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Nada Bennouna
Department of Dermatology and Venereology, University Hospital of Ibn Rochd, Casablanca, Morocco

Kenza Baline
Department of Dermatology and Venereology, University Hospital of Ibn Rochd, Casablanca, Morocco

Fouzia Hali
Department of Dermatology and Venereology, University Hospital of Ibn Rochd, Casablanca, Morocco

Asmaa Elkebir
Department of Anatomical Pathology, University Hospital of Ibn Rochd, Casablanca, Morocco

Farida Marnissi
Department of Anatomical Pathology, University Hospital of Ibn Rochd, Casablanca, Morocco

Soumiya Chiheb
Department of Dermatology and Venereology, University Hospital of Ibn Rochd, Casablanca, Morocco

Corresponding Author:
Nada Bennouna
Department of Dermatology and Venereology, University Hospital of Ibn Rochd, Casablanca, Morocco

Hairpin vessels in purely cutaneous Rosai-Dorfman disease of the face: A case report

Nada Bennouna, Kenza Baline, Fouzia Hali, Asmaa Elkebir, Farida Marnissi and Soumiya Chiheb

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Abstract
Rosai-Dorfman disease (RDD) is a benign non-Langerhansian histiocytosis mainly affecting lymph node areas. Although cutaneous involvement in RDD is common, purely cutaneous disease is rare and poorly documented. We report the case of a 25-year-old female patient presenting with a 6-month history of a slow-growing asymptomatic lesion of the right cheek mimicking a cutaneous sarcoidosis. The aim of this article is to describe this rare entity with the presentation of a new dermoscopic finding: the hairpin vessels. We also highlight the differential diagnosis with sarcoidosis.

Keywords: Rosai-Dorfman, cutaneous disease, hairpin, patient, skin

Introduction
Rosai-Dorfman disease (RDD) is a benign non-Langerhansian histiocytosis mainly affecting lymph node areas, described by Rosai and Dorfmann in 1969 [1]. Purely cutaneous RDD (CRDD) is an extremely rare form of RDD [2], with only a few cases reported in the literature. The diagnosis is based on anatomopathological and immunohistochemical features. Studies reporting the dermoscopic features of MRD are limited [3]. Our work aimed to present a case of purely CRDD, to describe a new dermoscopic finding of this disease, and to highlight the differential diagnosis with sarcoidosis.

Case report
We report the case of a 25-year-old female patient presenting with a 6-month history of a slow-growing asymptomatic lesion of the right cheek. The patient never complained of fever, cough, weight loss, or fatigue. Physical examination revealed yellowish, erythematous papules and nodules (Figure 1, panel A). The patient had no palpable lymph nodes. Dermoscopy showed scattered multiple orange-yellowish areas, whitish structures with yellowish follicular keratotic plugs, surrounded by radial arborizing dendritic vessels, and also radial hairpin vessels on a red-orange background (Figure 1, panel B). Additional tests comprising complete blood count, biochemistry, C-reactive protein, angiotensin-converting enzyme, protein electrophoresis, complement, antinuclear antibodies, syphilis serology, hepatitis and human immunodeficiency virus, and chest radiography, were normal or negative. Histopathologic examination revealed emperipolesis (Figure 3, panels A and B), and immunohistochemistry was positive for CD68 (Figure 3, panel C) and S-100 protein (Figure 4, panel D), and negative for CD1a. Microbiologic studies were negative for fungi, Mycobacterium tuberculosis, atypical mycobacteria, and Leishmania spp. The diagnosis of RDD was made on the basis of clinical, histological and immunohistochemical findings. The patient was put on oral corticosteroid therapy with a dose of 0.5 mg/kg associated with methotrexate with a dose of 15 mg/week. Clear improvement was observed at 3 months (Figure 1, panel C).

Discussion
RDD is a rare benign non-Langerhans histiocytosis of unknown etiology. Isolated cutaneous localization is very rare, accounting for 3% of the cases, with a diagnostic delay up to 5 years [4].

It typically presents with a massive lymphadenopathy or febrile bilateral cervical polyadenopathy. There are 3 main clinical presentations of the pure cutaneous form: 1. the papulonodular form which is the most frequent, located usually on the face or the upper body, which was the case of our patient; 2. the indurated plaque form; 3. the tumoral form. The confirmation of the diagnosis is based on the histological finding of emperipolesis, associated with CD68+ and PS100+ immunostaining, and the negativity of CD1a. The clinical forms make clinical diagnosis of CRDD difficult to assess, despite the distinctive histological features. In our patient, the angiolupoid presentation and the dermoscopic aspect were highly suggestive of a cutaneous form of sarcoidosis. Indeed, all the dermoscopic signs found can be seen in sarcoidosis, especially the hairpin aspect [5]. However, the anatomopathological and immunohistochemical examinations confirmed the diagnosis of RDD. The dermoscopic signs described so far in RDD are red-orange background, yellowish ovoid structures, whitish structures, yellowish follicular keratotic plugs, linear, irregular and arboreal vessels [6]. All of these signs were found in our patient, in addition to the the hairpin vessels; which have never been described before in this entity. Primary cutaneous B-cell lymphoma is the main clinical and histological differential diagnosis [7]. The treatment of RDD is not codified, with many different therapeutic options: abstention, surgery, oral or intra-lesional corticosteroid therapy, methotrexate, as well as thalidomide, sirolimus and radiotherapy, or treatments modeled on those of other histiocytosis (interferon-α and cladribine) [8]. The treatment in our patient consisted in corticosteroids and methotrexate for economic and treatment availability reasons.

