

## Case Report

### Rosai Dorfman Syndrome- A case report

Joshi Sushant<sup>1</sup>, Bhatia Gunjan<sup>2</sup>, Bhatia Ravi<sup>3</sup>

<sup>1</sup>Consultant ENT specialist, Arravali Hospital, Udaipur.

<sup>2</sup>Consultant Pathologist, GBH American Hospital, Udaipur.

<sup>3</sup>Assistant Professor, Dept of Pediatrics, Pacific Medical College, Udaipur.

Corresponding Author: Dr. Bhatia Ravi

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#### Abstract:

Rosai Dorfman disease also known as sinus histiocytosis with massive lymphadenopathy is a rare benign disorder of unknown aetiology, which present most commonly as bilateral massive lymphadenopathy of neck associated with fever. These cases are frequently misdiagnosed as lymphoma and thus it is important to distinguish Rosai Dorfman disease from other causes of neck swelling because of different treatment modalities. We report here a case of Rosai Dorfman disease presenting with massive right cervical lymphadenopathy.

**Keywords:** Sinus Histiocytosis, Massive Lymphadenopathy, Rosai Dorfman Disease

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#### Introduction:

Sinus Histiocytosis with massive lymphadenopathy was first described by Rosai Dorfman in 1969. Although the etiology remains unknown the disease is thought to be a disorder of immune regulation or response to an infectious agent with major manifestations in the lymphnodes resulting in proliferation of histiocytes. It is mostly a benign, self limiting disorder that commonly involves the lymph nodes<sup>1</sup>. The most common clinical manifestation of the disease is bilateral, non tender cervical lymphadenopathy often resembling a bull's neck. Although cervical region is by far the most common and most prominent site of involvement but peripheral or central lymph nodes are also affected. In about 25 to 40 % of cases, extranodal sites are also affected<sup>2-4</sup>. Since 1969 a registry has been maintained

by Rosai, the majority of the cases have presented within first two decades of life.

The cytological features of SHML are virtually diagnostic and can obviate the need for biopsy in most cases. The S-100 stain is helpful in identifying the Histiocytosis in SHML and there is no specific treatment present for this disease.

#### Case report:

Ten years male patient presented with history of swelling in the right side of neck of 25 days duration associated with low grade fever. On clinical examination swelling was 10x7.5cm in right anterior triangle of neck (Fig. 1). The swelling was non tender, soft to firm in consistency and mobile in all direction.

The peripheral blood cell count showed leukocytosis (17,000/mm<sup>3</sup>) with neutrophilia. ESR was raised to 45 mm after 1<sup>st</sup> hr. and serum

gammaglobulins were raised. Mantoux test was negative, X ray chest was normal. FNAC was done from right cervical node and smears were stained by May-Gruwald Giemsa stain. The smear showed sheets of histiocytes most of them showing intact lymphocytes in their cytoplasm, a feature known as Emperipolesis or Lymphophagocytosis (Fig. 2). On USG, abdomen did not reveal any organomegaly. On Immunocytochemistry the histiocytes were positive for S-100 and CD-68 thus confirming the diagnosis of Rosai Dorfman Disease.

Based on these characteristic FNAC findings, clinical features and immunocytochemistry a diagnosis of Rosai Dorfman disease was made.

Although this disorder is self limiting and in most cases undergoes complete spontaneous resolution but it can also follow a protracted clinical course for years. This patient was put on oral prednisolone 10mg t.i.d. in tapering doses for 21 days and patient showed marked clinical improvement.

#### **Discussion:**

SHML or Rosai Dorfman disease is a rare but well defined histiocytic, proliferative disorder of unknown etiology. The disease is thought to be a disorder of immune regulation or response to a presumed infectious agents (HHV-6/EBV) with its major manifestation in lymph nodes with resultant proliferation of sinusoidal histiocytes. The stimulation of monocytes/macrophages via Macrophage-Colony-Stimulating-Factor leads to immunosuppressive macrophages, which is considered as main pathogenesis of RDD<sup>5</sup>. It may occur at any age but mostly seen in young adults and children with a slight predilection for males (58%) and for individuals of African descent<sup>6,7</sup>. Clinically patients present as massive, painless, cervical

lymphadenopathy associated with low grade fever, leukocytosis, elevated ESR and hypergammaglobulinemia. Formerly it was thought to be a process limited to lymph nodes but in 40% of cases it also involves extra nodal sites. such as: Eyes and Ocular Adenexa, Head & Neck, Upper respiratory tract, Skin, Subcutaneous tissue, Bone, Skeletal muscles, CNS, GIT, Salivary glands, Genito urinary tract, Thyroid, Breast, Kidney, Liver, Heart and Uterine Cervix<sup>2,4</sup>. The onset of SHML is typically insidiously; the active phase is prolonged, there may be spontaneous remissions and relapses. Deaths have been reported in very few cases.

