



ROSAI DORFMAN DISEASE IN A PREGNANT FEMALE: A CASE REPORT

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ABSTRACT:

Rosai Dorfman disease (RDD) or sinus histiocytosis is a rare, idiopathic, non neoplastic histiocytic proliferation of unknown etiology typically presenting with massive cervical lymphadenopathy with or without systemic involvement. It may occur in any age group, but is most commonly seen in children and young adults. We report a case of RDD with bilateral massive cervical swelling in a 21 years old pregnant female. The swelling progressively increases in size after the onset of pregnancy. FNAC was advised. The cytomorphology revealed lymphocytes, plasma cells and uni to multinucleated histiocytes with lymphophagocytosis and the diagnosis of RDD was rendered. It is concluded that FNAC is a reliable and useful tool for the diagnosis of sinus histiocytosis with massive lymphadenopathy (RDD) and biopsy can be avoided, thus reducing the inconvenience to the patients especially in pregnancy.

Key words: Rosai Dorfman disease, Lymphophagocytosis, Pregnant

INTRODUCTION:

Sinus histiocytosis with massive lymphadenopathy, commonly known as Rosai Dorfman Disease; a rare and benign entity of unknown etiology was first described by Rosai and Dorfman in 1969.² It mainly involve the lymphnodes and characterized by painless, bilateral cervical lymphadenopathy accompanied by fever, leukocytosis, elevated ESR and hypergammaglobulinemia. It is very important to distinguish it from lymphoma and other causes of histiocytosis, for proper treatment of the patient. The disease is usually self limiting. Here, we report a case of progressive enlargement of bilateral cervical swellings in a 21 years old pregnant woman.

CASE REPORT:

A 21 yrs old female in third trimester of pregnancy presented in FNAC clinic with chief complaint of progressively increasing bilateral swellings for last one year. Initially the swelling was 2x1 cm as in OPD records but it progressively increased after she conceived and now she presented with bilateral multinodular swellings of size 4x3 cm approximately. There was no history of fever or weight loss. General physical examination was within normal limits with absence of jaundice, cyanosis and oedema. On local examination revealed swellings were firm, non tender and varying in diameter from 6 to 7 cm. Skin overlying the swellings was normal. There was no history of night sweats, rashes and bleeding manifestations. Her past and personal history was not significant. There was no family history of similar illness. Other systems were within normal limits. Hematological examination revealed Hb 8.4 gm%, PCV 40.6%, MCV 78 fl, MCH 28 pg, MCHC 30 %, Retic 1.2% . The peripheral blood count revealed leukocytosis (11,800 / cu mm) with neutrophilia (82%), platelet count 3.49 lakhs/cu mm. ESR was raised

(145 mm 1st hour by Westergren's method). Montoux test was negative. Peripheral blood film showed microcytic hypochromic red blood cells with mild anisocytosis. Chest radiograph was normal. Ultrasonography of the abdomen was normal and did not reveal any organomegaly or lymphadenopathy. Routine biochemical and urine examination were within normal limits. Serological examination for HIV I & II, HBV and Toxoplasma were negative.

FNAC of the cervical swellings was done. Smears stained with Romanowsky stain, were cellular; and revealed large number of histiocytes present in small and large groups having abundant pale to eosinophilic cytoplasm with mono to bi to multilobated vesicular nuclei. Some of these histiocytes revealed phagocytosis of lymphocytes, neutrophils and plasma cells against the background of lymphocytes and RBCs. Cytopathological possibility of Rosai Dorfman Disease of bilateral cervical LN involvement was rendered.

DISCUSSION:

RDD is a rare, non neoplastic histiocytic proliferation of idiopathic origin.¹ Most cases occur during the first or second decade of life, however any age can be affected. Males are involved more than females.^{2,3}

Most patients (95%) present with massive cervical lymphadenopathy. The extent of lymphadenopathy is often striking with nodes as large as 6 cm have been noted in some cases leading to respiratory compromise. In 43% of cases, extranodal sites are involved simultaneously and in only 23% isolated extranodal Rosai Dorfman Disease occur.³ Clinically patients may be mistaken for lymphoproliferative or other infectious disorders like tuberculosis which is more prevalent in developing countries like India. Non specific systemic symptoms include fever, weight loss and malaise. Patient may also present with tonsillitis, nasal discharge and obstruction.¹ Laboratory abnormalities include anemia, leucocytosis, elevated ESR, and hypergammaglobulinemia.²

Exact aetiology, pathogenesis and natural history of the disease is still not known despite intense investigations over the last 20 years.⁸ An immune mediated origin has been proposed by few, while others believe it to be a reactive process due to a viral infection (HHV-6) or an undefined immunological defect initiated by some other organism like EBV or CMV may be responsible for the disease.^{1,2} Few investigators consider it to be of bone marrow stem cell origin.² At present, it is considered a reactive proliferation showing spontaneous regression.³ RDD is recruitment of marrow monocytes from peripheral blood into

lymph nodal sinuses or extranodal sites and their transformation into the immunophenotypically distinct RDD histiocytes which demonstrate emperipoleisis and functional uniqueness in terms of cytokine expression profile. Release of cytokines like TNF- α from these cells is responsible for the genesis of fever and other systemic symptoms.⁷ There is no ideal therapeutic regimen, treatment option ranges from surgery, radiotherapy and steroids to chemotherapy.³