Figure legends

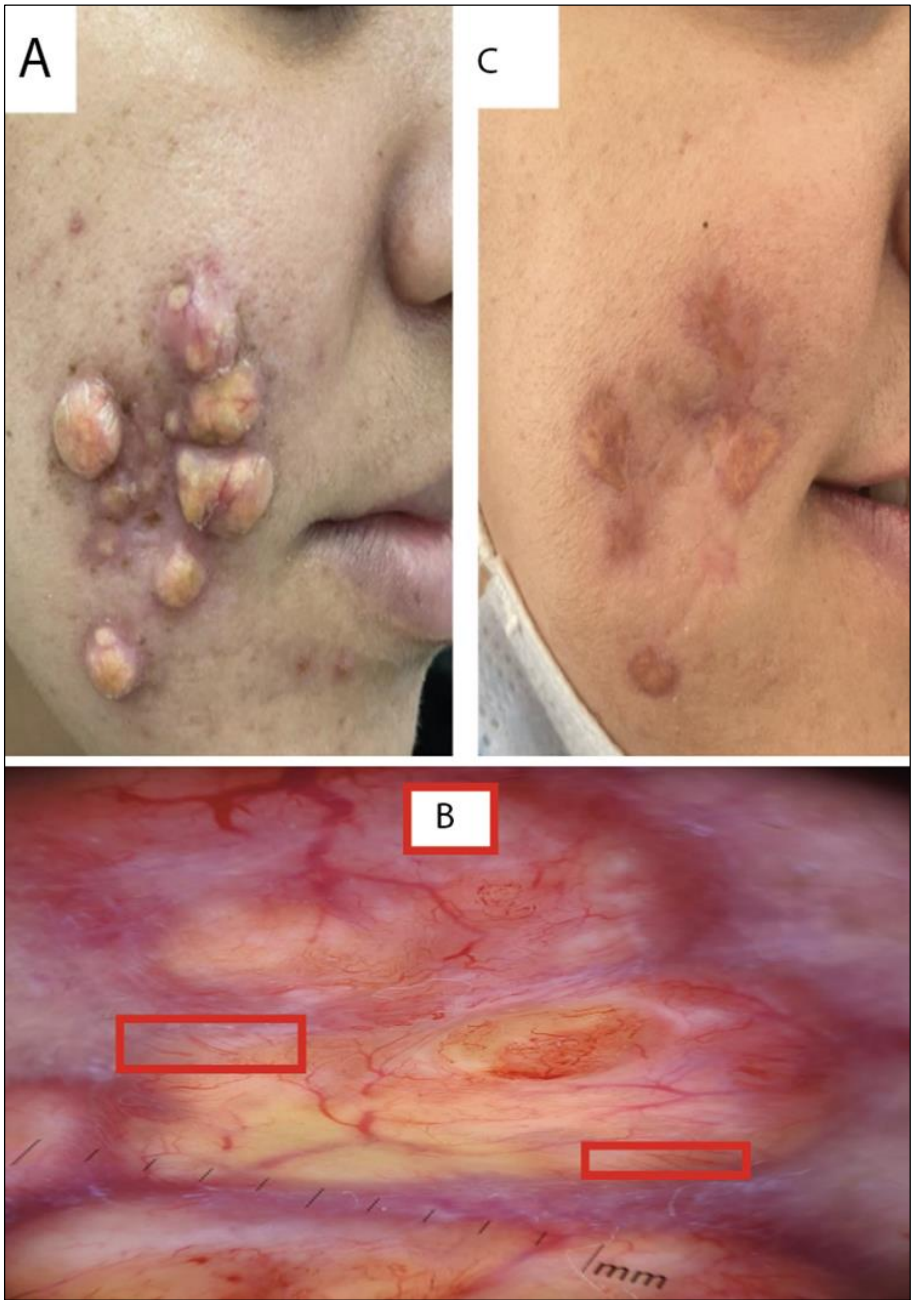

Fig 1: Yellowish, erythematous papules and nodules in the right cheek (panel A). Dermoscopy of the right cheek lesion showing scattered multiple orange-yellowish areas, whitish structures, surrounded by radial arborizing dendritic vessels, and radial hairpin vessels on a red-orange background (panel B). Clinical evolution after 3 months of treatment (panel C)

