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Case report

Breast location of Darier and Ferrand dermatofibrosarcoma: a rare case report

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Abstract

Darier and Ferrand Dermatofibrosarcoma (DFS) is a very uncommon pathology type of skin cancer. Breast location remains very rare, with only dozens of cases reported to date in the English literature. We report the case of a 21-year-old woman who presented with a left breast mass that had been present for more than 4 years. Clinical examination found cutaneous mass occupying the upper internal quadrant of the left breast measuring 3 x 2cm. The patient underwent an excision of the mass with positive margin. Histological and immunohistochemical findings were suggestive of a Darier and Ferrand Dermatofibrosarcoma (DFS). In front of this clinical board, wide excision of the mass was performed. 15 months after the end of surgery, the patient was in good health, with excellent local control. The purpose of presenting this case is not only to report an uncommon pathology subtype, but mostly to provide a long-term survival of Darier and Ferrand Dermatofibrosarcoma (DFS) of breast cancer that very rare.

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Introduction

Darier and Ferrand Dermatofibrosarcoma (DFS) is a rare variant of soft tissue sarcomas with an incidence rate of 4.2 to 4.5 cases per million persons per year in the United States. It accounts for 1% of soft tissue sarcomas and less than 0.1% of all malignant tumors [1, 2]. But DFS is even rarely found in breast cancers, with few clinical cases reported to date in the literature. Due to the scarcity of cases, there is no consensus on the best DFS treatment, and this entity is still challenging clinicians. In this paper, we report a case of DFS breast cancer and review of literature.

Patient and observation

A previously healthy 21-year-old woman presented with left breast cutaneous mass that had been present lasting for 4 years. Clinical examination found a cutaneous mass in the upper internal quadrants of the left breast measuring 3 x 2cm. There was no mammalian flow and the ganglionic areas were free. Mammary ultrasound found a mass in the left breast at the superficial oval heterogeneous echogenic level, well limited to regular contours, discreetly vascularized (Figure 1). Wide excision of the mass was performed. Microscopically, the tumor had a malignant mesenchymal nature, affecting the skin of breast. It was composed of subcutaneous tumor proliferation with storiform appearance of cellular beams and fusiform cell proliferation which dissociates the adipose tissue (Figure 2 A, Figure 2 B, Figure 2 C). Immunohistochemistry was positive for CD34 (Figure 2 D). The histopathologic and immunohistochemical diagnosis was consistent with Dermatofibrosarcoma (DFS) of Darier and Ferrand. In front of positive margins, the patient underwent a large excision. Fifteen months after the end of treatment, the patient was in good health, with excellent local and distant disease control.

Ethics approval: ethics approval was obtained from the ethics committee of the Ibn Zohr University in Agadir.

Discussion

Because of the rarity of Dermatofibrosarcoma of the breast, knowledge has been limited almost exclusively to case reports. To date, only dozens of cases reported to date in the English literature [3-14]. The purpose of presenting this case is not only to report an uncommon pathology subtype, but mostly to provide a long-term survival of Darier and Ferrand Dermatofibrosarcoma (DFS) of breast cancer that is very rare, and we are discussing treatment of mammary localization. The preferred site of DFS is the trunk 50 to 60%, the limbs 20 to 30% and the head 15 to 20% [12]. Its breast localization remains rare. Diagnosis is confirmed by biopsy. Microscopically, DFS typically presents as a storiform or fascicular proliferation of bland spindled cells that extends from the dermis into the subcutis [15-17]. The most common immunoprofile found in DFS is positivity for CD34 and negativity for S-100 protein and factor XIIIa. Dermatofibrosarcoma rarely metastasizes from where an extensive workup is not routinely indicated unless suggestive aspects in the clinical examination or adverse prognostic histologic features are present [15]. Surgery is the gold standard treatment. Every effort should be made to achieve clear surgical margins. There are varied approaches: Complete circumferential and peripheral deep margin assessment, Mohs micrographic surgery and wide excision with at least 2cm margins to investing fascia of muscle or pericranium with clear pathological margins, when clinically feasible [15]. Based on the limited available data, radiation therapy appears to be a reasonable approach for adjuvant therapy after surgery for positive surgical margins if further resection is not feasible. Indeed, many cases were treated by adjuvant radiation therapy with an excellent reported local and disease-free survival of 93% at 10 years [18]. Radiation therapy alone can be an option

in recurrent tumors and for patient who are not surgical candidates if not given previously. Given the high risk of recurrence, rigorous clinical supervision every 6 months is necessary with re-biopsy of any suspicious regions. In our patient, we did not detect local recurrence after 15 months.

Conclusion

Dermatofibrosarcoma of breast is an extremely rare soft tissue malignancy. It is characterized by a slow evolution, a significant risk of local recurrence and a low rate of metastasization.