The histocytological features include a histiocytic proliferation with presence of large histiocytes showing fine vacuoles in the cytoplasm and large vesicular nuclei. The phagocytosis of lymphocytes and plasma cells (Emperipolesis) is characteristic feature<sup>9,10</sup>. This disease also exhibit some of the phenotypic features of dendritic cells such as S100, cathepsin E, fascin and at times CD 1a, helping in confirmation of diagnosis<sup>11</sup>.

Radiological and other imaging modalities are useful to investigate extra nodal involvement. SHML is generally regarded as benign self limiting disorder in spite of its propensity to form large masses and to disseminate to both nodal and extra nodal sites.

The common differential diagnosis include infectious lesions, reactive lymphoid hyperplasia with sinus histiocytes (RLHSH), Langerhans cell Histiocytosis (LCH), Hemophagocytic syndrome and Malignant lymphoma.

Although this disorder is relatively resistant to therapy, treatment options can be divided into surgical and radio-chemotherapy. Surgery is mainly indicated for life threatening conditions. In

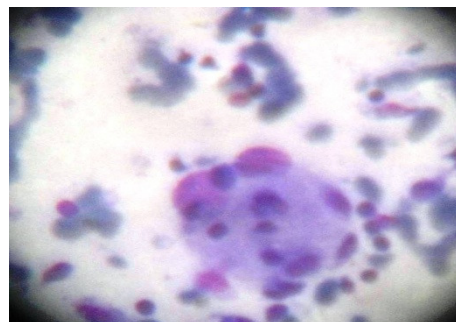
chemotherapy the most effective regimen include a vinca alkaloid combined with an alkylating agent and corticosteroid. Radiation therapy is also indicated for threatened function such as cord compression and respiratory embarrassment<sup>8,12,13</sup>.

### Conclusion

Rosai dorfman disease is characterised clinically by massive lymphadenopathy affecting mainly the cervical region. Being a rare entity clinical suspicion is difficult and it is mostly diagnosed by characteristic cytological features.



**Fig. 1: Neck Swelling**



**Fig.2: Smear showing Sinus histiocytes with Lymphocytophagocytosis (Emperipolesis) (MGG x 400)**

### References

1. Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy. Arch Pathol 1969; 87: 63-70.
2. Foucar E, Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy (Rosai Dorfman disease): Review of entity. Semin Diagn Pathol 1990;7:19-73.
3. Sanchez R, Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy. An analysis of 113 cases with special emphasis on its extranodal manifestation. Lab Invest 1977;36:21-2.
4. Goodnight JW, Wang MB, Sercarz JA, Fu YS. Extranodal Rosai Dorfman disease of the head and neck. Laryngoscope 1996;106:253-6.
5. Kong Y Y, Kong J C, Chen Z W, Wang J. Cutaneous Rosai Dorfman Disease: A Clinical and Histopathologic Study of 25 cases in China, Am J Surg Pathol 2007; 31(3): 341-50.
6. Sakai K, Koike G, Segichi K, Nakazato Y. Sinus Histiocytosis with massive lymphadenopathy: a case of multiple dural involvement. J Neurosurg 1984; 60:850-52.
7. Jo-Ann W, Andriko et al. Rosai Dorfman Disease isolated to the Central Nervous system. A report of 11 cases. Mod Pathol 2001;14:172-78.

8. Pulsoni A, Anghel G, Falcucci P, Matera, R Pescarmona, Ribersani M. Treatment of Sinus Histiocytosis with Massive Lymphadenopathy: Report of case and literature review. *Am J Hematol* 2000;69:61-71.
9. Jani PA, Banjan D. A case of Sinus Histiocytosis with Massive Lymphadenopathy ( Rosai- Dorfman Syndrome ) from Western India. *Mcgill J Med* 2008 11(2):156-59.
10. Lauwers Gregory Y, Perez-Atayde A, Dorfman R, Rosai J. The digestive system manifestation of Rosai Dorfman Disease (Sinus Histiocytosis with massive lymphadenopathy) review of 11 cases. *human path* 2000; 31:380-5.
11. Eisen R N, Buckley R J, Rosai J. Immunophenotypic characterization of sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease). *Semin Diagn Pathol* 1990; 7:74-82.
12. Raveenathiran V, Dhanalakshmi M, hayavadana Rao P V, Viswanathan P, Rosai Dorfman Disease: report of a 3 yer old girl with critical review of treatment options. *Eur J Pediatr surg* 2003;13: 350-4
13. Yjabali , MD, V Smrcka MD, J Pradna MD. Rosai Dorfman Disease: Successful longterm results by comination chemotherapy with prednisolone , 6 – mercaptopurine, methotrexate and vinblastine : acase report. *Int J Surg Path* 2005; 13: 285-289.