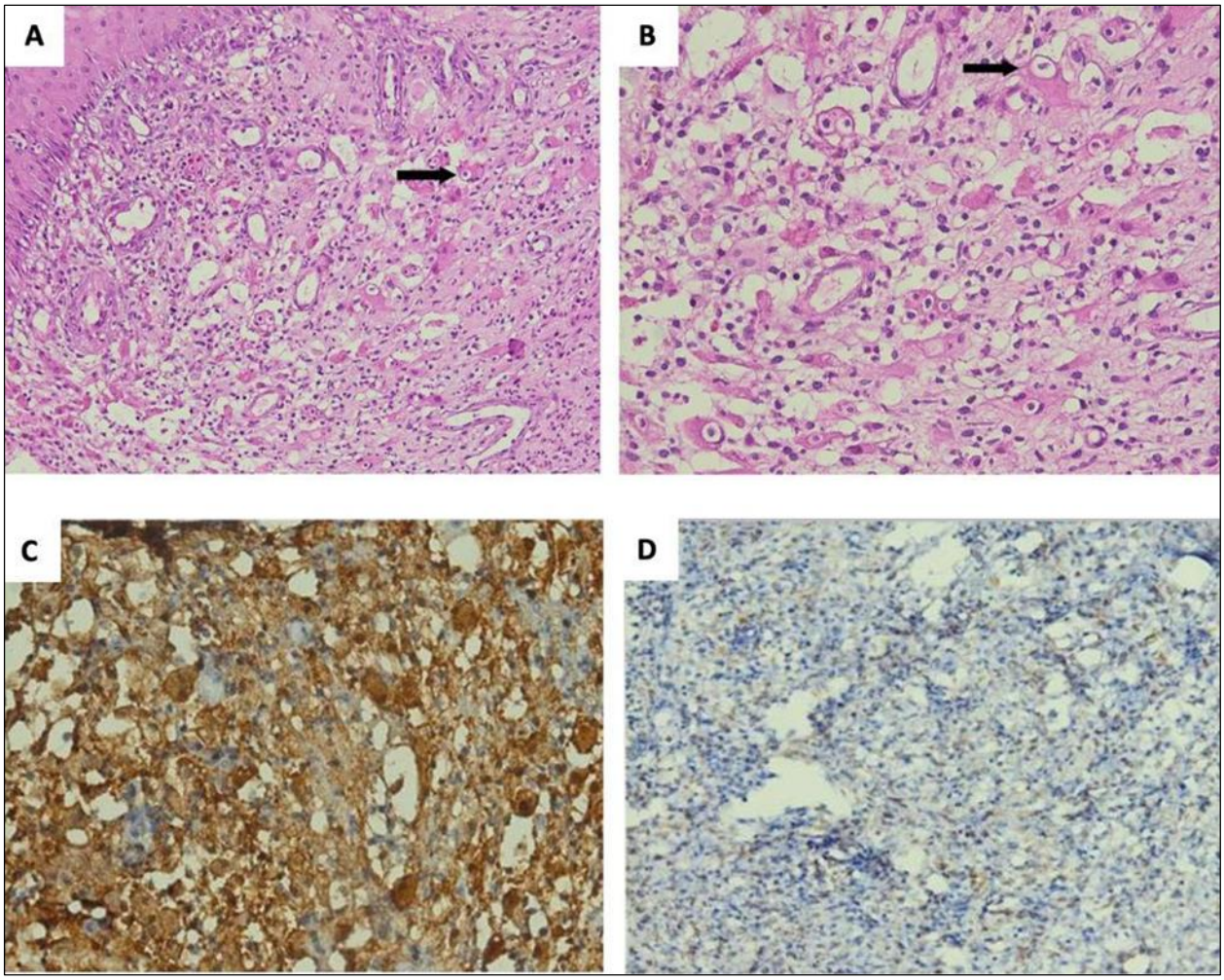

Fig 2: Histopathologic findings showing emperipolesis (arrows) in hematoxylin-eosin stain, x20 (Panel A) and x40 (Panel B) magnifications, and positive immunohistochemistry for CD68 (Panel C, x20 magnification) and S-100 protein (Panel D, x10 magnification)

Conclusion

As a conclusion, this case report sheds the light on a new dermoscopic sign of RDD, which is the presence of hairpin vessels. Furthermore, it is important to take into account the various dermoscopic similarities with cutaneous sarcoidosis. RDD is an entity to be considered in the differential diagnosis of granulomatous facial lesions, highlighting the complementarity of the clinical, dermoscopic, histological and immunohistochemical examinations in the presence of a pure cutaneous form of the disease.

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