CASE REPORT

Unifocal cutaneous Rosai–Dorfman–Destombes disease: A case report in an adult patient.

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ABSTRACT

In this article, we present the case of a 51-year-old woman with a 1-month history of a new-onset lesion, characterized by a violaceous nodule measuring 1.5 × 1 cm located in the left temporal region. A biopsy was performed, revealing dermal proliferation of histiocytes, negative for CD1a, with positive immunohistochemistry for staining S100 and Ki67, and focal positivity in reactive cells for Bcl2, CD20, CD3, and Bcl6. These findings confirmed the diagnosis of cutaneous Rosai–Dorfman–Destombes disease (RDD-C), which was successfully treated with surgical excision.

INTRODUCTION

Rosai–Dorfman disease (RDD) is a non-Langerhans cell histiocytosis characterized by excessive accumulation of histiocytes, classically presenting as massive, painless bilateral lymphadenopathy. It is classified into sporadic, familial, or cutaneous forms and may be associated with neoplasms or autoimmune diseases.[1,2] Extranodal RDD occurs in up to 40% of cases, with involvement of sites such as the skin, nasal cavity, orbital tissue, and central nervous system (CNS).[3] The isolated cutaneous form is exceptionally rare.[1]

When present, it most commonly involves the head and face, followed by the trunk and extremities.[4] Clinical variants include papulonodular (most common), plaque-type, and tumoral forms.[4,5] The pathogenesis is not fully understood; however, associations with viral infections such as Human Immunodeficiency Virus (HIV), Cytomegalovirus (CMV), Herpes Simplex Virus (HSV), and Epstein–Barr Virus (EBV) have been described.[1] Diagnosis is based on biopsy findings, which typically show sinusoids filled with large histiocytes, along with immunohistochemical positivity for S100, CD68, and CD163, and negativity for CD1a and CD207.[3,6] Molecular studies have revealed recurrent mutations involving genes in the Mitogen-Activated Protein Kinase (MAPK)/Extracellular Signal-Regulated Kinase (MAPK/ERK) pathway.[7] Mutations in certain kinase pathways, including the KRAS proto-oncogene (a GTPase), the neuroblastoma RAS viral oncogene homolog (NRAS), the serine/threonine-protein kinase A-Raf (ARAF), and the MAPK kinase pathway (MAP2K1), suggest that in some cases the disease may be more neoplastic than inflammatory.[4]

We report a new case of cutaneous Rosai–Dorfman disease, confirmed by biopsy and successfully treated with surgical excision.

CASE REPORT

A 51-year-old female patient from Sucre, Bolivia, with a history of hypothyroidism treated with levothyroxine, presented with a 1-month history of a lesion characterized by an erythematous-violaceous nodule located in the left temporal region, measuring 1.5×1 cm. The lesion was non-tender on palpation, firm, and adherent to deeper planes (Figure 1). No cervical or axillary lymphadenopathy or other significant findings were noted on physical examination.



Figure 1: Erythematous-violaceous nodule in the left temporal region, with well-defined borders, non-tender on palpation, adherent to deeper planes, measuring 1.5 × 1 cm.

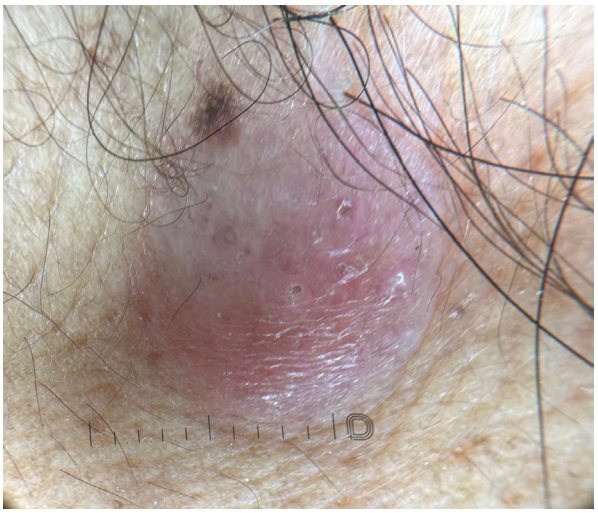


Figure 2: Dermoscopy shows reddish areas with some keratin plugs and increased vascularity.

Dermoscopy revealed reddish areas with some keratin plugs and increased vascularity. (Figure 2)

A biopsy of the lesion was performed, revealing dermal proliferation of histiocytic cells, negative for CD1a, with positive immunohistochemistry for S100 and Ki67, and focal positivity in reactive cells for Bcl2, CD20, CD68, CD3, and Bcl6. (Table 1) (Figure 3-5)

Table 1: Immunohistochemistry.

S100	POSITIVE
Bcl2	POSITIVE
Bcl6	POSITIVE
Ki67	POSITIVE
CD3	POSITIVE
CD20	POSITIVE
CD68	POSITIVE
CD1a	NEGATIVE

S100: S100 protein (positive).
CD3, CD20, CD68 (glycoproteins): Positive for histiocytosis (CD68), with reactive positivity in lymphocytic cells (CD3, CD20).
Ki67: Marker of mitotic activity.
Bcl2, Bcl6 (lymphocytic markers): Reactive positivity.
CD1a: Negative (dendritic cells, particularly Langerhans cells).

