Dermatology Practical & Conceptual

Multifocal Cutaneous Rosai-Dorfman Disease Treated With Localized Radiotherapy

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Introduction

Rosai-Dorfman disease (RDD) is a rare histiocytic disorder characterized by a benign proliferation of S100-positive histiocytes [1]. Skin involvement is present in 10% of cases: a purely cutaneous form (CRDD) is rare (3% of all cases) and has an older mean age of onset (43 years) than nodal disease (20 years) [2].

Case Presentation

A 38-year-old woman came to our attention presenting with a hyperpigmented infiltrated plaque in her left breast and right thigh for six months. The patient had already undergone biopsy with the diagnosis of: fibroadipose tissue, with lymphoplasmacytic and histiocytic infiltrate and presence of foamy histiocytes. Clinical evaluation revealed numerous pink-purple papulo-nodules with multiple yellowish-white areas, in the context of well-circumscribed infiltrated plaques (Figure 1, A and B). There were no systemic symptoms and no palpable lymphadenopathy. We carried out another skin biopsy of papules of breast and thigh and the histological features confirmed the diagnosis of RDD (Figure 2, A-C). To rule out systemic disease, a whole body computed tomography scan (CT) was performed with no evidence of organ involvement but showing a 6.3x5.9x7.2 cm lesion in the left breast and 16.6x7.5x15.9 cm in the right thigh. Laboratory work-up included: full blood count, liver function test, renal parameters, C-reactive protein, viral serology (HIV, HBV, HCV), ANA, rheumatoid factor were carried out. Results were all within normal parameters. These findings confirmed a diagnosis of CRDD without evidence of systemic RDD. As it was a localized cutaneous form, it was treated with topical clobetasol propionate twice daily for 2 months with little benefit. Given the subsequent worsening, the case was discussed in a multidisciplinary consultation with a radiotherapist and an oncologist, and it was decided to treat the patient with localized radiotherapy

