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Case Report

A case of Rosai-Dorfman Disease - a rare case diagnosed on FNAC

Binaya Shrestha[1*], Soniya Shrestha[2], Dipika Basnet[3], Nikita Gautam[4]

Author's Affiliations

[1]Lecturer, Department of Pathology, Dhulikhel Hospital, Kavre
[2]PG Resident, Department of Pathology, Dhulikhel Hospital, Kavre
[3]Assistant Professor, Department of Pathology, Dhulikhel Hospital, Kavre
[4]Lecturer, Department of Pathology, Dhulikhel Hospital, Kavre

Correspondence to:
Dr, Binaya Shrestha
Department of Pathology, Dhulikhel Hospital, Kavre, Nepal
Email: binayashrestha008@gmail.com

ABSTRACT

Background & Objectives: Sinus histiocytosis with massive lymphadenopathy (SHML) also known as Rosai-Dorfman Disease is a rare histiocytic proliferative disorder which can often be misdiagnosed as lymphoma clinically. On Fine Needle Aspiration Cytology (FNAC) very few cases of SHML have been diagnosed. The aim was focused on the cytological characteristics of the Rosai-Dorfman disease and differential diagnoses.

Presentation of Case: Here, we present a case of Rosai-Dorfman Disease in a 12 year old girl presented with a painless swelling over left side of neck.

Discussion: Rosai-Dorfman disease which is a rare histiocytic disorder involves mainly the lymph nodes and also extra nodal sites like head and neck. Most patients with RDD have spontaneous remission but few cases can be fatal. The differential cytological diagnosis includes reactive lymph node hyperplasia, infectious lymphadenitis, hemophagocytic syndrome, Langerhans cell histiocytosis and lymphoma.

Conclusion: RDD is rare and require knowledge of its cytological characteristics for a rapid and correct diagnosis. FNAC is enough for the final diagnosis of the disease.

Keywords: Emperipolesis, FNAC, Rosai-Dorfman, Sinus histiocytosis

INTRODUCTION

Sinus histiocytosis with massive lymphadenopathy (SHML) or Rosai-Dorfman

Disease (RDD) is a rare benign self-limited disorder involving the lymph nodes that was first described in 1969 by pathologists Juan Rosai and Ronald Dorfman [1-3]. It is a disorder of unknown etiology with some familial cases associated with Germ line mutations in SLC29A3. The aetiology is unlikely to be uniform across all cases, and various viruses such as Human Immunodeficiency Virus (HIV), Epstein-Barr Virus (EBV) and Cytomegalo Virus (CMV) have been linked to the disease [1,4].

It most often presents as a painless, massive bilateral enlargement of the cervical lymph nodes associated with fever, leucocytosis, elevated erythrocyte sedimentation rate (ESR), and polyclonal hypergammaglobulenemia. Most cases occur during the first or second decade of life with male preponderance [2]. Extra nodal disease is also common, often with a particular predilection for head and neck region [3,5,6]. It is a characterised by sinus enlargement with presence of histiocytes with round to oval vesicular nuclei, prominent nucleoli and abundant cytoplasm showing emperipolesis within histiocytes. Histiocytes show positive immunostaining for S-100 protein, vimentin and CD 68 antigen and is negative for CD1a [3,5].

CASE REPORT

A 12 years old female presented with history of painless swelling over left side of neck for one week (Figure 1). Ultrasonography was done which showed multiple enlarged lymph nodes likely to be tubercular nodes. Routine laboratory investigations revealed hemoglobin 11.6 g/dl, erythrocyte sedimentation rate (ESR) of 8 mm at the end of first hour, total leukocyte count of $7.6x10^3$ cells/mm3. Fine needle aspiration cytology (FNAC) was performed from the swelling. Smears were stained with Wright as well as Papanicolaou stain. Smears were highly cellular with numerous histiocyte with single to multiple nuclei, vesicular nuclei with prominent nucleoli and abundant pale cytoplasm showing lymphophagocytosis harboring numerous intact lymphocytes in their cytoplasm. Background showed plenty of mature lymphocytes, plasma cells and neutrophils admixed with hemorrhage (Figure 2 and 3).

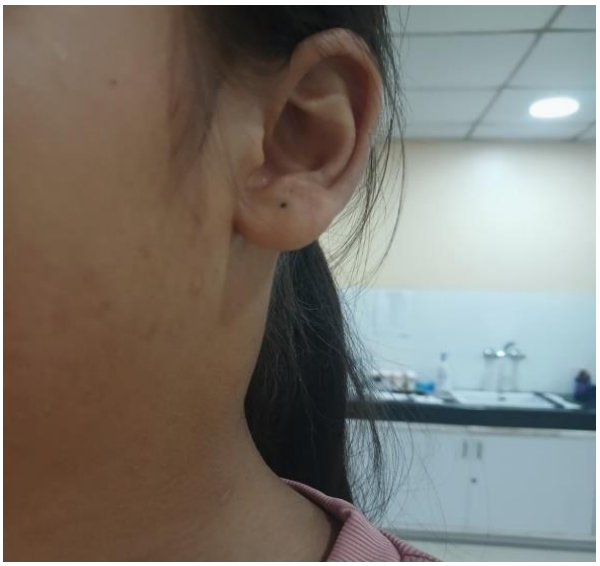

Figure 1. Swelling over left side of neck

DISCUSSION

Rosai-Dorfman disease is a rare histiocytic disorder of unknown etiology involving the lymph nodes [2]. Only few cases of Rosai - Dorfman Disease have been reported on FNAC [3]. But due to its distinct cytological features, FNAC alone is enough to make the diagnosis in most cases thereby avoiding more unnecessary invasive procedures [7,8]. Most patients with SHML have spontaneous remission and some can recur or have persistent disease with asymptomatic but persistent lymphadenopathy. In very few

cases it progresses to an aggressive tumor and can be fatal [7].

