



Facial Cutaneous Rosai-Dorfman Disease: Dermoscopic Findings with Successful Surgical Treatment

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Rosai-Dorfman disease (RDD) is a rare non-Langerhans cell histiocytosis characterized by an accumulation of activated histiocytes within the affected tissues. It is a heterogeneous disease that includes the classical (nodal) and extra-nodal variants. The cutaneous form of the disease without the characteristic lymphadenopathy is rare and is often misdiagnosed as other dermatologic diseases. Misdiagnosis as lymphoproliferative and infectious diseases such as lymphoma and tuberculosis have been reported in the literature. Herein, we report a case of facial cutaneous RDD with successful surgical treatment. In addition, we provide dermoscopic findings and literature review as dermoscopy can be a useful adjuvant tool in the diagnosis of cutaneous RDD.

Keywords: Dermoscopy, Histiocytes, Histiocytosis, Rosai-Dorfman disease, Surgery, Treatment

INTRODUCTION

Rosai-Dorfman disease (RDD) is a rare non-Langerhans cell histiocytosis characterized by an accumulation of activated histiocytes within the affected tissues¹. It is a heterogeneous disease that includes the classical (nodal) and extra-nodal variants, leading to cutaneous, intracranial, spinal, central nervous system, ophthalmic, gastrointestinal, bone, and hematologic involvement. The cutaneous form of the disease without the characteristic lymphadenopathy is rare and is often misdiagnosed as other dermatologic diseases such as sarcoidosis, lymphoma, and nontuberculous mycobacterial (NTM) infection. Herein, we report a case of facial cutaneous RDD with successful surgical treatment.

CASE REPORT

A 47-year-old female presented with well demarcated erythematous to yellowish plaques on both cheeks since 18 months ago (Fig. 1). She reported that the lesions occurred

two months after injection of filler on her face. She had previously undergone skin biopsy at a local hospital; however, the diagnosis was inconclusive. She was presumptively diagnosed as NTM infection and was treated with clarithromycin and doxycycline for several months. However, there was no improvement. Therefore, she was referred to our department for further examination. Dermoscopic examination revealed yellowish structureless patterns on an erythematous background with linear branched vessels (Fig. 2A). Since empirical antibiotic treatment was ineffective, we performed a skin biopsy. Histopathological analysis revealed a dense lymphocytic and histiocytic infiltration with moderate plasma cell infiltration. Under high magnification, emperipolesis was detected with positive CD68 and S-100 and negative CD1a staining (Fig. 2B~F). Systemic work-up, including the F-18 fluorodeoxyglucose positron emission tomography scan, did not reveal any systemic involvement of RDD. The diagnosis of cutaneous RDD was confirmed. Systemic steroid therapy with a tapering regimen for 3 months (initial dosage of 0.5 mg/kg prednisolone) did not render significant improvement. As excision of



Fig. 1. (A, D) Clinical images of cutaneous Rosai-Dorfman disease on the face. (B, E) Clinical images of the patient after systemic steroid treatment. The nodule on the left lower cheek was excised. (C, F) Postoperative photographs 19 months after surgical excision.

