A new presentation of isolated cutaneous Rosai-Dorfman disease: Eruptive xanthoma-like lesions

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Abstract

Rosai-Dorfman disease or sinus histiocytosis with massive lymphadenopathy is a benign lympho-histiocytic proliferative disorder initially described with bilateral painless lymphadenopathy (90 %), fever, leukocytosis, elevated ESR, anemia, and polyclonal hypergammaglobulinemia (90 %). Extranodal forms occur in 43% of cases, the skin being the most common site. Around 10% of patients have skin lesions and in 3%, the disease is limited exclusively to the skin. Here, we report a male patient who presented with pure cutaneous lesions which mimic eruptive xanthoma clinically. However, the diagnosis was established histo pathologically. So, high level of clinical suspension is critical to avoid missing such cases.

Key words: Emperipolesis, eruptive xanthoma, Rosai-Dorfman disease, skin

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Introduction

Rosai-Dorfman disease is a non-Langerhans cell histiocytosis first described in 1965 by Destombes and recognized as a distinct clinicopathological entity by Rosai and Dorfman in 1969.[1,2] Rosai-Dorfman disease presents with cervical adenopathy, most commonly in children or young adults (median age, 20 years), in those of African ancestry and male sex.[1] Extranodal disease occurs in 25-40%, is often widespread, involving skin, respiratory tract, soft tissue, paranasal sinuses, visceral organs, bone, central nervous system, genitourinary tract, and orbit.[2,3] Around 10% of patients have skin lesions and in 3%, the disease is limited exclusively to the skin.[3] Cutaneous Rosai-Dorfman disease presents as solitary or numerous papules, nodules, plaques or as a combination of these.[4]

Case Report

A 46-year-old male reported appearance of asymptomatic widespread skin lesions 4 months ago. The lesions grew rapidly on the trunk and left thigh and later spread to involve other extremities and face within a span of 2 months. The patient had no related medical history and was otherwise healthy. The patient reported no systemic symptoms such as fever, malaise or weight loss. There was no organomegaly or lymph node enlargement.

Cutaneous examination revealed innumerable generalized papules mainly affecting the face and limbs. The lesions were yellowish to orange papules with confluence forming non infiltrated plaques mainly on the thighs, cheeks, temples and peri-auricular area [Figure 1]. There was no mucosal or nail involvement. Our differential diagnoses were eruptive xanthoma, fibrofolliculoma, trichodiscomas, eruptive villous hair cysts as well as non-Langerhans cell histiocytosis. Routine laboratory tests, serum thyroid stimulating hormone, X-ray of

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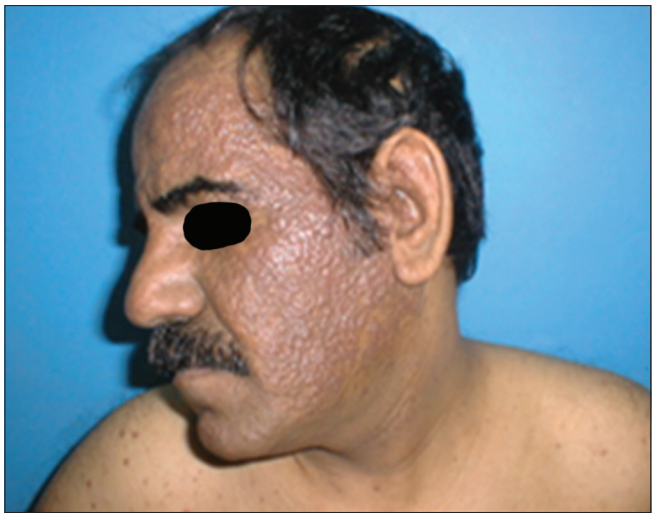

Figure 1a: Numerous yellowish to orange papules affecting cheeks, temples and forehead

