

# Multifocal Rosai-Dorfman disease with involvement of the pinna



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## INTRODUCTION

Rosai-Dorfman disease (RDD) is a benign non-Langerhans cell histiocytic disorder often involving the cervical lymph nodes. Extranodal disease was reported in 43% of patients with RDD in a review of the literature, with the skin reported to be the most common extranodal site.<sup>1</sup> In the absence of lymph node involvement, RDD limited to the skin is termed *cutaneous Rosai-Dorfman disease* (CRDD). To date, only 5 cases of CRDD involving the ear have been reported.<sup>2-6</sup> We present a case of multifocal CRDD with involvement of the right pinna.

## CASE REPORT

A 37-year-old Iraqi man presented with a 4-year history of an occasionally pruritic eruption on his legs and back. Over the 4 months before presentation, he noted drainage and swelling from his right ear. He previously treated the affected areas with clotrimazole 1% cream with poor response. He denied fevers, chills, night sweats, weight loss, or other constitutional symptoms. He had no significant medical or family history.

Physical examination of the right pinna found a soft, pink-to-violaceous plaque with a beaded appearance at the border (Fig 1, A). Several pink-to-brown, infiltrative plaques were present on the patient's back (Fig 1, B) and right posterior thigh. On the right anterior thigh and right lateral lower extremity were 3- to 5-mm pink, partially blanchable macules coalescing into a patch. There was no lymphadenopathy present on examination. No mucosal involvement was appreciated.

### Abbreviations used:

CRDD: cutaneous Rosai-Dorfman disease  
RDD: Rosai-Dorfman disease

A punch biopsy of the right posterior thigh from the initial visit found a mixed dense dermal inflammatory cell infiltrate, predominantly comprising plasma cells and histiocytes. Spirochete and acid-fast bacilli stains were negative. Rapid plasma reagin and HIV results were negative. The patient had a positive interferon- $\gamma$  release assay (T-spot), indicating positivity for mycobacterium tuberculosis. An excisional biopsy of the right middle back was performed for repeat histopathologic examination and tissue cultures. This specimen had large foamy histiocytes with emperipolesis and stained positively for S-100, CD68, and CD163 (Fig 2, A and B). CD1a stained a few Langerhans cells in the epidermis only. Bacterial, fungal, and acid-fast bacilli cultures were negative. Tissue culture using Novy-MacNeal-Nicolle medium to exclude leishmaniasis could not be performed, as the culture medium was not available at the time of biopsy. Based on the clinical presentation and pathology results, a diagnosis of RDD was made.

## DISCUSSION

To the best of our knowledge, only 5 cases of CRDD involving the ear have been reported in the literature.<sup>2-6</sup> In each of those cases, the ear was the only location on the skin involved. Cervical lymph nodes are typically involved, leading to painless,

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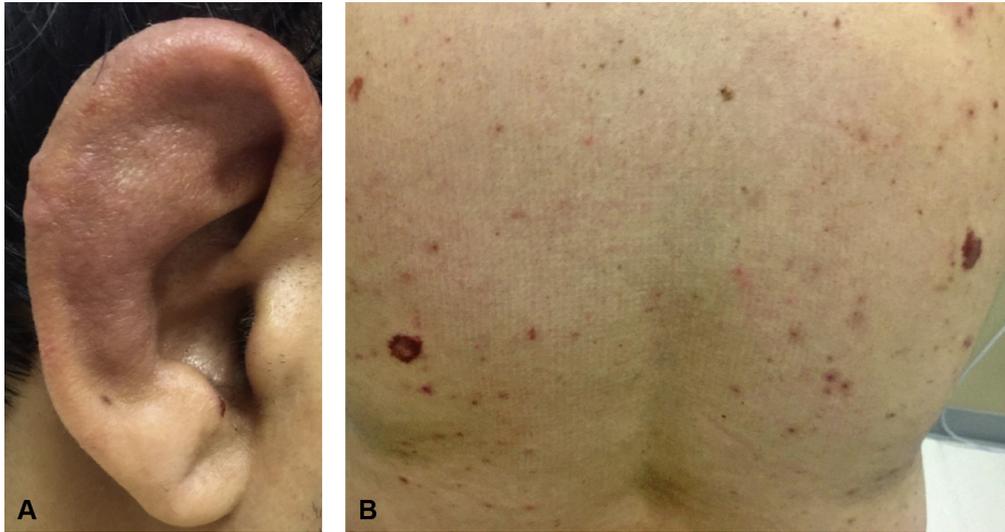
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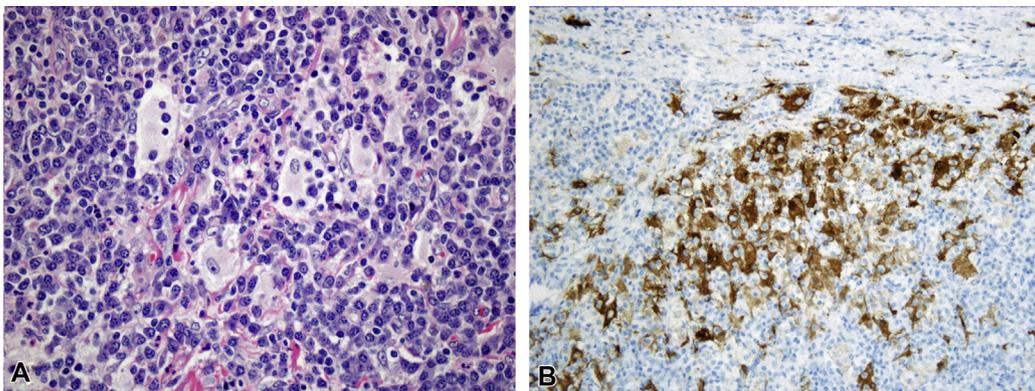
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**Fig 1.** **A**, Soft, pink-to-violaceous plaque with beaded appearance on the right pinna. **B**, Pink-to-brown infiltrative plaques on the back.



