



Cytological Diagnosis of Rosai-Dorfman Disease in Region of Neck

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Abstract

Introduction: *The disease was first described by Rosai and Dorfman in 1969. Also known as Sinus Histiocytosis with Massive Lymphadenopathy (SHML). Patients presenting with lymphadenopathy are commonly diagnosed as having tuberculosis or malignant lymphomas, whereas cases with extra nodal involvement are suspected of having various neoplasms, depending on site of involvement. Cytological features of SHML are virtually diagnostic and can obviate the need for biopsy in most cases.*

Objective: *To study the FNAC findings for diagnosis of this rare disease.*

Case: *The patient in our case was a young male who presented with massive cervical lymphadenopathy, fever, leukocytosis, hypergammaglobulinemia and raised ESR. The patient was clinically diagnosed as having lymphoma or tuberculosis.*

Materials and methods: *FNAC was done and the smears were stained by May-Grunwald Giemsa stain.*

Observations: *Smears were cellular and showed presence of mixed population of small and large lymphocytes, diffusely distributed histiocytes having single or multiple nuclei with fine chromatin, pale cytoplasm and exhibited numerous intact lymphocytes and polymorphs (Emperipolesis). Many foreign body giant cells, few plasma cells and large number of degenerated cells were also seen. Based on this characteristic cytomorphology a diagnosis of Rosai Dorfman Disease was made.*

Conclusion: *Thus, fine needle aspiration cytology is a useful and reliable tool for the diagnosis of Rosai-Dorfman disease due to which biopsy can be avoided.*

INTRODUCTION

The disease was first described by Rosai and Dorfman in 1969. Also known as Sinus Histiocytosis with Massive Lymphadenopathy (SHML). It is a benign, self-limiting disorder that commonly involves lymph nodes (Das et al) and

presents as massive, painless, bilateral, cervical lymphadenopathy accompanied by low grade fever with lab investigation shows leukocytosis, elevated ESR and hypergammaglobulinemia (Das et al).

CASE REPORT

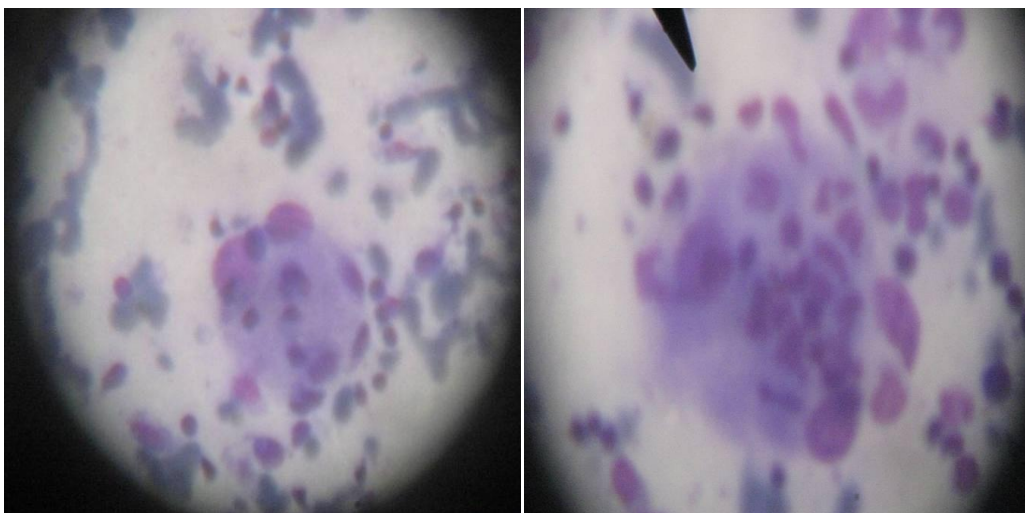
- A 10 yr old male presented with a swelling in the right side of neck since 25 days accompanied with low grade fever. The swelling was 10x7.5cm in size, gradually increasing, non tender, soft to firm and mobile. There was no history of weight loss.
- The peripheral blood cell count showed leukocytosis (17,000/cu-mm) with neutrophilia. ESR was 45mm after 1st hour. Serum Gamma globulins were also raised. But USG of abdomen did not reveal any organomegaly. The patient was clinically diagnosed as having lymphoma or tuberculosis.

FNAC was taken from the right cervical node and smears were stained by May-Grunwald Giemsa stain

Smears were cellular and showed presence of mixed population of small and large lymphocytes, diffusely distributed histiocytes having single or multiple nuclei with fine chromatin, pale cytoplasm and exhibited numerous intact lymphocytes and polymorphs (Emperipolesis).

Many foreign body giant cells, few plasma cells and large number of degenerated cells were also seen. No nuclear atypia or nuclear grooving was observed.

Based on this characteristic cytomorphology a diagnosis of Rosai Dorfman Disease was made.



Smears showing histiocytes and emperipolesis

Although this disorder is self limiting in most cases this patient was put on oral Prednisolone 10mg three times a day in tapering doses for 21 days and after 21 days patient showed marked clinical improvement.

DISCUSSION

SHML or Rosai Dorfman disease is a rare but well defined histiocytic, proliferative disorder of unknown etiology. Some investigators consider it to be of Bone Marrow stem cell origin (Das et

al).Most cases occur during the first and second decade of life and males are affected more than females. Exact pathogenesis of the disease is still unknown but viral etiology is implicated (Kumar et al).Clinically pts present as massive, painless, cervical lymphadenopathy associated with low grade fever, leukocytosis, elevated ESR and hypergammaglobulinemia.

Our case was a young male who presented with massive cervical lymphadenopathy, fever, leukocytosis, hypergammaglobulinemia and raised ESR. However, the possibility of Rosai -Dorfman was not considered until FNAC was performed.

Histiocytes showed positive immunostaining for S 100 protein; CD 11c, CD 14, CD 33 and CD 68 antigens in cytological smears.

Differential Diagnosis-

- Langerhans cell histiocytosis showing grooved, twisted nuclei and eosinophilic micro abscesses in the background.
- Lymphoma reveals lymphocytes, plasma cells, histiocytes, eosinophils and Reed-Sternberg cells.
- Reactive sinus histiocytosis shows loose clusters of histiocytes, reactive lymphocytes, germinal centre cells, immunoblasts and tingible body macrophages,emperipolesis is absent.
- Hemophagocytic syndrome shows hemophagocytosis, pancytopenia and hepatosplenomegaly.
- Tuberculous lymphadenitis syndrome shows hemophagocytosis, pancytopenia and hepatosplenomegaly

CONCLUSION

The characteristic cytomorphological features of the disease are distinctive and thus could be diagnosed by FNAC in our study and also in a study conducted by Kushwaha et al.(2009)

Thus, fine needle aspiration cytology is a useful and reliable tool for the diagnosis of Rosai-Dorfman disease due to which biopsy can be avoided.

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