The classical histology is characterized by distorted nodular architecture with marked dilation of lymphatic sinuses, partial effacement of follicles and germinal centers, as well as capsular and pericapsular fibrosis. Lymphatic sinuses are occupied by numerous lymphocytes and histiocytes with vesicular nucleus and abundant clear cytoplasm with phagocytized lymphocytes or plasma cells, also known as 'emperipolesis' [7,9].

The differential cytological diagnosis includes reactive lymph node hyperplasia, infectious lymphadenitis, hemophagocytic syndrome, Langerhans cell histiocytosis and lymphoma [7]. In the lymph node reactive hyperplasia there are sinusoidal hyperplasia with loose clusters of histiocytes, accompanying lymphocytes, germinal center cells, immunoblasts, and tingible body macrophages; however cytology usually does not show extensive emperipolesis while protein S-100 is negative. Hemophagocytic syndromes should be differentiated from Rosai-Dorfman disease on the basis of the presence of hemophagocytosis and absence of emperipolesis. The benign histiocytes

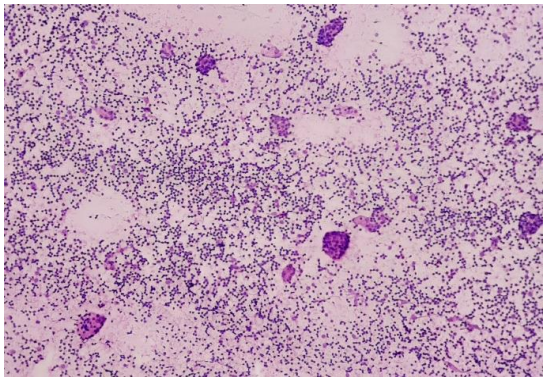

Fig. 2(a)

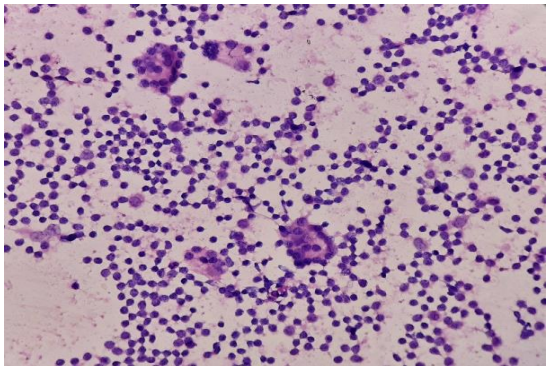

Fig. 2(b)

Figure 2. Cellular smear with clusters of histiocytes over lymphocytic background. Magnification a. 100X b. 400X

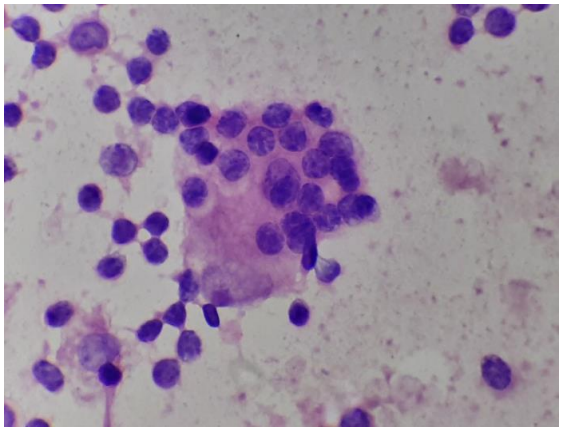

Fig. 3 (a)

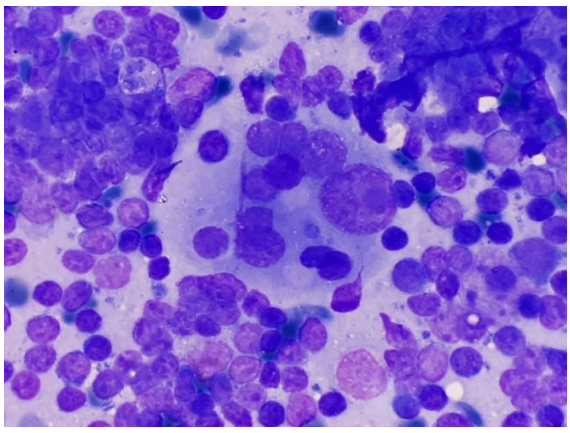

Fig. 3(b)

Figure 3. Histiocytes showing emperipolesis of lymphocytes. Magnification a. 100X b. 400X

engulf erythrocytes and platelets and this condition is associated with malignancy and infectious processes [7]. In Langerhans cell histiocytosis, Langerhans cells have grooved and twisted nuclei and the background has eosinophilic microabscess. Langerhans cells also express CD1a [7,10].

CONCLUSION

FNAC is enough to make the diagnosis of RDD due to its characteristics cytological features, thus other unnecessary invasive procedures can be avoided.

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Author's Contribution: contributed in performing FNAC, diagnosing the case and; taking the clinical and micro photographs- **BS,SS.** Both the authors read the final version and approved for publication.

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