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## Cutaneous Rosai-Dorfman disease: a case report of disease limited only to skin

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### Abstract

Rosai-Dorfman disease (RDD) is a disorder of histiocytic proliferation that can affect almost every organ. Skin may be involved in about 10% of cases, but only 3% of patients have skin lesions exclusively. Patients with cutaneous Rosai-Dorfman disease (CRDD) are usually healthy, without leading constitutional or other symptoms. The etiology of this disease is unclear, but it might be triggered by an infectious agent such as herpesvirus 6 (HHV-6) and Epstein-Barr virus (EBV). There is also a predisposition to autoimmune diseases and hematological malignancies. There is also a recognized predisposition to autoimmune disorders and hematologic malignancies. In our case, the patient was diagnosed with an infection due to *Borrelia burgdorferi*. To our knowledge, no previously reported association between *B. burgdorferi* infection and RDD has been described. The disease may manifest in any age group. The differential diagnosis includes Langerhans cell histiocytosis, metastasis, Hodgkin's lymphoma, and other diseases. Disease usually resolves spontaneously over a period of months to years. Treatment should be administered in non-responsive or multiple relapsing cases. Also, it is very important to take into consideration the patient's expectations and aesthetic appearance before making a treatment decision. Various treatment options are described, including surgical excision, cryotherapy, corticosteroids, antibiotics, methotrexate, and dapsons. In our case, we used CO<sub>2</sub> laser treatment successfully. To our knowledge, this is only the second case where CRDD lesions were treated successfully with CO<sub>2</sub> laser.

**Key words:** Rosai-Dorfman disease; cutaneous; case report.

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### Introduction

Rosai-Dorfman disease (RDD) is a rare benign disorder of histiocytic proliferation, also known as sinus histiocytosis with massive lymphadenopathy, first described in 1969.<sup>1</sup> Extranodal involvement may be seen in 43% of all cases. The disease can affect almost every organ, including the skin, kidneys, gastrointestinal tract, nervous system, and others. Skin may be involved in about 10% of cases, but only 3% of patients have skin lesions exclusively (without nodal involvement).<sup>2</sup> Etiology of this disease is unclear, but it might be triggered by an infectious agent such as herpesvirus 6 (HHV-6) and Epstein-Barr virus (EBV). There is also a predisposition to autoimmune diseases and hematological malignancies.<sup>3-5</sup> The disease may manifest in any age group, but more often in younger age groups.<sup>6</sup> The differential diagnosis includes Langerhans cell histiocytosis, Erdheim-Chester disease, melanoma, metastasis, Hodgkin's lymphoma, xanthomas, histoplasmosis, leishmaniasis, sarcoidosis, and other diseases.<sup>7,8</sup> We present a case of cutaneous Rosai-Dorfman disease (CRDD) that manifested in an older patient.

### Case Report

A 69-year-old male presented to our Dermatology Centre with a 5-month history of erythematous nodules on the skin. No pain or itching was reported. Dermatological examination revealed multiple 0.5 to 1.5 cm purple, firm nodules and papules located on the patient's face, legs, and buttocks (Figure 1). General physical examination was unremarkable. No lymphadenopathy or any systemic symptoms were observed. He did not have any relevant medical history and was not taking any medications. Laboratory investigations, including complete blood count, serum electrolytes, renal function, thyroid, liver function test, and glucose, were within normal ranges. Conventional chest radiography and abdominal ultrasound were unremarkable. Additional examinations revealed positive IgG and IgM antibody titers against *Borrelia burgdorferi*. He denied any recent tick bite. The patient was treated with amoxicillin 500 mg TID for one month. During the treatment, the lesions slowly regressed, but after discontinuation of the treatment, the regression stopped. Computed tomography of the head, chest, and abdomen had no abnormal findings. Histological examination of the skin lesion showed infiltration in the papillary and reticular dermis, consisting of medium-sized, abundant pale cytoplasm histiocytes and their groups. Phagocytosed lymphocytes (emperipolesis) were visible (Figure 2). An abundance of lymphocytes and plasma cells was observed in the background. The immunohisto-

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chemical examination revealed positive S100 and CD68 staining in histiocytes (Figure 3).

Based on the clinical manifestation of the lesions and histopathological features, the diagnosis of CRDD was established. Due to aesthetic concerns and slow regression of the lesions, the patient was treated with dapsons 100 mg BID for two weeks, combined with a mometasone furoate cream 1 mg/g QD, which showed no effect. Therefore, the patient was administered methotrexate 20 mg once a week for three months and triamcinolone acetonide intralesional injections. As an additional treatment option, the patient received four CO<sub>2</sub> laser treatment sessions with a total regression of the lesions (Figure 4). After the CO<sub>2</sub> laser treatments, the patient discontinued treatment with methotrexate. No lesion recurrence is observed. Further follow-up of the patient is ongoing.

