



CASE REPORT

Rosai-Dorfman disease as a differential diagnosis of cervical adenopathy

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Abstract

Introduction: Rosai-Dorfman disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy (SHML), is a rare benign idiopathic disease that affects both nodal and extranodal sites. **Case report:** A female patient aged 27 years with right side cervical adenopathy of levels IV and V, both mobile, painless and of fibroelastic consistency, with no signs of inflammation was evaluated. Histopathology of the lymph node biopsy showed preserved lymphoid architecture and presence of numerous mature histiocytes compatible with RDD.

Keywords: histiocytosis; emperipolesis; hemophagocytic lymphohistiocytosis.

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Introduction

Rosai-Dorfman disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy (SHML), it is a rare condition that affects all age groups, including pediatric patients. It is a non-Langerhans cell histiocytosis most commonly presented as painless cervical lymphadenopathy, although extranodal manifestation has also been observed in at least one third of patients, with most evident involvement of the head and neck region¹.

This disease is usually self-limiting, but can sometimes be life-threatening¹. The factors that influence disease recurrence are still not well understood; however, hypotheses have been raised that this condition may result from changes in immune response and infections caused by certain viral and bacterial pathogens, such as Varicella Zoster Virus (VZV), Epstein-Barr Virus (EBV), Cytomegalovirus (CMV), *Brucella*, *Klebsiella*, among others².

Case report

A 27-year-old Caucasian female patient sought emergency care with a history of cervical mass for a month. She reported tiredness, hyporexia and vomiting, and had an axillar temperature of 39°C. She also reported having

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The study carried out at the Universidade Metropolitana de Santos, Santos, SP, Brazil.



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lower limb pain, diffuse arthralgia, loss of appetite, and chills with night sweats for the past six months. Right side of the level IV cervical (1.5 cm) and supraclavicular (1 cm) lymph nodes were evidenced, both mobile, painless and of fibroelastic consistency, with no signs of inflammation. No lesions in the upper aerodigestive tract were observed at the locoregional examination. Laboratory tests showed no changes and serology was negative for mono-like diseases. No suspicious lesions were found at the locoregional examination. Cervical lymphadenectomy presented chronic lymphadenitis with intense sinus histiocytosis suggestive of RDD, with subsequent diagnostic confirmation by immunohistochemistry (IHC), which showed lymphoid architecture and presence of numerous mature histiocytes – Figures 1 to 3. As the patient did not present any systemic manifestations after treatment with corticosteroids (Prednisone 30 mg/day for 4 weeks), only surgical excision of the right supraclavicular lymph node was performed, without complications.