**Fig 2.** **A**, Hematoxylin-eosin staining shows an infiltrate of lymphocytes, plasma cells, and histiocytes that are large with pale, foamy cytoplasm and emperipolesis. **B**, Numerous cells staining positively for S-100.

bilateral cervical lymphadenopathy.<sup>2</sup> Associated findings often include fever, leukocytosis, elevated erythrocyte sedimentation rate, and polyclonal hypergammaglobulinemia. RDD may affect the lymph nodes and have internal organ involvement, or the disease may be limited exclusively to the skin, as in CRDD.<sup>7</sup> The nonspecific appearance of cutaneous lesions can lead to a broad differential diagnosis, including other histiocytoses, sarcoidosis, tuberculosis, leishmaniasis, syphilis, and other infectious and infiltrative etiologies. Safdar et al<sup>8</sup> reported a case of RDD that mirrored active spinal tuberculosis in a *Mycobacterium tuberculosis* hyperendemic region. The diagnosis of RDD was only made after performing immunohistochemistry on the tissue biopsy. Ear involvement is of particular significance because when present, it can help narrow the clinical and histologic differential diagnosis to include

infiltrative and inflammatory processes involving the ear such as leprosy, relapsing polychondritis, perichondritis, pseudocyst of the auricle, and lymphoproliferative disorders.

The histologic characteristics of RDD include an inflammatory infiltrate with numerous plasma cells and histiocytes, which have a large pale eosinophilic cytoplasm, vesicular nuclei, and positive staining for S-100 and CD68, but negative staining for CD1a.<sup>9</sup>

The exact etiology of RDD remains unknown, but it has been associated with human herpesvirus 6, lymphoma, Langerhans cell disease, autoimmune lymphoproliferative syndrome-1, Epstein-Barr virus, parvovirus B19, joint disease, asthma, juvenile-onset diabetes, HIV, and red blood cell autoantibodies.<sup>6,7,10</sup> Our patient did not report any history of these associated conditions.

In addition to involvement of the pinna, another unique feature of this case is that our patient is an Iraqi man. CRDD has been reported to have a 2:1 female predominance.<sup>7</sup> However, previous case reports of CRDD involving the ear were reported in 2 females and 3 males.<sup>2-6</sup> One case was in a 10-year-old African-American boy<sup>3</sup> and another a white man in his mid-20s,<sup>6</sup> whereas the other 3 case reports do not report patient ethnicity.

Numerous treatment options have been used for patients with CRDD including radiotherapy, corticosteroids (topical and systemic), chemotherapy, acyclovir, surgical excision, antibiotics, cryotherapy, interferon, dapsone, and retinoids.<sup>8</sup> Bunick et al<sup>5</sup> reported a case of CRDD involving the ear, which resulted in improved cosmetic appearance with localized radiation therapy. Most recently, Lin et al<sup>6</sup> also reported a case of CRDD on the ear, which was recalcitrant to antibiotics, antifungals, and topical, intralesional, and oral corticosteroids. Consequently, their patient elected to have a complete auriculectomy with prosthetic ear replacement.<sup>6</sup> Some cases of CRDD have been reported to resolve spontaneously. In this case, our patient was asymptomatic, and no treatment was initiated. He was referred to the hematology/oncology department for evaluation of systemic disease, but refused radiologic investigations.

CRDD is a rare clinical entity, with nonspecific cutaneous manifestations. The disease often follows a benign clinical course, with poorer prognosis associated with a heavy nodal burden and systemic involvement. The ideal treatment of CRDD has yet to be determined. We propose the development of a

national registry of patients for future meta-analyses to help gain a better understanding of the disease etiology, clinical features, and future treatment options.

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