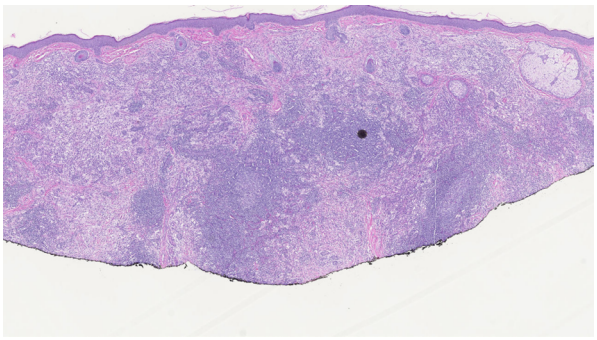


Figure 3: Dermal proliferation of histiocytic cells.

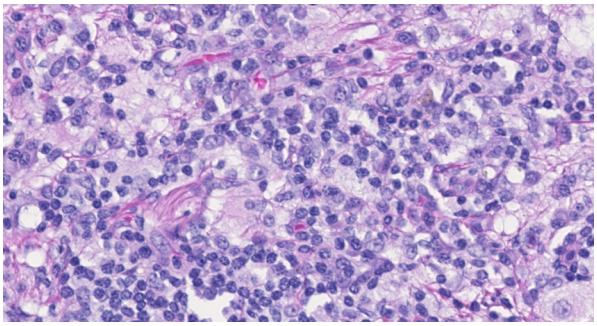


Figure 4: Multiple histiocytes, monocytes, and lymphocytic infiltrate are observed.

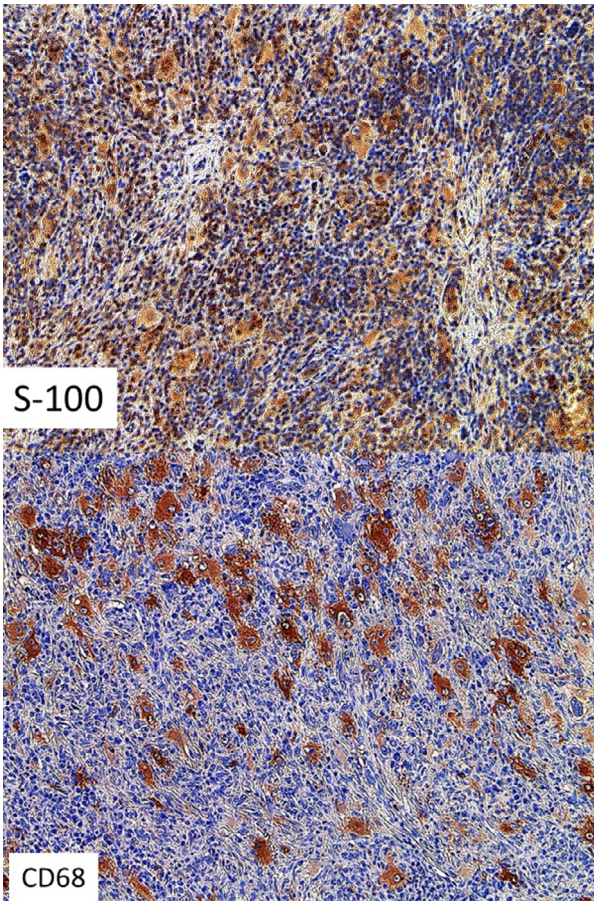


Figure 5: Positive immunohistochemical staining for S100 and CD68.



Figure 6: Follow-up at 3 months. Linear scar in the left temporal region.

Clinical findings and histopathology led to the diagnostic conclusion of cutaneous Rosai–Dorfman disease. Imaging studies for disease extension (cranial CT scan) were performed, ruling out involvement of other organs.

The therapeutic approach was surgical excision of the lesion, with no evidence of recurrence at 3-month follow-up. (Figure 6)

DISCUSSION

Cutaneous Rosai–Dorfman disease (RDD-C) represents approximately 3% of all forms of Rosai–Dorfman disease. It is a self-limited condition, with spontaneous remission occurring in up to 50% of cases; however, diagnosis remains challenging.[3] Biopsy provides the hallmark feature of this disease, in which emperipolesis (cell-in-cell phenomenon) is clearly evident. This refers to a biological process in which one cell penetrates another, remains viable throughout the process, and may exit at any time. Specifically, Rosai–Dorfman disease is characterized by the engulfment of plasma cells and lymphocytes by histiocytes.[9,10,11] Clinically, it shows a predominance in males and typically affects young adults.[3,8,9] In this context, the presented case is noteworthy due to its less common demographic presentation, which may initially lower diagnostic suspicion. However, several case series report a reversal in the male-to-female ratio and a trend toward older age at presentation.[10] There is no consensus regarding the treatment of RDD-C, and therapeutic management should be individualized according to the patient's clinical scenario.[3] Surgical excision is a highly effective treatment for small, unifocal lesions, with an efficacy of approximately 80%.[4] Additionally, systemic corticosteroids, methotrexate, rituximab, imatinib, vemurafenib, isotretinoin, and thalidomide have been used, with variable therapeutic responses.[1,3,4] Differential diagnosis includes dermatofibroma, Tangier disease, Langerhans cell histiocytosis, xanthogranuloma, B-cell lymphoma, and leprosy.[4]

CONCLUSION

Isolated cutaneous Rosai-Dorfman disease is an exceptionally rare condition, and its diagnosis and management remain challenging. However, surgical excision in unifocal lesions, as demonstrated in this case report, continues to be the treatment of choice.

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CONFLICTS OF INTEREST

The authors declare no conflicts of interest.

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