Although RDD is a histopathological diagnosis, yet FNAC has proved to be a reliable tool in diagnosing it. Only a few cases of RDD diagnosed by fine needle aspiration cytology have been reported in the literature. Cytology reveals numerous large histiocytes with abundant pale cytoplasm with phagocytosed lymphocytes, neutrophils and plasma cells (emperipoleisis). In emperipoleisis the lymphocytes are not attacked by enzymes and appear intact within the histiocytes in contrast of phagocytosis. The phenomenon of emperipoleisis is highly useful for the diagnosis of RDD using FNAC.^{1,2} All these features were also present in our case. None of the case has been reported in pregnant female as in our case.

Histologically, the lymphatic sinuses are dilated and are reveal presence of lymphocytes, plasma cells and numerous large histiocytes with emperipoleisis resulting in partial or complete architectural effacement.^{5,8}

Differential diagnosis includes nonspecific sinus hyperplasia, Langerhans cell histiocytosis, hemophagocytic syndrome, tuberculosis, lymphoma and metastatic malignant melanoma.⁹ Reactive sinus hyperplasia shows large clusters of histiocytosis, accompanied by reactive lymphocytes, germinal center cells, immunoblast and tingible macrophages, emperipoleisis is absent. In LCH, langerhans cells have grooved and twisted nuclei and the background has eosinophilic microabscesses. LCH is positive for both S-100 protein and CD 1a. Hemophagocytic syndromes should be differentiated from RDD on the basis of the presence of hemophagocytosis, absence of emperipoleisis and the presence of pancytopenia and hepatosplenomegaly. Tuberculous lymphadenitis shows epithelioid cell granuloma with or without caseous necrosis which are absent in RDD. Smears from patients with Hodgkins disease show lymphocytes, plasma cells, histiocytes, eosinophils and Reed- Sternberg cells.^{1,2} They are S100 protein negative, CD15 positive, and CD30 positive. Negative staining for HMB-45 and pankeratin differentiate RDD from melanoma and metastatic carcinoma.⁹



Clinical photograph of the patient

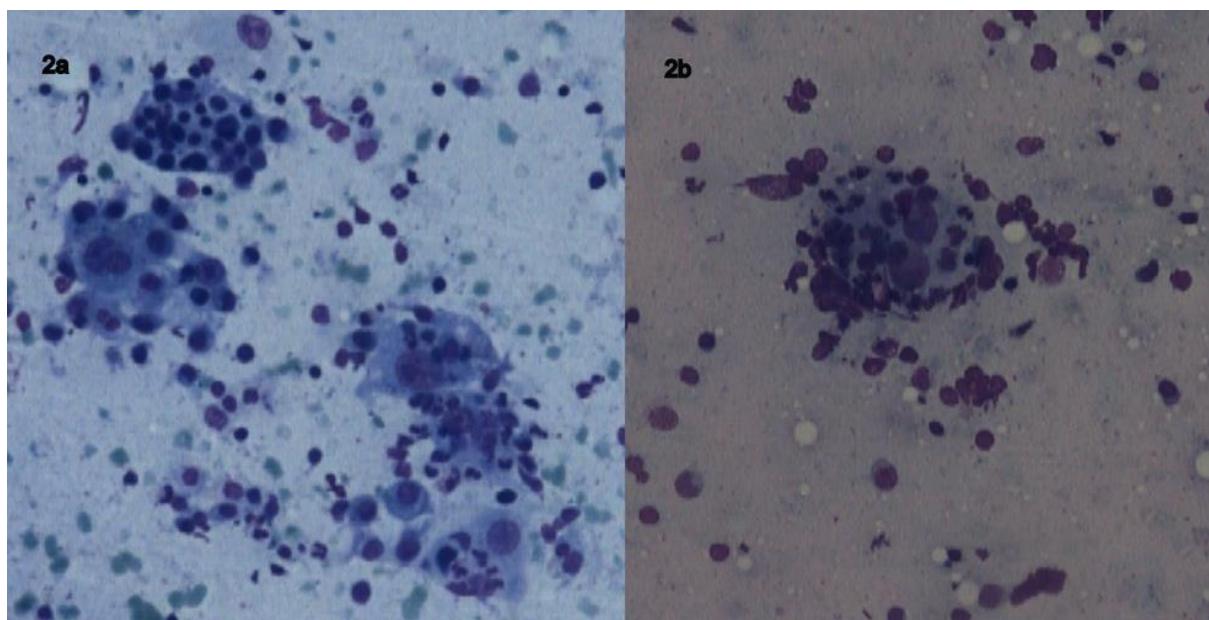


Figure showing FNAC and Histopathology from the specimen

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Legends:

Figure 1: Photograph of patient showing massive enlargement of cervical lymphnodes.

Figure 2a&b : Cellular smears showing large number of histiocytes in groups or scattered singly and showing phagocytosis of lymphocytes.