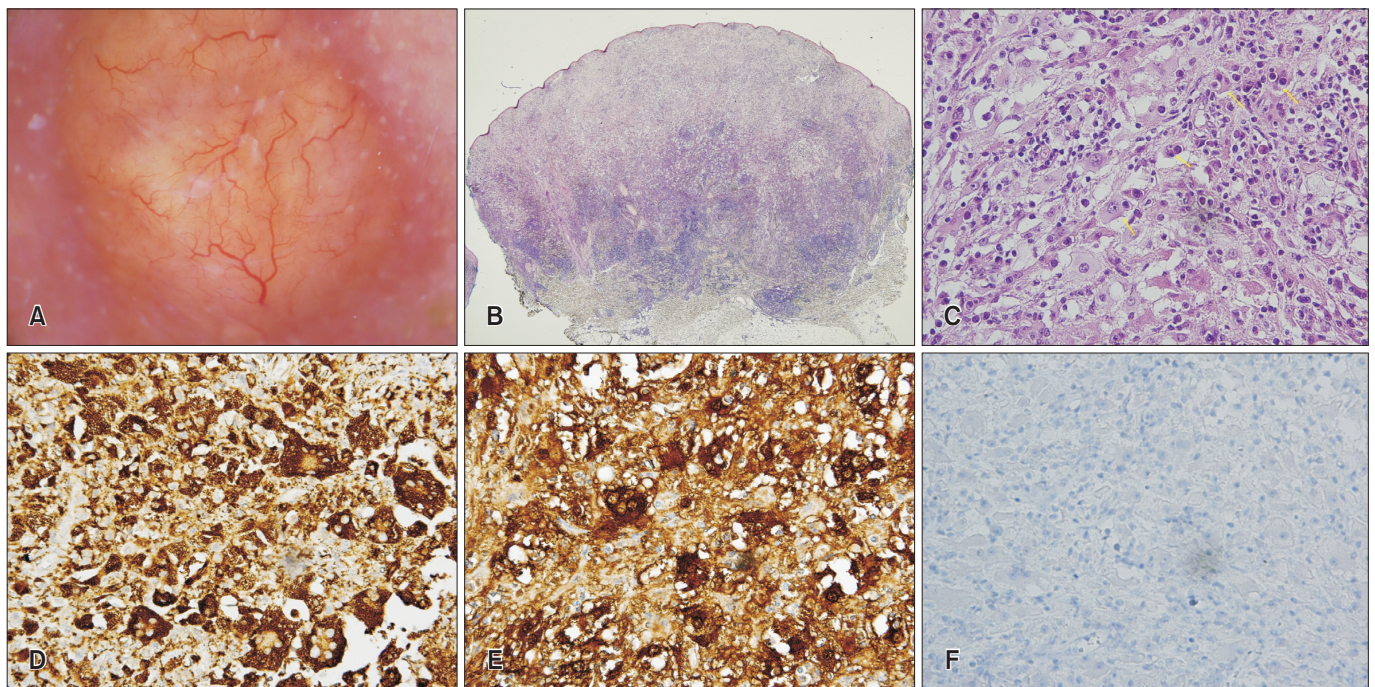


Fig. 2. (A) Dermoscopy revealing central yellowish structureless patterns on an erythematous background with arborizing vessels. (B, C) Histopathological images showing engulfment of lymphocytes by histiocytes (emperipolesis) (H&E, [B] $\times 20$, [C] $\times 400$, yellow arrows indicating emperipolesis). (D~F) Immunohistopathological studies showing positive staining with CD68 and S-100 and negative staining with CD1a (D: CD68, $\times 400$; E: S-100, $\times 400$; F: CD1a, $\times 400$).

the nodule on the left lower cheek was satisfactory (Fig. 1E), we performed staged elliptical excision under the patient's request (left cheek, two times; right cheek, one time), which resulted in a satisfactory cosmetic outcome (Fig. 1C, F). No recurrence was observed for two years.

Written informed consent from the patient for use of images and publication of case details was obtained.

DISCUSSION

Diagnosis of cutaneous RDD is challenging. Misdiagnosis is common due to its rarity and relatively nonspecific clinical findings. Emperipolesis, which indicates the presence of intact inflammatory cells within the cytoplasm of histiocytes, is the hallmark for the diagnosis of RDD. However, it is often subtle and difficult to identify, especially in the absence of a sufficient number of histiocytes showing emperipolesis in the biopsied specimen. The possibility of misdiagnosis is more likely for the cutaneous form of RDD as emperipolesis might be less conspicuous in the cutaneous form of RDD than in the lymph node of nodal RDD². In addition, due to dense inflammatory infiltration, including lymphocytes and plasma cells, histopathological diagnosis is challenging even to experienced

pathologists. Therefore, misdiagnosis as lymphoproliferative and infectious diseases such as lymphoma and tuberculosis have been reported in the literature^{3,4}.

