



CYTOLOGICAL DIAGNOSIS OF ROSAI DORFMAN DISEASE IN ELDERLY FEMALE – A CASE REPORT

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<p>Article Info <i>Received 15/05/2015</i> <i>Revised 11/06/2015</i> <i>Accepted 20/06/2015</i></p> <p>Key words: Rosai Dorfman disease, Axillary lymph node, Elderly female.</p>	<p>ABSTRACT Rosai Dorfman disease also called as Sinus histiocytosis with massive lymphadenopathy is a benign self limiting disorder that commonly involves the lymph nodes. Rosai Dorfman disease(RDD) was first described by Rosai and Dorfman in 1969. There is no definite etiological agent and the lymph nodes may attain large size and may remain enlarged for a long duration. RDD is not confined to a particular race or geographical area, although it is more commonly noted in African areas. RDD is characterized by massive, painless and bilateral enlargement of the lymph nodes more commonly the cervical lymph nodes. We here present a case of a 50-year-old lady suffering from this disease who presented with lymphadenopathy in left axillary region. Fine needle aspiration cytology (FNAC) from the lesions showed abundant benign histiocytes with lymphophagocytosis which was compatible with the diagnosis of RDD. This case is being reported for its rarity in presentation in an elderly female and diagnosing cytologically.</p>
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INTRODUCTION

Sinus histiocytosis with massive lymphadenopathy (SHML) is a rare but well-defined, histiocytic, proliferative disorder. Many people consider it to be related to bone marrow stem cell origin [1]. Majority of the cases of RDD occur during the first or second decade of life. However, there are chances of any age group may be affected; males are more affected than females [2]. Though cervical lymph node involvement is more common site for this disorder, other lymph node sites like the axillary, para aortic, inguinal, or mediastinal lymph nodes can be affected. There are chances of extranodal involvement which may occurs in 43% of cases, either in association with lymph node involvement or otherwise. The Various sites which have been affected are the nasal and paranasal cavities, central nervous system, eyes and retro-orbital tissue, skin and bones.

CASE REPORT

A 50 year old lady came to General surgery department with complains of swelling in the left side of axilla from past 2 months with on and off fever.

Patient was sent to Department of Pathology for Fine Needle Aspiration Cytology (FNAC). On examination the lymph node was of size 3x3 cm, firm and painless. FNAC was performed by using 10 ml syringe with 23 gauze needle and the aspirate was hemorrhagic. Routine H&E stains and Giemsa stains were performed and it was diagnosed as RDD cytologically.

DISCUSSION

Although the exact pathogenesis of RDD is not known there are few theories suggesting an infectious cause and immunodeficiency state. Presence of human herpes virus 6 genome was demonstrated by in situ hybridization; [3] and relationship with Klebsiella, Epstein-Barr virus, Brucella, or Cytomegalovirus were also suggested [4]. RDD can be seen in any age group but 80% of the cases are seen within the first two decades of life in our case the age of the patient was 50 years

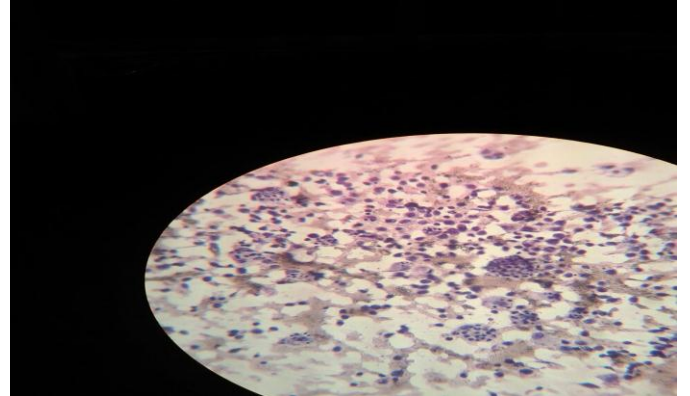
In the present case we can see numerous large histiocytes with abundant, pale cytoplasm, and



Figure 1. Photograph showing swelling in the left axillary region.



Figure 2. Photograph showing emperipolesis and few neutrophils. [H&E,x40].



phagocytosed lymphocytes (emperipolesis) on microscopy. Background shows lymphocytes, plasma cells, and occasional neutrophils [Figure 2].

Histiocytes show positive immunostaining for S100 protein; CD11c, CD14, CD33, and CD68 antigens in cytological smears. Differential diagnoses include reactive sinus histiocytosis, malignant histiocytosis, hemophagocytic syndrome, tuberculosis, and lymphoma.

In Langerhans cell histiocytosis, Langerhans cells have grooved and twisted nuclei and the background has eosinophilic microabscess. Reactive sinus histiocytosis shows loose clusters of histiocytes, accompanying which are reactive lymphocytes, germinal center cells, immunoblasts, and tingible body macrophages; emperipolesis is absent. Hemophagocytic syndromes should be differentiated from Rosai–Dorfman disease on

the basis of the presence of hemophagocytosis, absence of emperipolesis, and the presence of pancytopenia and hepatosplenomegaly. Tuberculous lymphadenitis show epithelioid cell granuloma with or without caseous necrosis, which are absent in Rosai–Dorfman disease.

Smears from patients with Hodgkin's disease show lymphocytes, plasma cells, histiocytes, eosinophils, and Reed–Sternberg cells. In Rosai-Dorfman disease, Reed-Sternberg cells and eosinophils are not seen.

CONCLUSION

Thus, the cytomorphology of Rosai–Dorfman disease is so distinctive that it can be diagnosed by FNAC. FNAC is a reliable and sensitive means to establish conclusive diagnosis, obviating the need for biopsy.

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