded by blue discoloration. Many times, it may enter a rapid growth phase giving rise to multiple nodules. DFSP breast has been observed in patients taking estrogen replacement therapy and radiation therapy, during pregnancy and on preexisting surgical scars [3]. Histologically, DFSP is characterized by proliferation of the dermal spindle cells arranged in a storiform or cartwheel pattern. These proliferation are made up of monotonous cells with little polymorphism and low mitotic index [1-2]. Immuno- histochemical stains are required to differentiate dermatofibroma (DF), DFSP and other fibrous tumours. CD34, factor XIIIa, stromelysin-3 (ST3) and apolipoprotein D (Apo D) are some of the important stains used. CD34 positivity is seen in most cases of DFSP [4]. Genetically, DFSP is characterized by a reciprocal translocation t(17;22)(q22;q13), or more often as a supernumerary ring chromosome involving

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chromosomes 17 and 22. Patients often ignore these tumors due to their slow growth. DFSP breast involvement is rare, and is often misdiagnosed as a benign breast tumor, which delays treatment [4].

The best treatment option is wide local excision or Mohs micrographic surgery. Modified radical mastectomy is also an option in select group of patients who are likely to default follow-up. Local recurrence is high and metastasis is rare with DFSP[1].

To prevent the risk of locoregional recurrence, an excision with margins of 4 to 5 cm in width and carrying a healthy anatomical barrier in depth has long been the reference in France. The morphological and sometimes functional sequelae generated by these vast excisions are important, especially in the young subject. Several teams using the Mohs technique with extemporaneous examination have shown that margins of 3 cm were sufficient in this case to remove all the tumor cells. Some authors even proposed surgical margins to 2.5 cm and did not observe a recurrence. It is indeed the excision in depth that poses a problem.

Thus, the dogma of the removal of a healthy barrier remains polemical and the continuation of the studies on the vertical extension seems to impose itself. Finally, the size of the initial tumor could be another prognostic factor for recurrence [5].

Other modalities of treatment used are radiotherapy and the use of imatinib in selected patients mainly in advanced disease [1].

In the study evaluating the association of imatinib with surgery, Rutkowski et al had concluded that targeted therapy was effective by minimizing not only the resection margins but also the decrease in recurrence rates. As for radiotherapy, most of the literature defines Darier-Ferrand's sarcoma as a radioresistant tumor. However, others have argued that radiotherapy reduces the rate of local recurrence and allows for more limited surgery. Thus, according to Haas et al., Local tumor control was 82% with adjuvant radiotherapy for insufficient or invaded surgical margins, with a follow-up of 1 to 22 years [6]. In the present case, the patient is programmed for a wide local excision.

CONCLUSION

DFSP is a rare breast neoplasm. Despite its classification as a tumor of intermediate malignancy with a limited potential for metastasis, DFSP does possess the potential for aggressive local behavior and it may frequently recur if not extirpated completely. Dermatofibrosarcoma of the breast is a diagnostic challenge. Pre-operative investigations are usually non conclusive. Histopathological examination and immuno-histochemistry aid in establishing the definitive diagnosis. Complete surgical excision represents the treatment of choice of DFSP. However, the high locoregional recurrence potential transforms this lesion into an entity that is difficult to control clinically. Imatinib may assist in disease control in patients with locally advanced or metastatic disease.

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