Competing interests

The authors declare no competing interests.

Authors' contributions

Karima Mouden examined the patient, participated in the sequence alignment and drafted the manuscript. Soukaina Wakrim carried out radiological, histological and immunohistochemical studies and helped to draft the manuscript. Afaf Semmar participated in the sequence alignment, revised and helped to draft the article. All authors read and approved the final manuscript.

Figures

Figure 1: mammary ultrasound: mass in the left breast at the superficial, oval, heterogeneous echogenic level, well limited to regular contours, discreetly vascularized

Figure 2: (A) subcutaneous tumor proliferation (Gx100); (B) storiform appearance of cellular beams (Gx200); (C) fusiform cell proliferation which dissociates the adipose tissue (Gx200); (D) antibody CD34: diffuse positivity of fusiform cells

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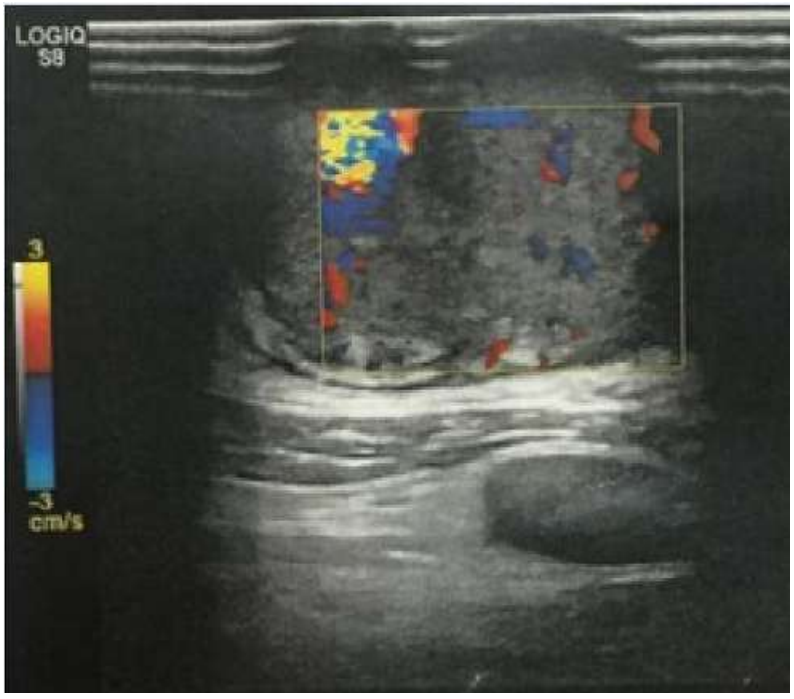

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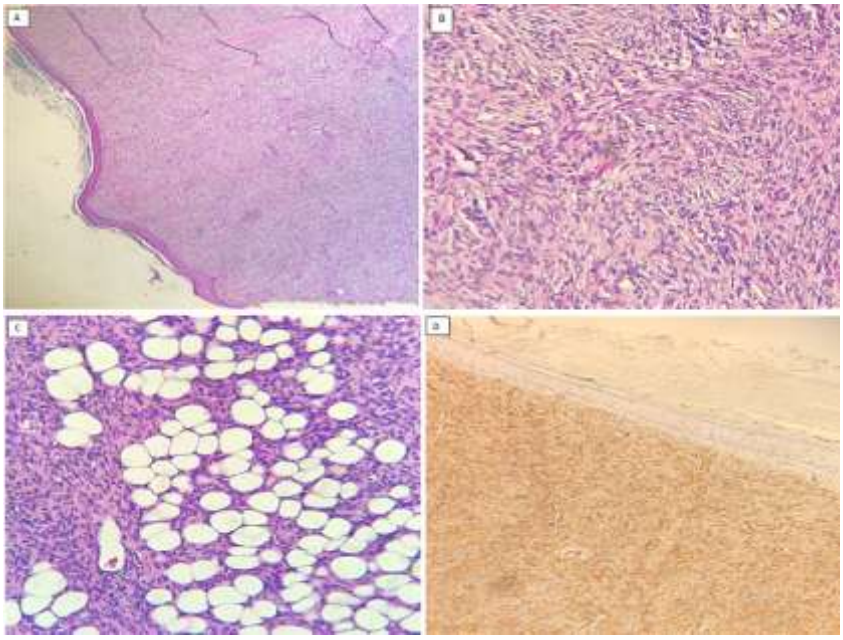

Figure 2: (A) subcutaneous tumor proliferation (Gx100); (B) storiform appearance of cellular beams (Gx200); (C) fusiform cell proliferation which dissociates the adipose tissue (Gx200); (D) antibody CD34: diffuse positivity of fusiform cells