Clinical Medicine and Health Research Journal (CMHRJ)
Volume 04, Issue 05, September - October, 2024
Page No. 1005-1006,

Case Report

Rosai Dorfman Disease: A Rare Disease Case Report

[1]Hamza Sümter, [2]Meryem ümit kurban

[1,2]Uşak Training and Research Hospital Internal Medicine

Received: 01 August, 2024 : Accepted: 04 September, 2024 : Published: 09 September 2024

Abstract:

Rosai Dorfman Disease (RDD) is a rare benign condition. It was first described by Rosai and Dorfman in the 1960s. The etiology, pathogenesis and true frequency of the disease remain unknown. The disease is characterized by the proliferation of histiocytes in the lymph nodes. Clinically, this enlargement of the lymph nodes must be distinguished from other infectious or malignant causes. Although it is often observed at early ages, in this study, a case of RDD detected in a 66-year-old male patient who came to the hematology outpatient clinic due to axillary swelling was presented. The patient is still being monitored without any symptoms,without treatment.

Keywords: Rosai Dorfman Disease, Histiocytosis, Emperipolesis, Benign

Introduction:

Rosai Dorfman Disease (RDD) is known as the enlargement of lymph nodes resulting from a histiocytic proliferative disorder withinthe lymph node sinuses. It is frequently observed during the first two decades of life [1]. It generally presents with a mass in the clinic. Diagnosis is based on lymph node histology. In pathology, varying quantities of lymphocytes, plasmocytes and erythrocytes are phagocytosed by histiocytes. The reason is unknown. Most of the time, fever, weight loss, night sweats, etc. (symptom B) is not observed. The prognosis following surgical intervention is generally favorable. The true prevalenceof the disease is unknown.

Case report:

A 66-year-old male patient with no known systemic disease was evaluated in the hematology outpatient clinic with swelling in the left armpit area. There was no use of medical or herbal drugs in his anamnesis. There was no lymphoproliferative disease in his family. During the evaluation of the patient, no evidence of fever, weight loss, or night sweats was observed. Upon physical examination, a palpable mass in the left axilla was observed (picture 1-2). Biochemical values (transaminases, renal function tests) were within the normal range. In the complete blood count, Hb: 14.3 g/Dl, Hct: 43, WBC: 6100/uL, PLT: 230,000/Ul. No evidence of pathology was detected in hepatitis (HAV, HBV, HCV), TORCH, EBV, brucella, salmonella tests. Peripheral blood smear was evaluated as 60% polymorphonuclear leukocyte (PMNL), 35% lymphocytes, 5% monocytes. No atypical cells were seen.

An active lymph node measuring 7 x 3 cm was detected in the left axillary region viaultrasonography. First, a fine needle aspiration biopsy was conducted, followed by a tru cut biopsy. However, both biopsy samples were non-diagnostic. The subsequent excisional biopsy was evaluated as CD68(+), CD163(+), S100(+) Rosai Dorfman disease (sinus histiocytosis with massive lymphadenopathy). The patient, who continues to experienceno symptoms, is being monitored without treatment in our hematology outpatient clinic.

Discussion:

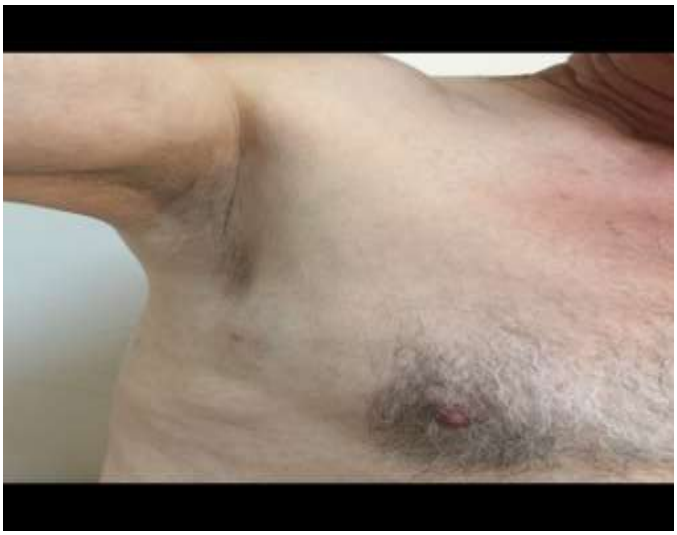
Rosai Dorfman Disease (RDD) was initially delineatedby Rosai and Dorfman in 1969[2]. Although it can manifest at any age, it is most commonly diagnosed duringthe first two decades of life [1]. It has been stated that it is more common among males [3, 4]. Although it is stated that fine needle aspiration biopsy can be useful in diagnosis, the relevant method could not be used in this case [1, 4]. The incidence of RDD, which is rare, in the United States of America (USA) is 100 cases per year [5]. Although various causes are considered in etiology (CMV, EBV, Parvovirus B19, HHV-8, VZV, Brucella, Klebsiella, polyomavirus, autoimmune diseases), no definitive pathogen can be blamed [1, 6, 7]. Immune dysfunction is held responsible [3]. The classic presentation is painless cervical swelling [6]. This swelling may be nodal or extranodal[1]. Extranodal involvement has been observedin various percentages of patients, such as 25-40-43% [3, 6]. While it can manifest in any part of the body, it is frequently observedin the cervical region. Histological evaluation is essential in diagnosis. A histiocytic proliferative disorder is present in the lymph nodes [1, 3]. The important and characteristic histological finding of RDD is that varying amounts of lymphocytes, plasmocytes and erythrocytes are phagocytosed by histiocytes [6]. This feature is defined as emperipolesis or lymphocytophagocytosis[8]. The normal structure of the lymph node is disrupted as a result of the significant enlargement of the lymph sinuses. In immunohistochemical evaluation, there are CD11c (+), S100(+), CD68(+), CD14(+), CD33(+). Also, CD163(+), cyclin D1(+), OCT2(+), CD1a (-), CD207(-), ZBTB 46(-). Phospho ERK and p16 are frequentlypositive. Rarely, BRAFV600 E positivity may occur [2, 4].