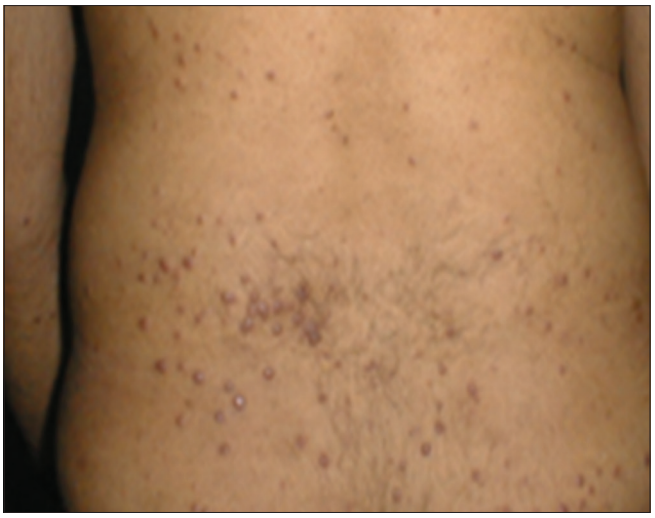

Figure 1b: Numerous yellowish to orange papules scattered on the trunk

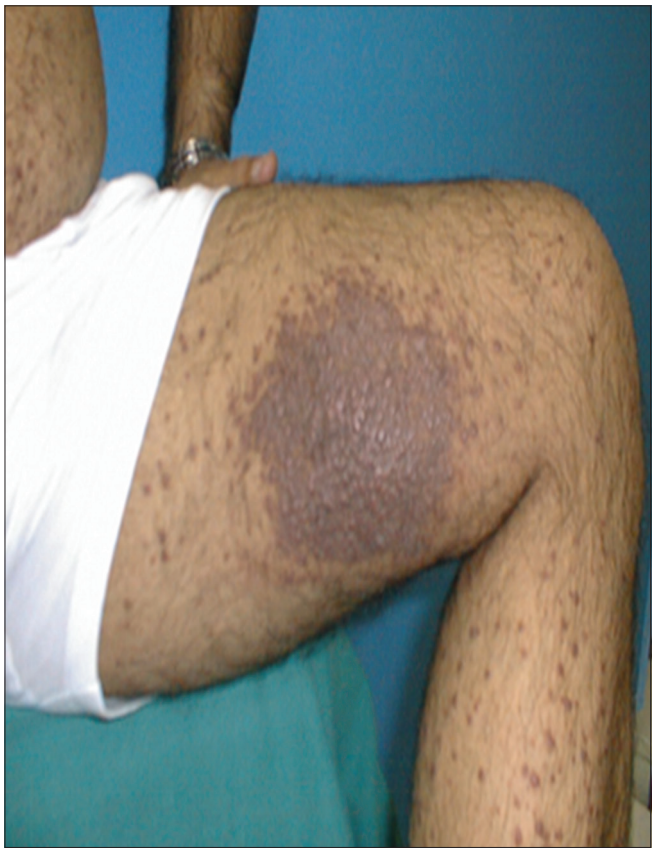

Figure 1c: Numerous yellowish to orange papules affecting limbs with confluence forming plaques on the medial aspect of left thigh

the chest and long bones and abdominal ultrasonography were normal. Computed tomography scan was carried out from the level of the craniocervical junction down to the groin and was normal. Lipid profile and serum protein electrophoresis tests were normal. Eye examination was unremarkable.

A skin biopsy showed dermal infiltrate composed mainly of pale-staining macrophages with large cytoplasm and vesicular nuclei with evident nucleoli. Emperipolesis was evident. Lymphocytes, neutrophils and plasma cells were also found [Figure 2]. Special stains including Giemsa stain, Fite Stain, Gram stain and periodic acid–Schiff were all negative. Immunohistochemistry revealed diffuse S-100 protein expression in the histiocyte cells, as well as positivity for CD68 and negativity for CD1a [Figure 3].

Correlating the clinical and histopathological data, the diagnosis of exclusively cutaneous Rosai–Dorfman disease was established. Follow-up of the patient up to 2 years after his initial presentation showed that the disease remained stationary without any clinical, laboratory or radiological progression to the systemic classic form.

Discussion

The term cutaneous Rosai–Dorfman disease is used when involvement is limited only to the skin.[1-3] Approximately 10% of Rosai–Dorfman disease patients have skin lesions, and in 3%, the disease exclusively affects the skin.[4,5] Cutaneous Rosai–Dorfman disease can present with papules, plaques, nodules and/or tumors of a brownish- or yellowish-erythematous color, varying in size from less than 1 cm to 30 cm, either localized or disseminated. The sites most commonly affected are trunk, head, neck, lower and upper extremities.[3] The lesions occasionally resemble psoriasis or acne.[6] We were unable to find any previous reports of cutaneous Rosai–Dorfman disease presenting as eruptive xanthoma like lesions in the literature.

