

CASE REPORT

Cutaneous Rosai-Dorfman Disease - A Rare Non-Langerhans Cell Histiocytosis

Afthab Jameela Wahab, S. Athilakshmi, Nivvedhetha S.

Abstract

Cutaneous Rosai-Dorfman disease is classified as non-Langerhans cell histiocytosis. It is a benign lymphoproliferative disorder involving only skin and subcutaneous tissue, which is rare and not well documented. It manifests as erythematous to brown papules, plaques, or nodules with histiocyte-rich inflammatory infiltrate which constantly exhibit emperipolesis i.e., uptake of intact lymphocytes and plasma cells; they express both Langerhans cell and macrophage markers (S100 and CD68 respectively). We report a case of a 45-year-old female presenting with a plaque on right cheek since 10 months without systemic symptoms. The lesion was excised in-toto and defect covered with rhomboid flap repair.

Key Words

Histiocytosis, Emperipolesis, Cutaneous Rosai-Dorfman

Introduction :

Cutaneous Rosai Dorfman disease (CRDD) is a rare non-Langerhans cell histiocytosis without systemic involvement, reported in 3%.^[1] It is an extranodal variant of Rosai Dorfman disease; a distinct clinicopathological entity mainly affecting middle aged females and usually presenting as plaques and nodules with satellite papules. Histopathologically it is characterized by emperipolesis, the histiocytes being CD1a⁺, S100⁺ and CD68⁺. Here we present a 45 year old female with skin lesion of ten months duration. The diagnosis of CRDD was confirmed by histopathology and Immunohistochemistry and the lesion excised completely.

Case Report

A forty-five year old female presented with an erythematous growth over the right cheek. Ten months ago it started without antecedent trauma and associated fever or pharyngitis, as three grouped shiny white

asymptomatic papules. These coalesced to form a red plaque. This increased in size with appearance and fusion of satellite papules. In the past five months, the lesion became itchy, painful with burning sensation; there was no discharge, no aural or mucosal lesions, and no history suggestive of malaise, lymphadenopathy, tuberculosis, weight loss, loss of appetite or atopy.

Patient was on anti-hypertensives, statins and thyroxine for the past five years, and oral hypoglycemics for the past six months.

Patient's vitals and systems examination were normal. Dermatological examination revealed an erythematous 4cm-diameter firm swelling with a polycyclic margin over the right parotid area: the surface was crusted in places and studded with small white papules and telangiectasia (*Fig1*). It was tender on deep pressure, not fixed to underlying structures. There was perilesional erythema

Department of Dermatology, Venerology & Leprosy, Saveetha Medical College and Hospital, Thandalam, Chennai, Tamil Nadu, India

Correspondence to: Dr. Nivvedhetha S, Post Graduate, Department of Dermatology, Venerology & Leprosy, Saveetha Medical College and Hospital, Thandalam, Chennai - 602105, Tamil Nadu, India

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Fig 1. Tumour showing surface telangiectasia, white papules and satellite papules

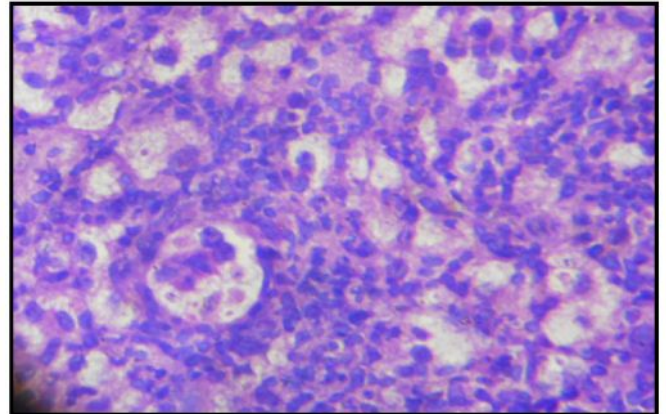


Fig 2. Histiocytes with foamy cytoplasm (H&E, 40x)

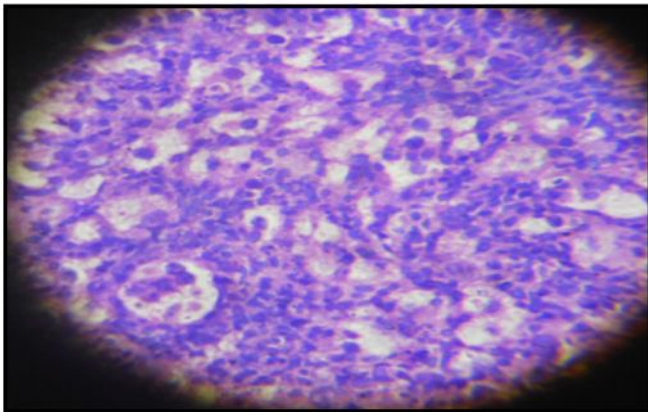


Fig 3. Histiocytes showing emperipolesis (H&E, 40x)