In the differential diagnosis, reactive lymph node hyperplasia, infectious lenadenitis, langerhans cell histiocytosis, lymphoma, hemophagocytic syndrome, and metastatic carcinoma should be considered [6]. RDD can be distinguishedfrom Langerhans cell histiocytosis by CD1a negativity [1]. There is no emperipoesis in hemophagocytic syndrome. There is pancytopenia and hepatosplenomegaly [4].

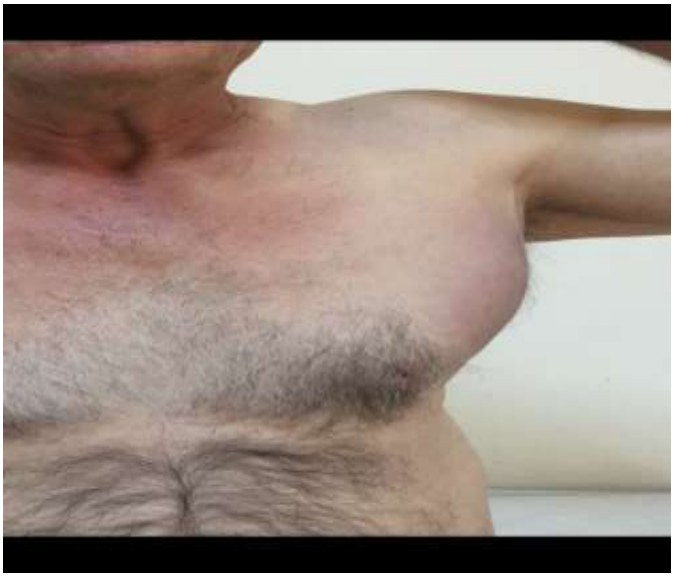
Although there are steroids, chemotherapy, antibiotics, radiotherapy and surgical methods in the literature, there is no standard treatment [1, 6]. Drugs that have been tried in treatment include: sirolimus, rituximab, 6-Mercaptopurine, cladribine, lenalidomide, thalidomide, clofarabine, comimetinib[9].

RDD generally has a favorableprognosis and often regresses spontaneously[5, 7]. While a wait-and-see approach can be applied in asymptomatic patients, 1-2 mg/kg/day prednisone treatment is recommended for symptomatic patients. After 2-3 weeks of steroids, a dose reduction in 2 months has been recommended [9].

Accompanying immune disorders and extranodal involvement are significant in the clinical course of RDD. The severe clinical picture is noteworthy in cases with SLE and AIDS [8]. Bone involvement is also associated with poor prognosis [7].



Picture -1



Picture-2

Conclusion:

Rosai Dorfman disease is a rare cause of lymph node enlargement and is caused by proliferation in the lymph node sinuses. The etiology, pathogenesis and frequency of the disease remainunknown. Histology (immunological histochemistry) is essential in diagnosis. The prognosis is generally favorable following surgical intervention. In most cases, no further treatment is necessary, such as chemotherapy or radiotherapy. It is crucial to distinguish it from other benign and malignant causes.

The patient provided consent for this case report.

References:

1. Santra, G., et al., Rosai-Dorfman disease. Singapore Med J, 2010. 51(10): p. e173-5.
2. Doglioni, C., Rosai-Dorfman disease. A legacy of Professor Rosai that is still not exploited completely. Pathologica, 2021. 113(5): p. 388-395.
3. Oğultekin Vazgeçer, E., et al., Çocukluk Çağında Ender Görülen Bir Lenfadenopati Nedeni: Masif Lenfadenopatili Sinüs Histiositozu (Rosai-Dorfman Hastalığı). 2014. 14(4).
4. Kushwaha, R., C. Ahluwalia, and V. Sipayya, Diagnosis of sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman Disease) by fine needle aspiration cytology. J Cytol, 2009. 26(2): p. 83-5.
5. Synghal, G., et al., Rosai-Dorfman disease in a symptomatic elderly man. Proc (Bayl Univ Med Cent), 2022. 35(1): p. 78-79.
6. Giri, K., et al., Rosai-Dorfman disease in 6-year-old child: Presentation, diagnosis, and treatment. Clin Case Rep, 2021. 9(9): p. e04795.
7. Garcia, R.A. and E.F. DiCarlo, Rosai-Dorfman Disease of Bone and Soft Tissue. Arch Pathol Lab Med, 2022. 146(1): p. 40-46.
8. ÇIRAK, K., et al., ROSAİ-DORFMAN HASTALIĞI (OLGU SUNUMU). 16(2): p. 1-8.
9. Salama, H.A., et al., Highlights of the Management of Adult Histiocytic Disorders: Langerhans Cell Histiocytosis, Erdheim-Chester Disease, Rosai-Dorfman Disease, and Hemophagocytic Lymphohistiocytosis. Clin Lymphoma Myeloma Leuk, 2021. 21(1): p. e66-e75.