The etiology of Rosai–Dorfman disease remains unknown. There are some reports that link the systemic form of the disease with herpes virus hominis-6 and 8 and Epstein–Barr

virus. Researchers have also attempted to demonstrate this coexistence in a small series of cases in cutaneous Rosai–Dorfman disease; however, the results were conflicting.[3,7] The finding of polyclonal gammopathy supports the hypothesis of a reactive process against infectious one.[1]

Histological findings in cutaneous Rosai–Dorfman disease are usually similar to those in the classic Rosai–Dorfman disease form. Typically, the dermis shows a diffuse infiltrate of histiocytes accompanied by a background infiltrate of lymphocytes and plasma cells. Emperipolesis, which represents intact lymphocytes within histiocytes, is common in cutaneous Rosai–Dorfman disease. Less often, the cytoplasm may contain plasma cells, neutrophils and red blood cells. Cutaneous Rosai–Dorfman disease histiocytes stain positively for S100 protein and CD68 but negatively for CD1a.[8]

Here, we report cutaneous Rosai–Dorfman disease manifesting in a new presentation – eruptive xanthoma-like eruption. The diagnosis of isolated cutaneous Rosai–Dorfman disease was made after excluding systemic involvement, either on first presentation or within 2 years of follow-up. Regarding treatment, we preferred wait and see policy, depending on the benign nature of the disease. Thus, cutaneous Rosai–Dorfman disease can be mistaken for a plethora of skin diseases, and can be included in the expanding list of skin imitators.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts

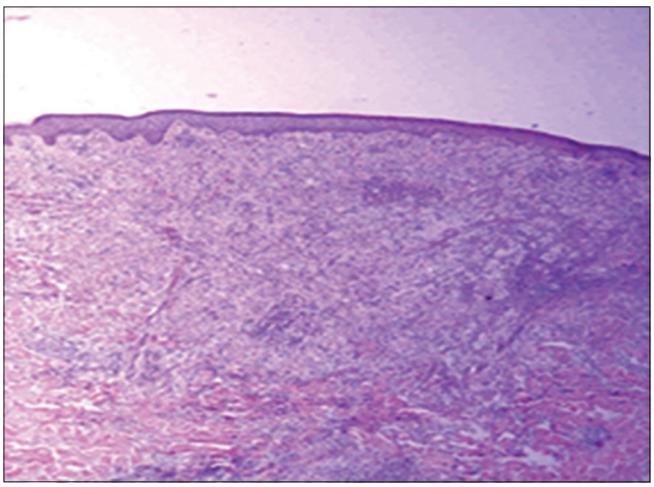

Figure 2a: Collections of lymphocytes in dermis (H and E, ×40)

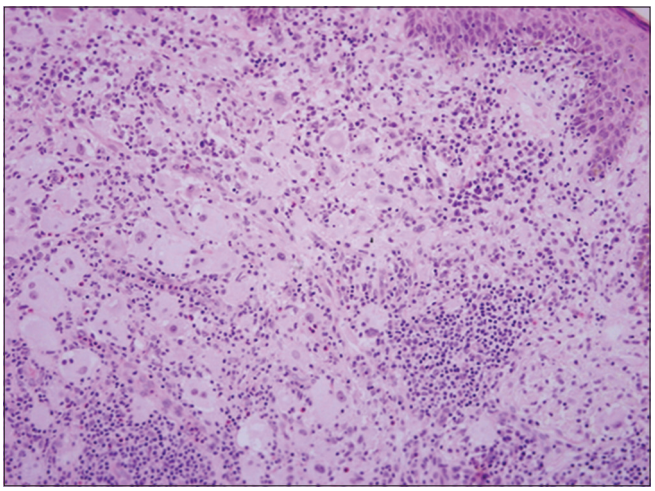

Figure 2b: Sheets of numerous large pale-staining macrophages (H and E, ×200)

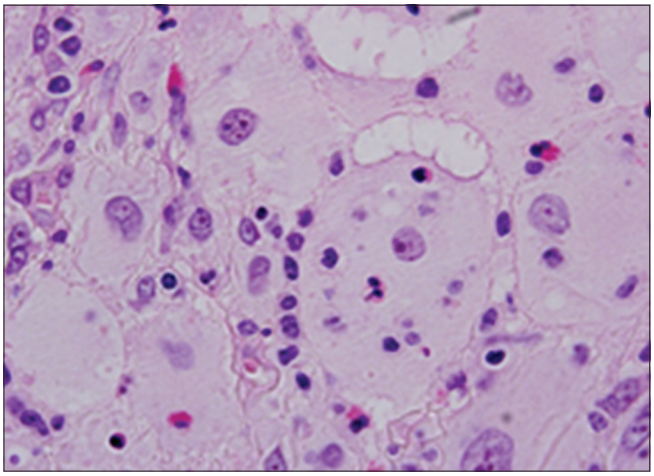

Figure 2c: Sheets of numerous large pale-staining macrophages with emperipolesis (H and E, ×400)