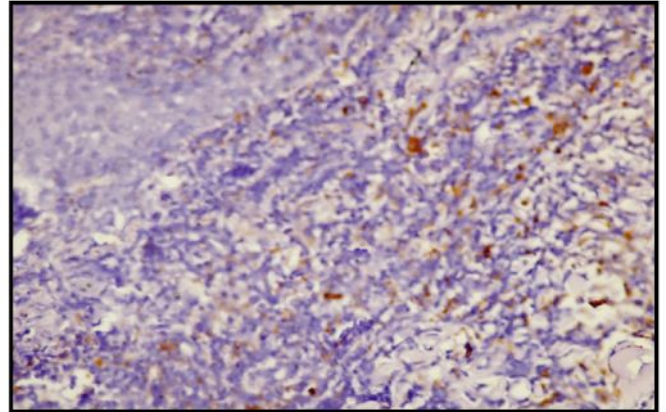


Fig 4. CD68 positive histiocytes (40x)

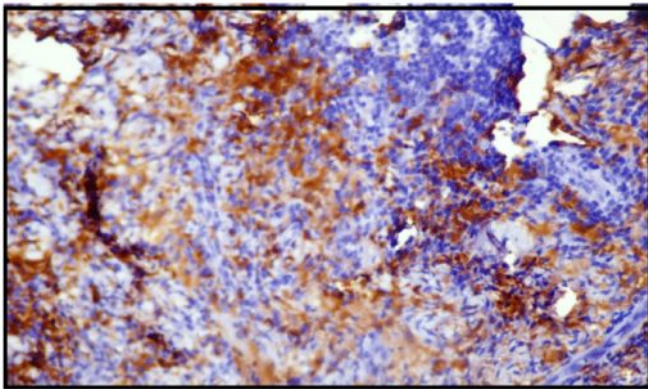


Fig 5. S-100 positive histiocytes (40x)



Fig 6. Post surgical photograph of the lesional site

and few erythematous shiny papules. There were no mucosal lesions, regional or generalized lymphadenopathy or organomegaly. Complete hemogram, liver and renal function tests, blood sugar, serum lipid profile were normal, VDRL non reactive and viral markers were negative. Histopathology showed dense dermal infiltrate of histiocytes and inflammatory

mononuclear cells. There were large histiocytes with foamy cytoplasm (*Fig 2*) and few large histiocytes demonstrating emperipolesis (*Fig 3*). Immunohistochemical staining revealed CD 68 + histiocytes (*Fig 4*) and S100 positive histiocytes (*Fig 5*). She was diagnosed as CRDD, started on systemic steroids. The response being unsatisfactory she was

referred to plastic surgery; a total excision was done with a rhomboid flap repair to cover the defect. This case is being reported for its rarity and specific histological and immunohistochemical findings.

Discussion

Cutaneous histiocytes include Langerhans cells, mononuclear cells/macrophages and dermal dendrocytes (type 1- Factor XIIIa +cells and type 2- CD3+ cells).^[2] Histiocytoses are proliferative disorders involving bone marrow derived CD34 positive progenitor cells. They include two closely related groups, namely, Langerhans cell histiocytoses and non Langerhans cell histiocytoses. Non Langerhans cell histiocytoses may be primarily cutaneous, cutaneous with systemic involvement or systemic with rare skin involvement.

Rosai-Dorfman disease (RDD), originally called sinus histiocytosis with massive lymphadenopathy,^[3] is a rare, benign, reactive non Langerhans cell histiocytosis. Antecedent non-specific fevers and pharyngitis may herald the onset of RDD.^[4] Of unknown etiology, RDD mostly affects lymph nodes, along with extranodal sites in 43%, the skin in 10%. Systemic RDD affects younger persons, mostly blacks, with associated fever, systemic symptoms, lymphadenopathy, anaemia, leukocytosis, elevated ESR and hypergammaglobulinaemia.

Purely cutaneous RDD (C-RDD) limited to the skin without systemic involvement, is reported in 3 %.^[5] A distinct clinicopathological entity^[6], C-RDD commonly presents mainly in Asians and whites as a noduloplaque with satellite papules^[7] over the face, trunk or extremities at a mean age of 43.5 years with a female predominance.^[8] There are no significant systemic, extracutaneous or serologic manifestations. Histologically lesions involve the dermis and subcutis, the pathognomonic cells being histiocytes with abundant cytoplasm, indistinct borders, large vesicular nucleus and prominent nucleoli expressing both macrophage markers CD68 and MAC387 and Langerhans cell marker S100.^[9] There are also numerous plasma cells, lymphocytes and polymorphs; the hallmark is emperipolesis, i.e., large histiocytes with intracytoplasmic intact inflammatory cells.^[10] CRDD is benign and self limited.^[11] Treatment depends on severity of disease, patient acceptance and treatment complications. Various modalities include surgical excision, cryotherapy, isotretinoin, chemotherapy and radiotherapy.

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Nil.

Conflicts of Interest

There are no conflicts of interest.

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