



Case Report:

Diagnosis of Rosai-Dorfman Disease by Fine Needle Aspiration Cytology in a Case with Cervical Lymphadenopathy and Nasal Mass.

Madhusmita Jena, Associate Professor, Department of Pathology,
MVJ Medical College & Research Hospital, Bangalore, India.

Address for Correspondence:

Dr. Madhusmita Jena,

11C4, Krishna Nagar Apt,

Annasandrapalya,

HAL Post, Bangalore -560017,

India.

E-mail: jena_madhusmita@hotmail.com

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Abstract: We report a case of Rosai-Dorfman Disease, a rare non neoplastic proliferative disorder of the cells of macrophage-histiocyte family, in a case with cervical lymphadenopathy and nasal mass diagnosed by fine needle aspiration cytology .

Key Words: Rosai-Dorfman Disease; Cervical Lymphadenopathy; Nasal Mass; Fine needle aspiration cytology

Case Report:

A 46 years old male presented with complaints of stuffiness and blockade of nose on the left side since six months. On examination a mass measuring 2.5x1.5cms was seen occupying the left nasal cavity. Multiple bilateral cervical lymph nodes were enlarged. The largest one measured 1.0x0.5cms. Routine blood counts were within normal limits. The ESR was 40mm/1st hour. Chest X-Ray showed no abnormality. A clinical diagnosis of lymphoma was suggested.

A fine needle aspiration was performed on cervical lymphnodes using a 22G needle. Smears were stained in Papanicolaou and Giemsa stains. This was followed by an excisional biopsy of the cervical lymph node and nasal mass.

The aspirated smears showed a polymorphous population of cells consisting of mature lymphocytes, plasma cells, neutrophils and histiocytes. A diagnosis of reactive lymph node was made initially. However a review of the Pap stained smears showed few histiocytes with vesicular nuclei and abundant pale cytoplasm. The cytoplasm of the histiocytes exhibited intact engulfed lymphocytes (emperipolesis) within them (Fig 1 & 2). A cytological diagnosis of Rosai-Dorfman disease was suggested based on the characteristic cytomorphology.

Following biopsy, Haematoxylin & Eosin sections of the excised cervical lymph nodes showed a normal architecture with dilated sinuses filled with histiocytes having abundant pale cytoplasm and some of them showing emperipolesis (Fig 3 & 4).The excised nasal mass showed the similar features on histology.

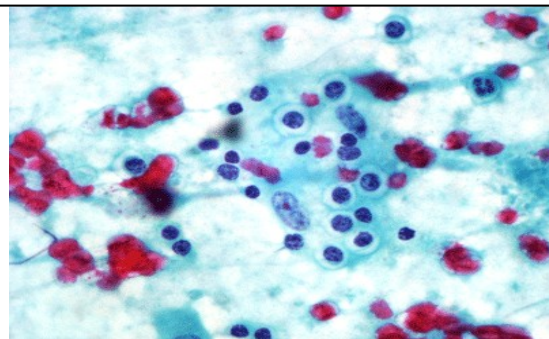


Fig 1: Smears showing histiocyte with evidence of emperipolesis (Pap, X400)

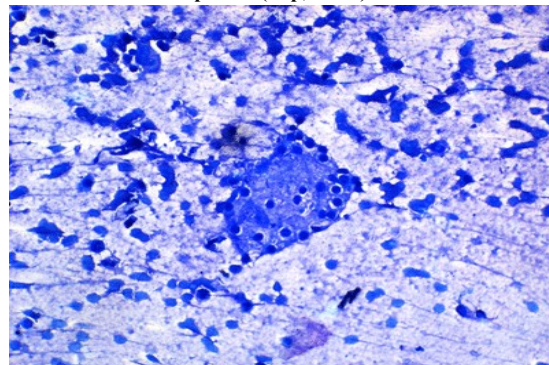


Fig 2: Smear showing histiocyte with engulfed intact lymphocytes (Giemsa, X400)

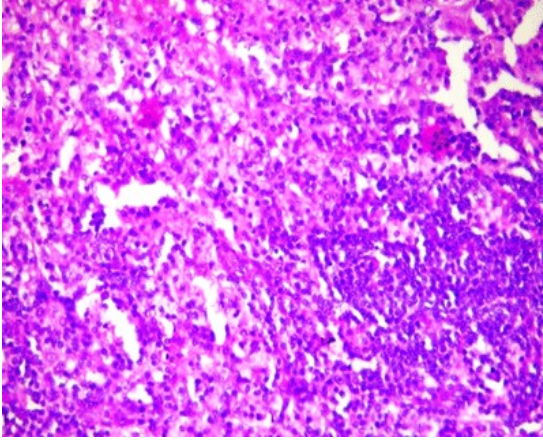


Fig 3: Excisional biopsy of the cervical lymph node showing dilated sinuses filled with histiocytes with abundant pale eosinophilic cytoplasm (H&E, X100)

