Journal of Cytology & Histology

Leite, et al., J Cytol Histol 2016, 7:3
DOI: 10.4172/2157-7099.1000427

Case Report OMICS International

Rosai-Dorfman Disease Involving Thyroid and Parotid Glands in Male with 48-Year-Old

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Received date: December 14, 2015; **Accepted date:** July 10, 2016; **Published date:** July 20, 2016

Abstract

Rosai-Dorfman disease (RDD) is a rare and self-limiting non-Langherhans histiocytosis with lymph node and cutaneous manifestations. This sinus histiocytosis is very extremely usual in children that progress to massive lymphadenopathy. Emperipolesis is a common Histopathological findings and protein S-100 is positive in most cases of RDD. Here, report a case of a 48-year-old with adult with RDD that exhibited cervical lymphadenopathy without fever or Osteolyticlesions. Histopathological and immunohistochemistry studies of lymph node showed emperipolesis, S-100 protein, CD68 positive and macrophages negative for CD1a. After that the diagnosis of RDD was done. However, cervical lymphadenopathy was not solved spontaneously, and the treatment with corticosteroid therapy was initiated and the follow-up showed a clinical improvement condition of the patient.

Keywords: Rosai-Dorfman disease; Sinus histiocytosis; Involvement of glands; Adults

Introduction

Rosai-Dorfman disease (RDD) is a rare entity of non-Langerhans histiocytic disorder described by Rosai and Dorfman in 1969 and usually present a benign course [1]. This sinus histiocytosis with massive lymphadenopathy remains such as an idiopathic disease. Some evidences suggests that immunological disorders and viral infections such as human herpes virus (HHV), parvovirus B19 and Epstein - Barr virus (EBV) may be involved in the genesis of RDD.4 Rosai-Dorfman disease are frequently affect children, teenagers, but it's a uncommon disease after 20 years-old [2,3].

This sinus histiocytosis may be divided in two forms: cutaneous indolent disease and a nodal form with or without systemic symptoms. In most cases, this histiocytosis presents a self-limited course with or without treatment. The RDD is commonly characterized by massive lymphadenopathy with painless, fever, night sweats, weight loss, leukocytosis, increase of erythrocyte sedimentation and polyclonal hypergammaglobulinemia [4]. The involvement of extra nodal sites are most commonly in skin, bones, and soft tissue and it's seen in 40% of the cases and the involvement of thyroid and parotid glands represents about 25% of all cases of RDD [5,6].

For diagnosis of RDD, a lymph node biopsy, histopathological and immune histochemical examination are essential. The architecture of the lymph node presents modified sinusoidal dilatation containing a large number of histiocytes, lymphocytes and plasma cells. Emperipolesis is a classic finding of DRD, characterized by numerous histiocytes that phagocytose intacts lymphocytes within their cytoplasm. The pattern of the immunohistochemical in RDD is positive for S-100, CD68 (KP-1) and CD16 protein and the absence of CD1a [7,8].

Several patients with DRD do not require treatment, but some cases may be recommended received steroid therapy with prednisolone, surgical resection or more intensive therapy may be necessary in a patient that shows massive and refractory lymph node that can result in organ compression. In the situations, chemotherapy regimens are required and some case exhibits a good response to cladiribine and imatinib. Radiotherapy has been reported to be useful in selected patients [9,10].

The purpose of this study was described an atypical presentation of DRD involving the thyroid and parotid glands and without fever and osteolytic lesion in a 48-year old male.

Case Report

48-years-old man was admitted in a Instituto de Medicina Integral Professor Fernando Figueira, IMIP, Recife, Brazil presented a small with complaint of painless unilateral lymph node enlargement in the neck without fever. He denied weight loss, bone pain, night sweats, history of cancer, diabetes, hypertension or infectious diseases. Physical examination revealed cervical lymphadenopathy by palpation (> 2 cm) and ultrasound examination shows hepatomegaly,

involvement of the parotid glands and thyroid. Routine laboratory investigations, including a complete blood count, Erythrocyte sedimentation rate (ESR) and serum protein electrophoresis, were made. Values obtained were WBC: 6.9x109/L, hemoglobin, 13.1 g/dL, platelet counts, 98x109/L, Lactate dehydrogenase 181 U/L, glucose 96 mg/dL, Creatinine 0.6 mg/dL calcium 9.3 mg/dL erythrocyte sedimentation: 45 mm/h, albumin 4.1 g/L and protein electrophoresis revealed polyclonal. The serological tests were negative for hepatitis B and C, HIV, HTLV, toxoplasmosis, syphilis and Epstein-Barr virus. Besides, the investigations for colagenosis also were negative. Histopathological tests exhibited a large number of histiocytes and emperipolesis, lymphocytes and plasma cells inside of cytoplasm of macrophagic cells, (Figure 1). Immunohistochemical analysis revealed numerous histiocytes that were positive for S-100 protein (Figure 2), CD68, and negative for CD1a. Moreover, this test showed a mix population of lymphocyte B (CD20+) and lymphocyte T (CD3+), and the c-kit mutation were negative. The findings were compatible with Rosai Dorfman disease. The c-kit mutation was negative. The patient keep to be followed by Onco-hematologic services at IMIP and the enlargement of the cervical lymph node not disappeared, and it was started the treatment with corticosteroid for 30 days.

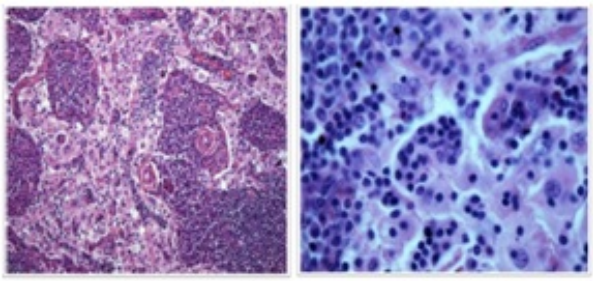

Figure 1: Histological examination of the lymph node revealing a large number of histiocyte containing numerous lymphocytes (emperipolesis), (Hematoxylin and eosin stain 400x).

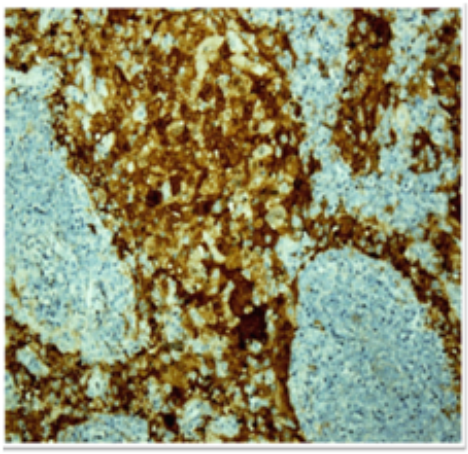

Figure 2: Immunohistochemical staining for protein S100 expression (400x).

Conclusion

We report an atypical case of RDD in adults, with cervical lymphadenopathy without fever or osteolytic lesions. Due to low prevalence of RDD in adults is not still possible to determine whether this disease is more aggressive or indolent, and further studies should be conducted to improve understanding of the pathophysiology, epidemiology and therapeutic approach in these patients.

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