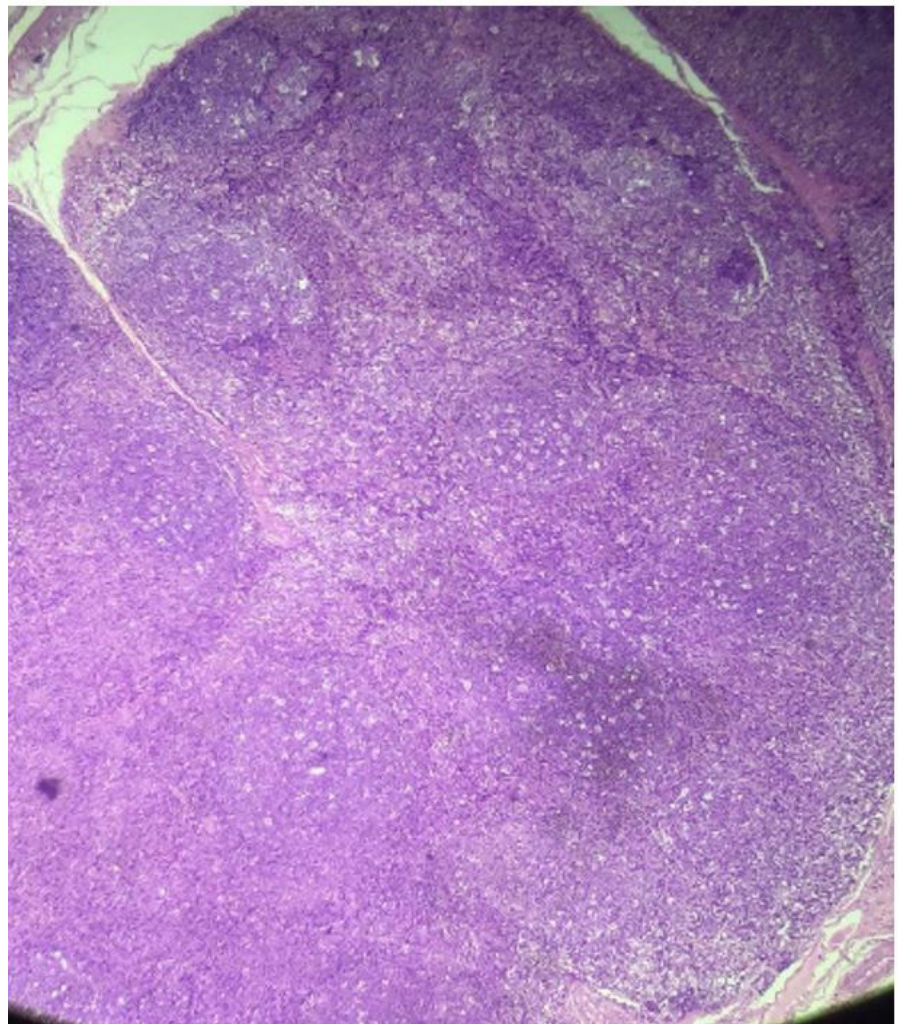


Figure 1. Photomicrography showing reactionary lymphoid population with the presence of several immunoblasts (red circle), HE, 100x.

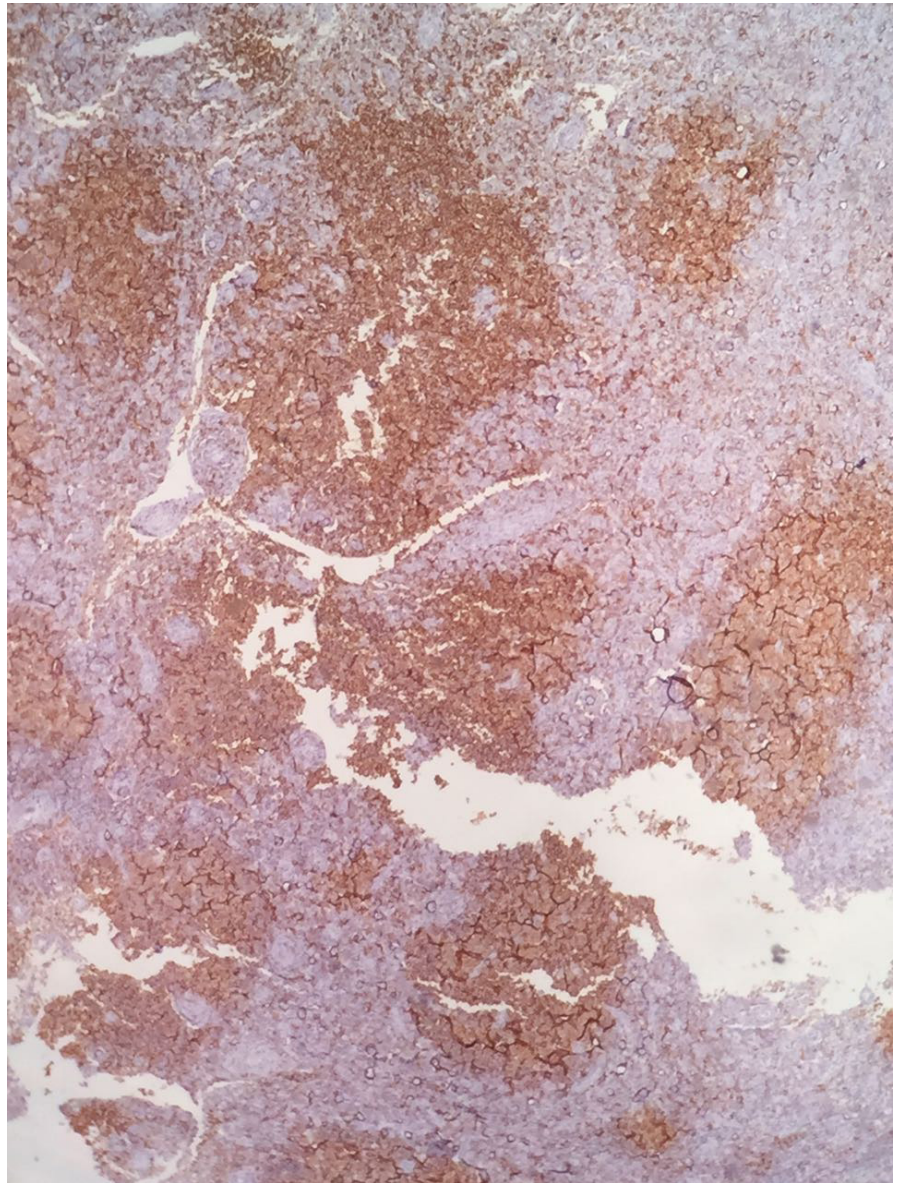


Figure 2. Photomicrography showing B lymphocytes in the germinative center; CD20+ immunohistochemistry. X100.