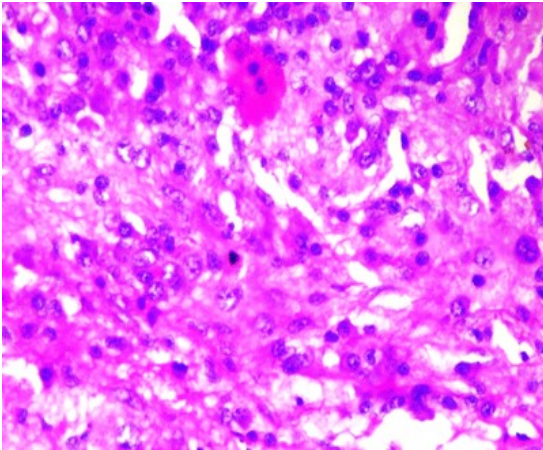


Fig 4: Excisional biopsy of the cervical lymph node showing histiocytes with evidence of emperipolesis (H&E, X400)

Discussion:

Rosai-Dorfman Disease, also referred to as Sinus histiocytosis with massive lymphadenopathy, is a rare non neoplastic proliferative disorder of the cells of macrophage-histiocyte family that was first described by Destombes in 1965.¹ It was recognized as a distinct clinicopathologic entity by Rosai and Dorfman in 1969.² It commonly affects the cervical lymph nodes giving rise to painless enlargement of lymph nodes which may clinically mimic a lymphoma. It is characterised by expansion of sinuses of the lymph nodes and the lymphatics of extranodal sites by proliferation of histiocytes with abundant pale eosinophilic cytoplasm containing engulfed lymphocytes.

The exact etiology of the disease is unknown.³ It is presumed that an aberrant exaggerated immune response to an infectious agent causes a proliferation of histiocytes. Although several infectious agents are suspected, but none of them are documented so far.⁴ The expression of HHV6 (Human Herpes Virus) specific p101k antigen was found in follicular dendritic cell in SHML.⁴

SHML is a disease of childhood and early adulthood. Clinically the mean age of onset is second decade.^{4,6,7} The patient in this case was a middle aged (46yrs) man. The most common presenting symptom of this disease is painless bilateral cervical lymphadenopathy which is seen in 90% of the patients and 43% of the cases the patients have at least one site of extra nodal involvement.^{4,7} Rarely sites other than lymph nodes are involved such as skin and soft tissue, upper respiratory system, genitourinary tract, eye, orbit, kidney, thyroid, breast, bone.³ The other

accompanying features are fever, leucocytosis, elevated ESR and polyclonal hypergammaglobulinaemia.⁷ In the present case the patient had bilateral cervical lymphadenopathy and a nasal mass with an elevated ESR. Clinically the patient was suspected of lymphoma and the possibility of Rosai-Dorfman disease was not considered until FNAC was performed.

As per the review of literature there are only few reports or small series of cases on the (FNA) cytologic features of this entity.⁶ The characteristic cytomorphology of this entity is the presence of large histiocytes with abundant cytoplasm having variable number of intact lymphocytes within it; a phenomenon referred to as lymphophagocytosis or emperipolesis.^{4,6-8} The background typically shows lymphocytes, plasma cells and occasional neutrophils.⁶⁻⁸ In the present case the diagnosis was initially missed on FNAC as the number of histiocytes were not many and so were probably overlooked, but on a review of the smears the histiocytes showed the characteristic feature of emperipolesis and therefore a diagnosis of Rosai-Dorfman disease was made. Besides cytomorphology, the histiocytes on immunostaining show positivity for S100 protein, CD14, CD33 & CD68 in cytological smears.⁸ Extranodal involvement is seen in upto 40% of cases which show similar morphological features to its nodal counterpart although fewer histiocytes with emperipolesis are encountered.^{3,5} The patient had an extra nodal involvement in the form of a nasal mass which on biopsy showed features of sinus histiocytosis with few histiocytes showing the characteristic emperipolesis.

Although the cytomorphological features are well described, diagnostic difficulties can sometimes arise. The main differential diagnosis on FNAC of the lymph nodes include reactive lymphoid hyperplasia with sinus histiocytosis, malignant histiocytosis, lymphoma and tuberculosis.^{3,6,8} Prominent emperipolesis, polymorphous cell population, absence of grooved nuclei of histiocytes, absence of eosinophils, absence of Reed Sternberg cells and absence of epithelioid granulomas rule out the above conditions in the present case.

The course of this disease is usually self limiting in most of the patients and so treatment is not necessary in majority of them. Surgery is generally limited to biopsy to confirm the diagnosis or to relieve obstructive symptoms. Steroids are given to the patients with progressive disease.⁷ In this patient surgery was done to confirm the diagnosis of the nasal mass. The patient received steroid therapy to which he responded well.

The author likes to emphasize firstly that the possibility of SHML as a differential diagnosis has been kept in mind of the cytopathologist while examining FNA smears of a lymph node. Secondly a careful interpretation of the morphology of the histiocytes is required whenever they are less in number as the diagnosis may be missed at the initial stages of the disease or if the aspirate is not from a representative area of the lymph node. Thirdly if in a case there is an extra nodal manifestation where the lymph node aspirate shows a possibility of SHML, then a FNA can be performed on the extranodal site and the cytological findings of both the sites can be correlated. Fourthly the cytological findings should be interpreted in the appropriate clinical context. Thereby FNA can be used as a reliable tool to establish a diagnosis and an unnecessary biopsy which is an invasive procedure can be avoided.

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