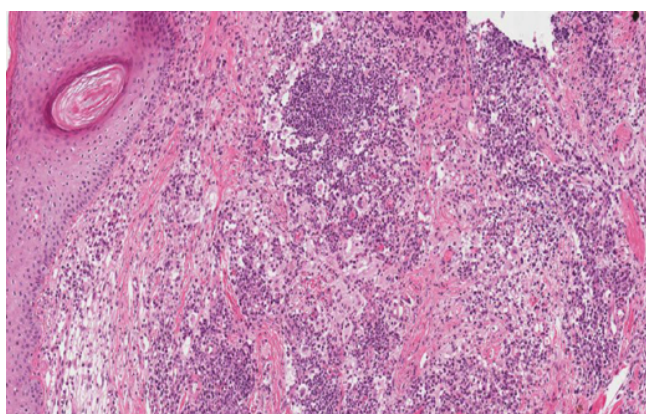
## Discussion

Systemic RDD typically affects children and young adults, with a predominance in males. In contrast, CRDD tends to present

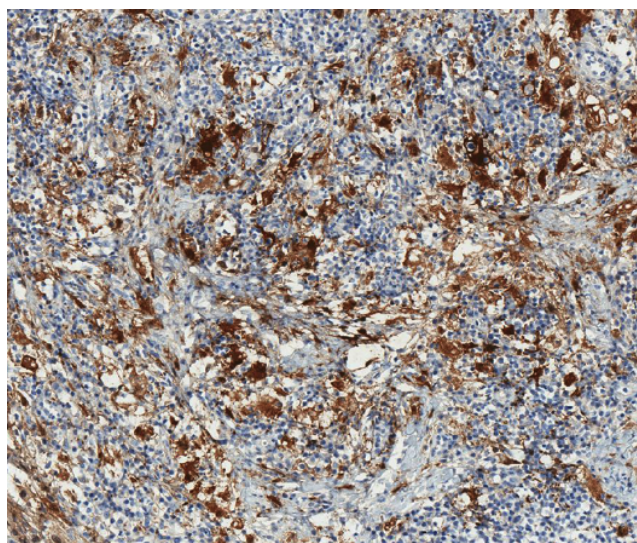
later in life and shows a higher prevalence among females. The Asian population is more frequently affected.<sup>3</sup> In our clinical case, an older Caucasian man was diagnosed with CRDD, which is uncharacteristic of the illness. Patients with CRDD usually are healthy, without leading constitutional or other symptoms. The etiology of this disorder is unclear. It is assumed to be a benign origin rather than a neoplastic condition caused by infection or cellular immune infringement.<sup>9</sup> Any known association between *B. burgdorferi* infection and any form of RDD has not been described before. Therefore, it might be just a concomitant illness, even though during the treatment with amoxicillin, the lesions slowly diminished. In most cases, lesions are asymptomatic nodules, plaques, or papules, varying from erythematous to brown, grey-blue, violaceous, or yellowish in color, usually slow-growing.<sup>10</sup> Any location can be affected, predominantly the extremities.<sup>11</sup> Diagnosis rests on distinctive histological features. The main finding is the presence of the characteristic foamy histiocytes. In addition, emperipolesis or lymphophagocytosis, the presence of intact inflammatory cells in the cytoplasm of histiocytes, is common. On immunohistochemistry, the histiocytes demonstrate positivity for S-100, CD4, and factor XIIIa; they may be either positive or neg-



**Figure 1.** The patient with erythematous nodules on the skin.



**Figure 2.** Histological examination of the skin lesion.



**Figure 3.** Immunohistochemical examination.



**Figure 4.** Patient after treatment.

ative for CD68 and are consistently negative for CD1a.<sup>12</sup> Most cases have a benign course, and treatment is not necessary. Disease usually resolves spontaneously over a period of months to years.<sup>13</sup> Treatment should be administered to patients with life-threatening disease (which is not characteristic of disease limited only to the skin) or in non-responsive or multiple relapsing cases. Also, it is very important to take into consideration the patient's expectations and aesthetic appearance before making a treatment decision. Various treatment options are described, including cryotherapy, radiotherapy, systemic, topical, or intralesional corticosteroids, antibiotics, interferon, chemotherapy, methotrexate, and dapsone.<sup>13,14</sup> The most effective treatment option if the lesion is solitary is a surgical excision. On the other hand, this treatment method is associated with a scar formation risk and lesion recurrence.<sup>15</sup> The treatment with CO<sub>2</sub> laser has some advantages compared to other treatment options for its safe ablation (high efficacy in ablating benign raised lesions), good hemostasis, low cost, and short recovery time.<sup>16</sup> The CO<sub>2</sub> laser treatment was effective in our case. To our knowledge, this is only the second case where CRDD lesions were treated successfully with CO<sub>2</sub> laser.

## Conclusions

RDD is a rare benign disorder of histiocytic proliferation that usually affects younger people but can occur at any age. The etiology of the disease is unclear. The majority of cases have a benign course and treatment is not necessary. Treatment should be administered to patients with life-threatening disease or in non-responsive or multiple relapsing cases. Also, it is very important to take into consideration the patient's wishes and aesthetic look before making a treatment decision.

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Consent for publication: the patient gave his written consent to use his personal data for the publication of this case report and any accompanying images.

Availability of data and materials: data and materials are available from the authors upon reasonable request.

Conference presentation: this case was presented in 27th EADV Congress in Paris, France, in 2018.

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