A Rare Case of Purely Cutaneous Rosai–Dorfman’s Disease: Xanthoma-like Presentation

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Abstract

Rosai–Dorfman’s disease (RDD) is sinus histiocytosis with massive lymphadenopathy. Cutaneous RDD is a rare entity that presents with isolated skin involvement without any systemic involvement. We report a 35-year-old man with an asymptomatic large nodule on the preauricular region of the face with an irregular surface and multiple yellow studded nodules. On detailed physical and systemic examination, there was no lymph node or systemic involvement. Skin biopsy on histopathology showed dense dermal inflammatory infiltrate and foamy histiocytes with emperipolesis. Immunohistochemical markers were positive for S100 protein and CD68 protein but negative for CD1a. Thus, a diagnosis of cutaneous RDD was made.

Keywords: Emperipolesis, lymphadenopathy, Rosai–Dorfman’s disease, sinus histiocytosis

Introduction

Rosai–Dorfman’s disease (RDD) is a rare non-Langerhans cell histiocytosis characterized by lymph node involvement along with extranodal organ involvement. Extranodal disease majorly involves the skin, respiratory tract, solid organs, bone, central nervous system, genitourinary system, orbit, and ears. Skin involvement is seen in 10\% of cases and about 3\% of cases have disease limited exclusively to the skin.\textsuperscript{[1]} The term cutaneous Rosai–Dorfman’s disease (CRDD) is used when involvement is limited only to skin without systemic involvement. We report a rare case of CRDD in a male patient presenting as nodular growth on face.

Case Report

A 35-year-old male presented with a right-sided preauricular slowly enlarging growth for 3 months. The lesion started as an erythematous papule around pea-sized which gradually enlarged in size, with occasional pain. There was no history of trauma before the occurrence of the lesion. He had no history of fever or weight loss. On examination, a well-defined nodule with an erythematous base measuring 5 cm×6 cm was seen in the right preauricular area. The surface was irregular and studded with multiple yellow nodules [Figure 1A]. On palpation, it was soft in consistency and fixed to underlying tissue. On physical examination, there was no lymphadenopathy or hepatosplenomegaly. Differential diagnosis of deep fungal infection, cutaneous lymphoma, cutaneous xanthoma, and lupus vulgaris was kept. All routine laboratory investigations and serum lipids were within normal limits. Chest X-ray and ultrasonography of the abdomen and pelvis did not show any abnormality. Local X-ray of the preauricular region did not show any evidence of foci of infection or fracture.

A skin biopsy performed from the nodule showed a dense dermal infiltrate composed of foamy histiocytes, lymphocytes, neutrophils, and plasma cells [Figure 2]. At foci, lymphocytes were seen engulfed by histiocytes: a phenomenon called emperipolesis [Figure 3]. PAS and Giemsa stain did not reveal any organisms. There were no evidence of foci of infection or fracture. Xanthoma-like presentation. Indian J Dermatopathol Diagn Dermatol 2021;XX-XX-X.

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no caseating granulomas or Langhans giant cells which ruled out cutaneous tuberculosis. Cutaneous lymphoma was ruled out because of absent lymphocytic aggregates and epidermotropism. Tissue culture was negative for fungi; therefore, deep fungal infection was ruled out. Normal serum lipids and absence of lipid deposits on histopathology helped to ruled out cutaneous xanthomas. Immunohistochemical study was positive for S100 [Figure 3A] and CD68 [Figure 3B] but negative for CD1a. The diagnosis of CRDD was confirmed considering the clinicopathological co-relation and CD68 and S100 positivity and CD1a negativity. The patient was started on Tab. Methotrexate 15 mg weekly and topical 2% fucidin cream for local application. Intrallesional triamcinolone acetonide 10 mg/mL was injected in the lesion once in 3 weeks for 3 months. After 3 months follow-up, the size of the lesion was decreased. It was interesting to note the involution at the center of the lesion at the site of biopsy [Figure 1B].

