Rosai Dorfman Syndrome with Extranodal Manifestation

SS Bist*, Manisha Bisht**, S Varshney***, VP Pathak+

Abstract
Rosai Dorfman Syndrome (RDS) is a benign condition and a rare cause of cervical lymphadenopathy. It usually occurs in the first decade of life and manifest as massive enlargement of cervical lymph nodes. The disease has a benign course and involvement of the nasal cavity as an extranodal site is exceptional. A 22-year-old male presented as progressive massive bilateral cervical lymphadenopathy accompanied with nasal obstruction and occasional episodes of epistaxis. A FNAC from cervical lymph node and biopsy from nasal mass was compatible with RDS. ©

INTRODUCTION
In 1969, Rosai and Dorfman described 4 cases of a disease they called sinus histiocytosis with massive lymphadenopathy (SHML). Later in 1972, they analyzed 30 additional cases, establishing SHML as a clinicopathologic entity. RDS generally manifests in children or young adults with massive cervical lymphadenopathy, fever, leukocytosis, an increased erythrocyte sedimentation rate, and hyper-gammaglobulinemia. Other lymphatic groups, such as mediastinal, axillary and inguinal lymph nodes can also be affected. In about 25 to 40% of cases, extranodal sites are also affected. The concomitant involvement of one or more sites in the same individual is observed in up to 44.7% of cases. Extranodal involvement is often responsible for the most important clinical manifestation of the disease. The cause of RDS has yet to be established. Castleman’s disease, dermopathic lymphadenitis, mucocutaneous lymph node syndrome (Kawasaki’s disease), histiocytic necrotising lymphadenopathy (Kikuchi’s disease), vascular transformation of lymph nodes and inflammatory pseudotumor of the lymph node are among the other rare causes of lymph node enlargement. RDS is one more addition to this list.

CASE REPORT
A 22-year-old male presented with bilateral, massive, painless neck swelling for past six months. He complained of gradually increasing nasal obstruction on right side with occasional episodes of epistaxis for past three months. On examination lymph nodes were grossly enlarged in both anterior and posterior triangles which were multiple, firm, non-tender and non-matted with smooth surface (Fig. 1). Anterior rhinoscopy showed pinkish, irregular and friable mass filling the right nasal cavity, which was soft, non sensitive and bled when manipulated. Posterior rhinoscopy showed the mass in the right choana. Rest of ENT and systemic examination revealed no abnormality. Investigations showed HB-11 gm/dl, WBC count -13730/cu mm, N 72, L14, E14, ESR- 122 mm 1st/hr. Peripheral blood film showed microcytic, hypochromic red blood cells. X-rays Paranasal sinuses showed radio-opaque shadow in right nasal cavity with clear sinuses. Chest X-ray and USG scan of the whole abdomen was normal. A nasal biopsy was obtained under local anesthesia and histopathology revealed subepithelial tissue heavily infiltrated with large numbers of histiocytes showing emperipolesis (Fig. 2). FNAC of right and left cervical node showed many lymphocytes, plasma cells, immunoblasts along with histiocytes. Many of these histiocytes showed emperipolesis (Fig. 3). These features were consistent with a diagnosis of Rosai Dorfman syndrome. The patient was advised surgery for nasal mass under general anesthesia and the mass was excised endoscopically. The patient was treated with oral prednisolone 60 mg / day for a period of 8 weeks and then tapered off. Four months after surgery, the nasal cavity is free of any mass and the lymph nodes enlargements have regressed in size but still minimally palpable. The patient is under follow up.

