

## Case Report

# Isolated maxillary rosai dorfmann disease masquerading as a malignancy

Deepak Janardhan<sup>1\*</sup>, Bipin T. Varghese<sup>1</sup>, Sharath S.<sup>1</sup>, Anitha Mathews<sup>2</sup>

<sup>1</sup>Department of Head and Neck Surgical Oncology, Regional Cancer Centre, Trivandrum, Kerala, India

<sup>2</sup>Department of Pathology, Regional Cancer Centre, Trivandrum, Kerala, India

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### \*Correspondence:

Dr. Deepak Janardhan,

E-mail: [dr.deepakjanardhan@gmail.com](mailto:dr.deepakjanardhan@gmail.com)

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### ABSTRACT

Rosai Dorfmann disease (RDD), first described in 1969 by Rosai and Dorfmann, is a rare, benign disorder of unknown etiology. It is a distinct clinopathological entity, typically characterized by non Langerhan histiocytic cell proliferation with massive cervical lymphadenopathy. Axillary, mediastinal and inguinal nodes are affected rarely. Fever, anemia, leucocytosis, hypergamma globulinemia, raised ESR and rarely autoimmune hemolytic anemia may coexist; mimicking differentials like Wegener's granulomatosis, Langerhans cell histiocytosis, Tuberculosis, Hodgkin's lymphoma, monocytic leukaemia and rhinoscleromatosis. Hence, a high clinical suspicion with immunopathological analysis confirms the diagnosis, as in this case report.

**Keywords:** Rosai Dorfmann disease, Maxilla, Histiocytes

## INTRODUCTION

Rosai Dorfmann disease (RDD), is a rare, pseudolymphomatous disorder of unknown etiology, characterized by non Langerhan histiocytic cell proliferation with massive cervical lymphadenopathy.<sup>1</sup> It is a self-limiting disease with insidious course.<sup>3</sup> Young males are commonly affected. Extranodal involvement occurs in 25%.<sup>2</sup> S100 positive histiocytes with lymphophagocytosis, known as emperipolesis, is the characteristic histological finding. No specific treatment has been advocated for this condition.<sup>8</sup>

However long term follow up for local recurrence or other site involvement is considered beneficial for timely and appropriate treatment.

## CASE REPORT

A 43 year old female with no known comorbidities, was referred to our tertiary center, with pain and swelling over left malar region. On clinical examination, only a left

malar prominence was noted with normal blood parameters.

CT scan (Figure 1) revealed irregular destruction of left alveolar process of maxilla with adjacent heterogeneously enhancing soft tissue density extending to the left upper gingivobuccal sulcus and retromolar trigone, infratemporal fossa, masticator space with infiltration of the pterygoid muscles and temporalis muscle with obliteration of the retromaxillary fat and focal loss of fat plane with the masseter muscle. Anteriorly, the soft tissue density was found extending to the premaxillary fat. Focal destruction of the inferior aspect of the left posterolateral maxillary wall was noted. As the CT features were suggestive of malignancy, multiple biopsies were done. Histopathology revealed only inflammatory infiltrates with plasma cells, lymphocytes, histiocytes and macrophages and no obvious feature of malignancy was noted. With consent for wide excision of the lesion amounting to subtotal maxillectomy and possibility of negative histopathology report explained, the patient was taken up for surgery.

Intraoperatively, scarred and indurated lesion grossly adherent to the anterior and lateral wall of maxilla with extension into the floor of the sinus and bulge in left half of palate was noted. Subtotal maxillectomy with removal of pterygoid plates and partial clearance of infratemporal fossa was done. Defect (Figure 2) was lined with split skin graft. Final histopathological analysis revealed S100 positivity (Figure 3), CD68 variable staining and Cytokeratin negativity with evidence of 'emperipolesis' (Figure 4). These features were consistent with a diagnosis of Rosai Dorfman disease. Patient was found to be doing well up to 6<sup>th</sup> month of follow up with a well mucosalised post-operative defect and advised to have prothodontic consultation for obturator usage.



