# Cutaneous Rosai-Dorfman Disease of the Face: A Comprehensive Literature Review and Case Report

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**Purpose:** Cutaneous Rosai-Dorfman disease (C-RDD) is a rare, benign, non-Langerhans cell histiocytosis that can affect any skin area. The purpose of this study was to characterize C-RDD of the face through a literature review of published case reports of this lesion and present a new case of C-RDD.

**Materials and Methods:** An English-language search of 3 databases (PubMed, Scopus, and EBSCO Search) was conducted for cases of C-RDD of any skin area that had been published since 1969. Repeated citations of the same article in more than 1 database were deleted. Cases of C-RDD with solely facial involvement or involving the face and other skin parts were included and analyzed. Details of C-RDD cases (demographic features, regional distribution, clinical presentation, treatment, and follow-up course) were methodically reviewed and collected in Excel spreadsheets. Simple statistical analyses were conducted using Excel.

**Results:** The literature search yielded 578 published cases of C-RDD affecting any skin area; of these, 65 cases (11.2%) had facial skin involvement. The male-to-female ratio was 1:1.5, and the average age at presentation was 43.5 years (standard deviation, 12.4 yr). The racial distribution of facial C-RDD was, in descending order, 74.5% in Asians, 20% in Caucasians, and 5.5% in blacks. The most commonly affected facial skin regions were the cheeks and periorbital area, and most lesions were multiple in number and bilaterally distributed. The vast majority of facial C-RDD lesions presented as asymptomatic, nonulcerative, red, nodular plaques with durations ranging from 1 month to a few years. Many methods have been attempted for the treatment of facial C-RDD. However, the combined cure rate for all published treatment methods was only 28.6%. Surgical excision was the most effective treatment method, and corticosteroids were the least effective.

**Conclusion:** This article has tried to characterize facial C-RDD lesions for easier management by maxillofacial surgeons.



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Rosai-Dorfman disease (RDD), initially termed *sinus histiocytosis with massive lymphadenopathy*, is a benign, multiorgan, non-Langerhans cell histiocytosis that was described as a distinct entity by Rosai and Dorfman[1] in 1969. RDD typically consists of bilateral painless lymphadenopathy, fever, polyclonal hyperglobulinemia, high erythrocyte sedimentation rate (ESR), and immunologic dysfunction. It has been estimated that more than 40% of patients with RDD have extranodal involvement, especially the skin.[2] A purely cutaneous RDD (C-RDD) has been described, but is considered a rare occurrence.[3]

Histopathologically, RDD typically exhibits an inflammatory infiltrate with numerous plasma cells and a large number of histiocytic cells. The cytoplasm of histiocytes shows positive expression of S-100 and CD68 proteins, but is negative for CD1a.[4] Within the cytoplasm of intact histiocytes, other cells can be engulfed, including neutrophils and lymphocytes. This histopathologic finding is called *emperipolesis* and confirms the diagnosis of

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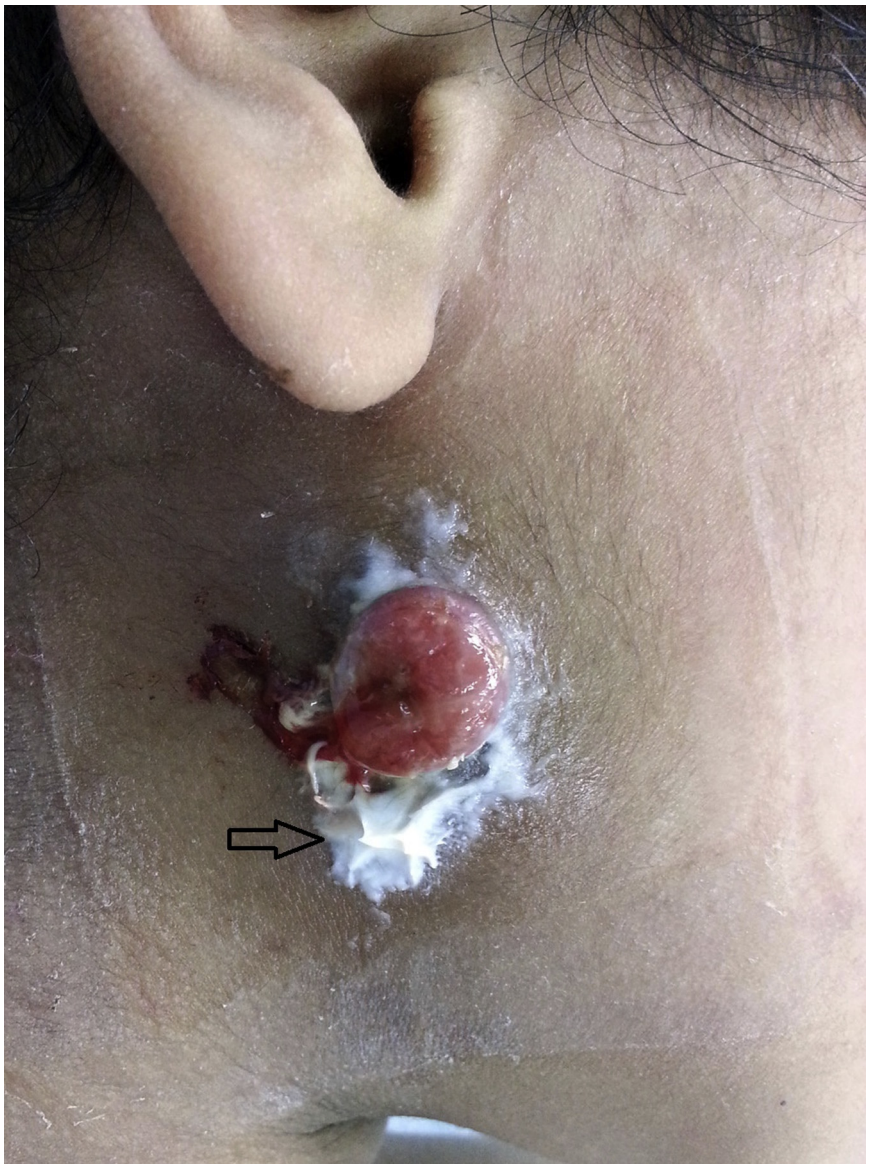

**FIGURE 1.** *A,* The mass at initial presentation (*arrow,* antibiotic cream). **(Fig 1 continued on next page.)**

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RDD. Emperipolesis differs from phagocytosis in that an engulfed cell survives within another cell and retains its intact normal structure; in phagocytosis, the engulfed cell is destroyed by lysosomal enzymes.[5] Electron microscopic studies have shown that histiocytes of RDD are devoid of Birbeck granules,[3] a feature that differentiates RDD from Langerhans histiocytosis.

Maxillofacial surgeons very rarely see RDD; in fact, RDD has been scarcely reported in the maxillofacial literature. An English-language search of 3 databases (see below) failed to find more than 11 reports of this disease affecting various organs of the maxillofacial area since 1969.[6-16] Pure C-RDD of the facial skin also is rare; the literature search yielded only 65 cases of facial C-RDD reported from different medical disciplines (see below). A case of facial C-RDD with its clinicopathologic features is described in the present article.

To characterize facial C-RDD and illustrate the clinical features that differentiate C-RDD from other benign and malignant lesions that can affect the facial skin, this article presents a comprehensive literature review of the 65 published cases of C-RDD of the facial skin.

## Report of Case

A 4-year-old boy presented to the maxillofacial unit at King Abdul Aziz Medical City (Riyadh, Saudi Arabia)