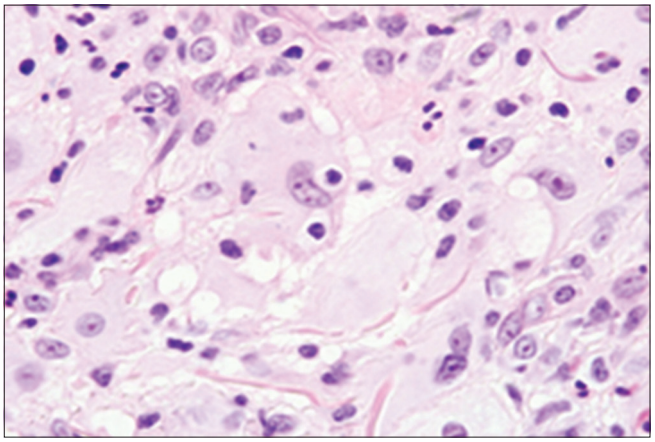

Figure 2d: Collections of lymphocytes ,and sheets of numerous large pale-staining macrophages (H and E, ×400)

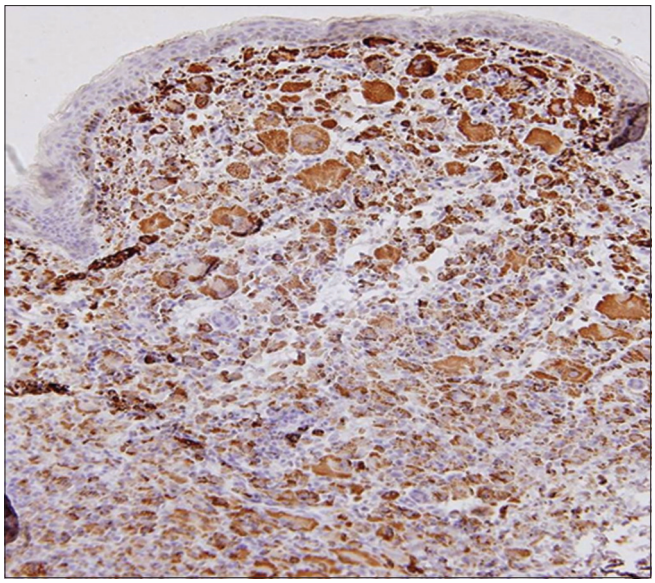

Figure 3a: Immunohistochemistry showing CD68 positive histiocytes (×100)

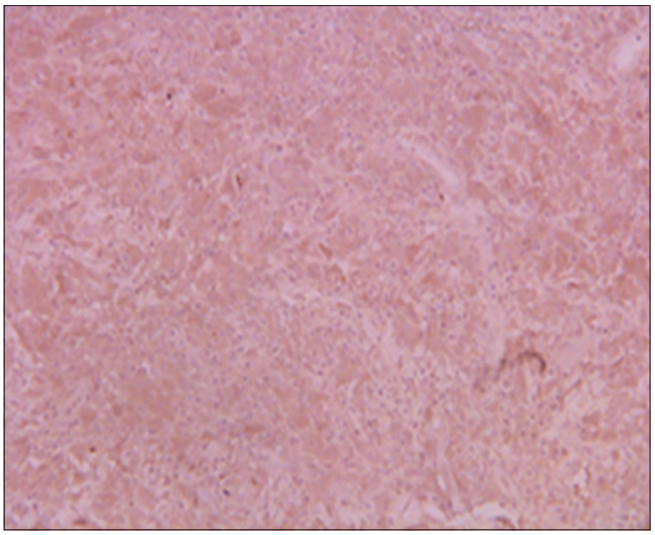

Figure 3b: Immunohistochemistry showing CD1a negative histiocytes (×100)

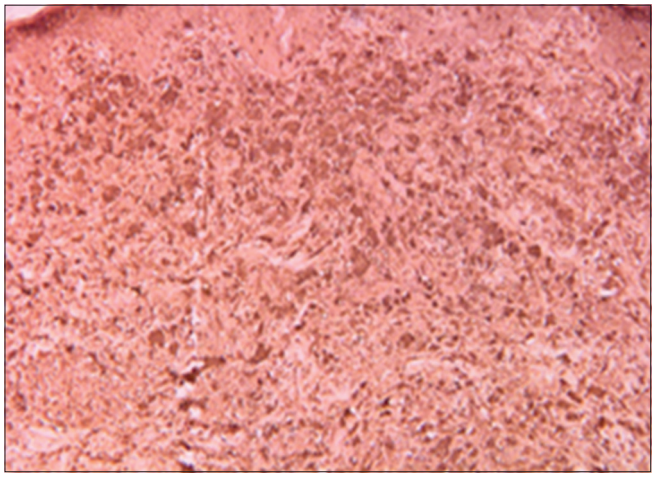

Figure 3c: Immunohistochemistry showing S100 positive histiocytes (×100)

will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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