**Figure 1: CT scan showing destruction of left alveolar process of maxilla with adjacent heterogeneous soft tissue extending to left upper gingivobuccal sulcus, retromolar trigone, with minimal infratemporal fossa and masticator space extension.**

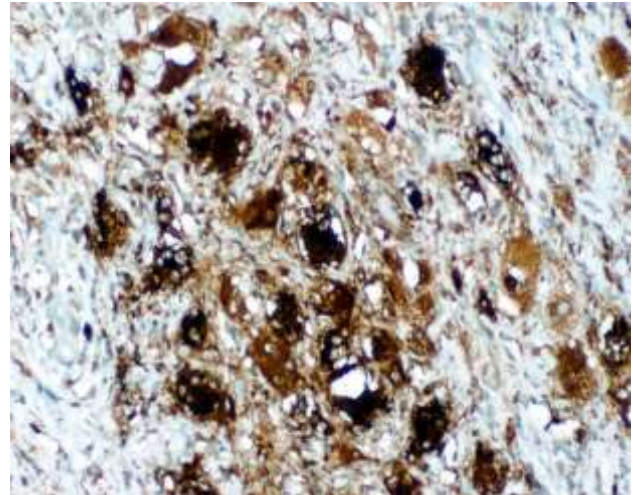


**Figure 2: Partial maxillectomy defect lined with split skin graft.**

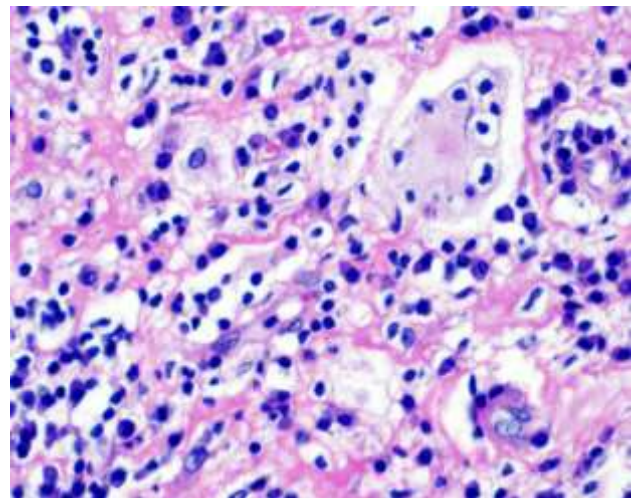
## DISCUSSION

Rosai-Dorfman disease or Sinus histiocytosis with massive lymphadenopathy (SHML) is a rare, benign,

self-limiting disease, wherein accumulation of histiocytes occurs in lymph nodes throughout the body, most commonly in the neck. The etiology of RDD is largely unknown, but it has been attributed to genetic alterations or due to aberrant response to infective agents like EBV, HHV6, or cytokines.<sup>3</sup>



**Figure 3: Rosai – Dorfman disease: strong expression of s100 protein by the histiocytes.**



**Figure 4: Rosai – Dorfman disease: Histiocyte with emperipolesis in a background of lymphocytes and plasma cells.**

Although involvement of neck nodes are the most common sign, atypical presentations, with the absence of nodal involvement can occur, making the diagnosis difficult. Extra nodal involvement of at least one site occurs in 43% of RDD cases and 23% have only isolated extra nodal disease involving skin, respiratory tract, bone, genitourinary system, oral cavity, central nervous system, orbit, salivary gland, tonsil, breast, viscera, heart and rarely bone marrow 75% of extra nodal forms are in head and neck region with sinonasal area being the commonest.<sup>2,3</sup>

Sinonasal RDD has a predilection for young adult men and may present with nasal obstruction, epistaxis, facial pain, hyposmia or facial swelling. It may be accompanied by fever, neutrophilic leucocytosis, and polyclonal hypergammaglobulinemia.

Most patients with RDD have spontaneous remission, but in few it can persist or recur. Very rarely, the disease follows an aggressive course, ending fatally<sup>3</sup>. The involvement of kidney, lower respiratory tract, or liver is considered to be of poor prognosis.<sup>1</sup> It may be associated with autoimmune disorders and hematopoietic malignancies.<sup>4,7</sup>

The diverse clinical picture mimics numerous differentials. Although clinicoradiological correlation at presentation is helpful, histopathologic assessment alone is confirmatory. The classic histology is characterized by effacement of nodal architecture and dilatation of lymph node sinuses by lymphocytes, plasma cells and numerous histiocytes. These histiocytes contain intact lymphocytes, and plasma cells in their cytoplasm. This process, where cells enter another cell and evade cellular degradation is known as 'Emperipolesis'. This was first described by Humble et al.<sup>8</sup> When extra nodal sites are involved, emperipolesis is less prominent.<sup>3</sup> Immunohistochemical stains useful for diagnosing RDD are S100 (+), CD68 (+) and CD1a (-). Rarely RDD can progress to malignant lymphoma or amyloidosis.

Although various treatment modalities such as surgery followed by radiation, steroids, interferon and chemotherapy have been attempted, till date, no definite treatment has been established. This is probably due to the peculiar oddity, self-limiting and benign nature of RDD.

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