**Discussion**

RDD was first described in 1965 by Destombes and recognized as a distinct clinicopathological entity by Rosai and Dorfman in 1969.[1-3] It is a rare non-Langerhans cell histiocytosis characterized by an accumulation of activated histiocytes within affected tissues.[4] CRDD can present with papules, plaques, nodules of erythematous with brownish to yellowish color. Size may vary from less than 1 cm to several centimeters. Cutaneous lesions can be either localized or disseminated. In few cases, satellite lesions have been described.[4] Trunk, head, neck, and lower and upper extremities are the most commonly affected sites.[5] Rare presentations include eruptive xanthoma-like pustular and acneiform lesions, and lesions mimicking vasculitis and panniculitis have also been reported.[1,4]

In our case, solitary xanthoma-like presentation is seen which is rarely reported. Patients with CRDD are in normal health and do not show lymphadenopathy, fever, night sweats, and the laboratory findings are within normal limits.[2] Some authors regard CRDD and RDD variants of the disease as distinct clinical entities. CRDD has a later age of onset (median age, 43.5 years) compared with RDD. Also, CRDD shows a marked female predominance (2:1) and most commonly affects Asian and Caucasian individuals. In contrast, RDD has a median onset age of 20.6 years and is slightly more common in males (1.4:1). The majority of RDD patients are of African descent and the disease is rarely reported in Asian patients.[3] The exact etiology of RDD is unknown. The polyclonal nature of the cell infiltrate and the clinical progression of RDD suggest a reactive process rather than a neoplastic disorder.[4] In some patients, infectious agents including EBV, HHV 6, parvovirus B19, HSV, Brucella, Klebsiella rhinoscleromatis, and Nocardia have been implicated.[5] The cell of origin in RDD is uncertain. Recent studies have identified NRAS, KRAS, MAP2K1, and ARAF mutations in patients with features of RDD.[7-8] Histological findings in CRDD show diffuse dermal infiltrate of histiocytes, lymphocytes, and plasma cells. Emperiploise, which represents intact lymphocytes within the cytoplasm of histiocytes, is also seen in CRDD.

Less often, the cytoplasm may contain plasma cells, neutrophils, and red blood cells. Emperiploise is a helpful finding but is not required for diagnosis, because it can be focal, especially at extranodal sites, and may be seen focally in another histiocytosis such as Erdheim-Chester disease, juvenile xanthogranuloma, and malignant histiocytosis.[9]
Immunohistochemistry of histiocytes in RDD shows S100 and CD68 positivity with variables CD14 and CD163 being positive.[3] CD1a staining is negative and electronic microscopy shows the absence of Birbeck granules which rules out Langerhans cell histiocytosis.[4] The differential diagnosis of CRDD includes sarcoidosis, lupus vulgaris/non-tuberculous mycobacterial infections, Hansen’s disease, leishmaniasis, and cutaneous lymphomas.[2]

There is no specific treatment and the disease is considered self-limited due to spontaneous resolution and usually does not require aggressive intervention. Treatment modalities for CRDD include corticosteroids, dapsone, thalidomide, isotretinoin, acitretin, and pulsed dye laser with variable success rates.

Surgical excision can be an effective treatment modality.[4] In our case, Tab. methotrexate 15 mg weekly dose and intralesional steroid injection showed involution and a decrease in the size of the lesion along with decrease in pain experienced by the patient.

**Conclusion**

CRDD mimics various dermatological diseases and has a wide variety of presentations. Histopathology and immunohistochemistry help in the diagnosis. Isolated purely cutaneous xanthoma-like presentation in a young male patient is rare. Intralesional corticosteroids and oral methotrexate can be considered for management of such a case. Dermatologists and dermatopathologists should be aware of this condition.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

**References**


Figure 3: A: Histiocytes showing S100 positivity (40×). B: Histiocytes showing positivity for CD68 protein (40×)

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