Dermoscopy can help in the diagnosis of lymphoproliferative and histiocytic diseases. To the best of our knowledge, only four studies have reported the dermoscopic features of cutaneous RDD to date (Table 1)⁵⁻⁸. Previous studies reporting dermoscopy of RDD have described similar dermoscopic features, such as yellowish area and surrounding branched vessels. In the view of dermoscopic and histopathologic correlation, a yellowish structureless area on dermoscopy is assumed to correspond to dense histiocytic accumulation. Surrounding blood vessels on dermoscopy may represent increased vasculature, which is required for the proliferation of histiocytes.

Dermoscopic features that require differentiation from cutaneous RDD have been reported. Sarcoidosis was reported to manifest translucent yellow to orange globular-like or structureless areas associated with linear vessels⁹. Orange-whitish central areas with surrounding looped vessels were reported in one case with NTM infection¹⁰. Serpentine vessels with salmon-colored background is suggested for cutaneous B-cell lymphoma. Fine focused telangiectasia on a yellow to orange background is typically described for lupus vulgaris^{11,12}. Orange-

Table 1. Literature review of cutaneous Rosai-Dorfman disease with the dermoscopic features

Literature (year)	Sex/age in years	Location	Clinical finding	Initial misdiagnosis	Dermoscopic finding
Rodriguez-Blanco et al. ⁸ (2009)	M/25	Left sole	Hemorrhagic hyperkeratotic border and a clean ulcer bed	Viral wart	Light red background with milky-white irregularly distributed globular structures of different sizes
Aviles-Izquierdo et al. ⁵ (2012)	M/29	Chest	Erythematous-to-orange plaque with well-demarcated border	Atypical xanthogranuloma	Yellow homogeneous areas surrounded by large telangiectatic vessels on a red-orange background
Wang et al. ⁶ (2014)	F/41	Right cheek	Non-tender, smooth, erythematous papules, and nodules	-	Yellow discoloration on an erythematous background with branched vessels that were larger at the periphery and became smaller as they reached the center of the yellowish foci
Yang et al. ⁷ (2021)	M/53	Whole body	Numerous asymptomatic dome-shaped, firm papules, and nodules	-	Scattered yellow homogenous areas and surrounding branched blood vessels
Present case	F/47	Face	Multiple erythematous plaques with yellowish hue	Nontuberculous mycobacterial infection	Yellowish structureless patterns on an erythematous background with linear branched vessels

M: male, F: female.

yellow structureless pattern with erythematous border, named “setting sun sign”, represents the most frequent dermoscopic manifestation of juvenile xanthogranuloma¹³. However, considering the limited data available in the literature, further studies are necessary to characterize the patterns of various skin diseases to investigate the diagnostic accuracy of dermoscopy.

There are various treatment modalities for cutaneous RDD including excision, oral or intralesional steroids, chemotherapy, radiation, and cryotherapy^{1,14,15}. Due to the absence of randomized controlled trials, treatment guideline was not fully established. Therefore, treatment should be tailored to the individual clinical circumstances¹. The treatments have varying efficacy with uncertain treatment duration. Targeted therapy with a mitogen-activated protein kinase inhibitor can be tried in refractory cases with *KRAS*-mutated RDD¹⁶. A recent review article demonstrated that excision was shown to be the most successful option with 59% of the cases achieving complete remission¹⁴. However, surgery is not easy for patients with RDD due to the presence of multiple and often large lesions. It is worth noting that observation can be a reasonable approach as studies have shown that 20% to 50% of patients with nodal/cutaneous disease experience spontaneous remission¹. In our case, we engaged in a thorough discussion with the patient regarding various treatment modalities and opted for meticulous surgical excision, which yielded a satisfactory outcome considering the disfiguring features on the face.

In conclusion, we report a case of facial cutaneous RDD which was treated with surgery. Dermoscopy can be a useful adjuvant tool in the diagnosis of cutaneous RDD.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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