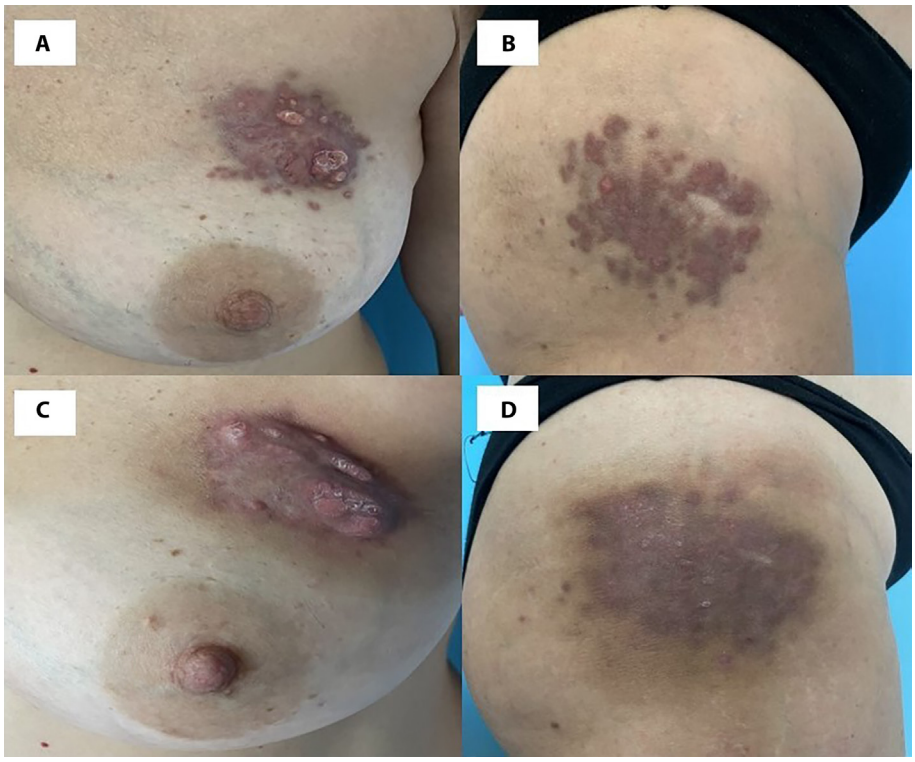

Figure 1. A-D. (A,B) Pink-violaceous papulonodules and plaques on left breast and right thigh at the time of the initial dermatology consultation (6.3x5.9x7.2 cm in the left breast and 16.6x7.5x15.9 cm in the right thigh). (C,D) Near-complete response at 4-month follow-up after completion of radiotherapy left breast lesion measured 3.21 x 3.68 x 3.4 cm (-47%), right thigh measured 10.56 x 5.4 x 7.8 cm (-38%).

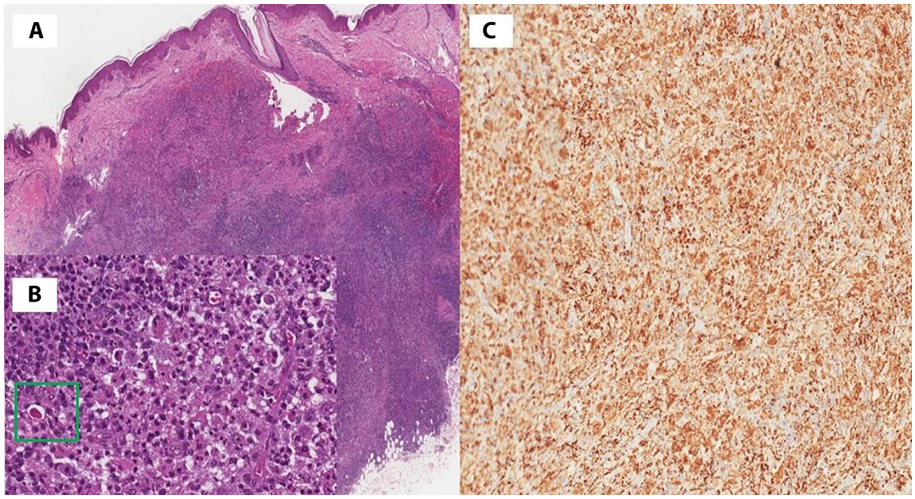

Figure 2. A-C. (A) Infiltrate consisting mainly of histiocytes mixed with neutrophils, plasma cells and T lymphocytes. H&E, original magnification: 4x. (B) Emperipolesis phenomena (presence of an intact cell within the cytoplasm of another cell) and histiocytes with granular cytoplasm (square.) H&E, original magnification: 40x. (C) Immunohistochemical shows positivity for S100 original magnification: 20x.

at a dose of 40 Gy in 20 fractions of 2 Gy. The patient tolerated radiotherapy well with only mild radiodermatitis. A follow-up whole body CT was performed four months after radiotherapy which showed a marked reduction in treated lesions, left breast lesion measured 3.21 x 3.68 x 3.4 cm (-47%), right thigh measured 10.56 x 5.4 x 7.8 cm (-38%) and an almost complete disappearance of radiodermatitis was observed (Figure 1, C and D). We hope for further improvement, however a possible surgical treatment is being evaluated.

Conclusions

CRDD is a very rare entity, the diagnosis may be suspected clinically but remains a histological diagnosis [3]. There are currently no standardized guidelines and treatment is individual. Spontaneous remission was observed in 20%-50% of cases [4]. Treatment options for CRDD include topical and systemic corticosteroids, dapsone, thalidomide, isotretinoin, imatinib, surgical excision (for localized form), chemotherapy. But also, physical treatments such as cryotherapy, local radiotherapy and laser therapy. As suggested by Choi et al, localized radiotherapy should be considered a valid therapeutic option for CRDD, which is challenging to excise or with aesthetic impact and for neoadjuvant purposes [5].

A multidisciplinary approach in the treatment of rare skin diseases such as CRDD is important. Localized radiotherapy could be a valid therapeutic option, furthermore in order to achieve standard dosing guidelines, new case submissions are essential.

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