DISCUSSION
RDS shows a worldwide distribution and can manifest...
in any age group. 81% of reported cases occur during the first and second decades of life and has a 2:1 male-to-female ratio. Our patient reported the occurrence of the disease during the third decade of life. The predominant clinical manifestation of the disease is massive cervical lymphadenopathy (87.3%) that, in most cases, is painless and bilateral, affecting one or all-cervical chains. Lymph nodes are isolated, mobile, and small during the initial stages but becomes adherent with disease progression, forming a voluminous multinodular mass. The axillary (23.7%), inguinal (25.7%), and mediastinal (14.5%) regions can also be affected, but always to a lesser extent than cervical involvement. Fever occurs in upto 30% of cases but was absent in our patient. Laboratory alterations are frequent and include anemia (65.7%), leucocytosis (59.1%), neutrophilia (68.4%), increased ESR (88.5%) and hyper-gammoglobulinemia (90%).

Our patient showed anemia, neutrophilia and raised ESR. The most common sites of extranodal involvement are skin (27.4%), nasal and paranasal cavities (26.8%), subcutaneous tissue (22.2%), orbit and eyelids (20.1%) and bone (18.4%). In extremely rare circumstances the central nervous system can be affected. Suprasellar involvement-mimicking meningioma has been reported. Our patient presented with extranodal disease confined to nasal cavity with involvement of bilateral cervical group of lymph nodes in anterior and posterior triangles. The patient had nasal obstruction and epistaxis due to the involvement of nasal cavity. The diagnosis of RDS is confirmed by histopathology and immunohistochemical studies. In lymph nodes, the sinuses are markedly dilated and crowded with histiocytes, lymphocytes and plasma cells. Histiocytes show abundant foamy cytoplasm, some of which show small lymphocytes in cytoplasm (emperipolesis). Immunoreactivity of the histiocytes for S-100 protein and CD-68 positive large histiocytes displaying lymphocyte phagocytosis are characteristically seen. However, these marker studies were not done in our patient. In general, extranodal involvement does not determine a more aggressive character or poor outcome; however, generalised lymphadenopathy, extranodal involvement of multiple organs (kidney, lungs and liver) and immunologic alterations lead to a poor prognosis. The causes of death due to RDS were reported to be a combination of cellular infiltration, mass forming ability of RDS and also may be due to defect in immune function.

The treatment modalities for RDS are nonspecific and include corticosteroids, chemotherapy with a combination of vinca alkaloids and alkylating agents, low dose interferon, antibiotics therapy, radiation therapy and surgical treatment with partial or total resection. Surgical option may be reserved for compressive symptoms, like airway obstruction, neurologic or ocular compressions, or severe deformation. However, the best treatment for RDS has yet to be established. In our patient, surgical excision of nasal mass was done with satisfactory result and cervical lymphadenopathy responded favorably to...
oral corticosteroids.

**CONCLUSION**

Massive cervical lymphadenopathy is the hallmark of Rosai Dorfman syndrome and head - neck region is the preferred site of the extranodal form of disease. The diagnosis of RDS is made on the basis of clinical suspicion and confirmed by histopathology. Clinicians and pathologists should always be aware of RDS in making a differential diagnosis of lymphadenopathy.

**REFERENCES**


---

**Announcement**


Chairman : Dr. RV Balar
Hon. Secretary : Dr. PB Joshi
Treasurer : Dr. S Panjwani
Executive Committee : Dr. RA Shridhrani
                  : Dr. RC Gupta
                  : Dr. HI Patwari
                  : Dr. D Gupta

---

**Announcement**

**National Conference on Medical Education (NCME 2007)**

**Building Capacity in Medical Education : A National Perspective, 15th to 17th November, 2007**
Organized by K.L. Wig Centre for Medical Education and Technology, All India Institute of Medical Sciences, New Delhi. Foundation for Advancement of International Medical Education and Research, (FAIMER), Philadelphia, U.S.A.

Conference Secretariat : **Prof. Rita Sood**, Organizing Chairperson and **Dr. A Shariff**, Organizing Secretary, K.L. Wig Centre for Medical Education and Technology (CMET), All India Institute of Medical Sciences, New Delhi 110029, India.
Contact e-mail : info@ncme2007.in
For details, please visit website : http://ncme2007.in