Discussion

Lymphadenectomy is rarely caused by RDD in children and young adults. Since RDD was first described in 1969, several cases of nodal and extranodal involvement have been reported². RDD has two main forms of presentation: the first form affects the lymph nodes (SHML), with rare systemic manifestation in other organs; the second one involves only the skin, without any systemic or nodal disease even after long-term follow-up. These two forms are considered as different clinical entities².

Extranodal involvement has been observed in 25-43% of cases, whereas focal lymph node involvement has been found in 57% of cases. Painless

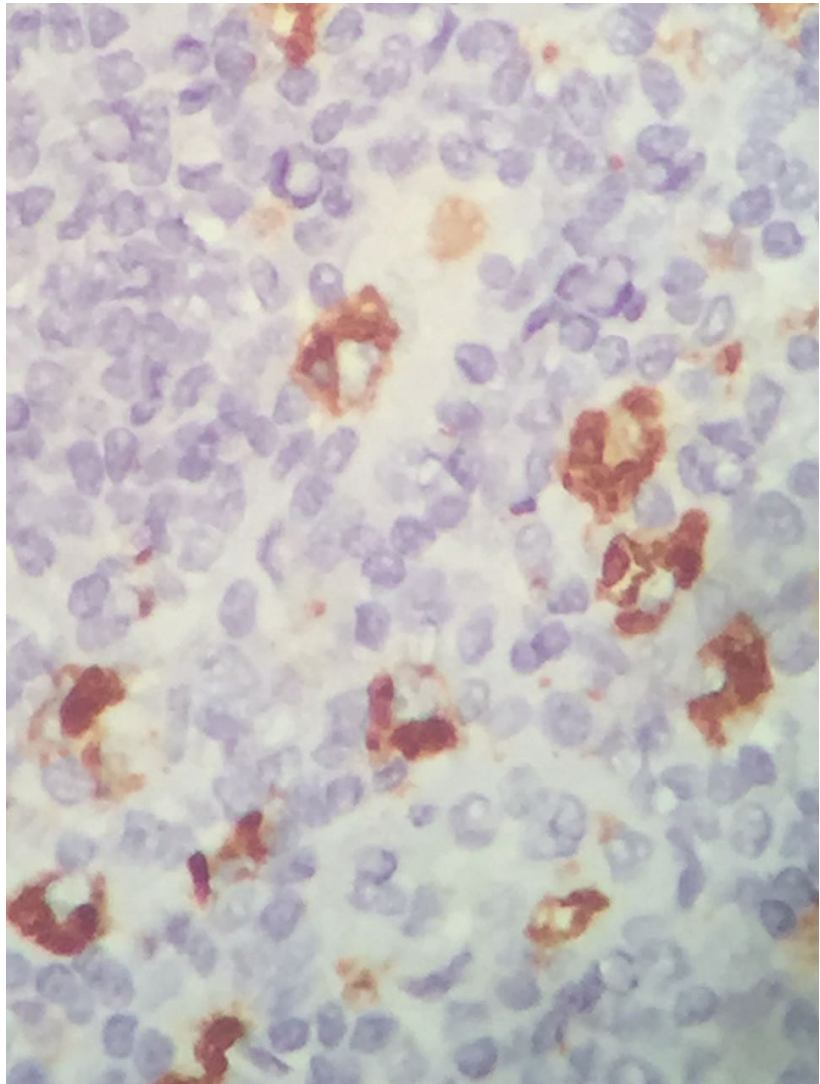


Figure 3. Photomicrography showing B macrophages with possible lymphocytes phagocytosis (emperipolesis); CD68+ immunohistochemistry, x400.

cervical lymphadenopathy is the most frequent initial symptom of this disease, reported by nearly 90% of patients². In this patient, it was possible to palpate an enlarged lymph node from the cervical supraclavicular and internal jugular regions to the clavicle. Apparently, there is no relationship between the nodal disease and the extranodal sites, which can occur separately. The RDD etiology is uncertain, although infectious agents such as the EBV and VZV are important in its pathogenesis². Typically, the disease occurs with insidious onset, prolonged active phase, and eventual spontaneous remission with subsequent recurrences³.

Imaging examinations can be useful in assessing the extent of RDD and histopathology is necessary to confirm its diagnosis. The presence of cytologic atypia and the aggressive clinical course of the disease establish the diagnosis in most cases³. S-100 protein positivity a differential diagnostic aspect¹.

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Differential diagnosis includes several lymphoreticular malignancies such as lymphoma, Hodgkin's disease, histiocytic neoplasms and monocytic leukemia, which present similar histopathological characteristics.

Because SHML is considered an uncommon and self-limiting disease, there is no ideal protocol for its treatment. Half of the cases are resolved spontaneously. In patients with high fever alone, corticosteroid therapy may be indicated⁴. Surgical debulking is necessary in cases where lymph node enlargement induces important symptoms, such as when the airways are obstructed or vital organs are compressed. Radical surgery is not usually indicated considering the typically benign course of RDD⁵.

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