Rosai - Dorfman Disease – A Rare Cause of Cervical Lymphadenopathy

Ritesh Shelkar*, Vipin R. Ekhar**, Sachin Rane#, S.K.T. Jain***

*Assistant Professor, **Associate Professor, #Junior Resident, ***Professor and HOD, Department of Otolaryngology and Head-Neck Surgery
Indira Gandhi Government Medical College, Nagpur, Maharashtra, INDIA.

* Corresponding Address:
ritesnshelkar@gmail.com

Case Report

Abstract: Rosai-Dorfman Disease also known as Sinus Histiocytosis with Massive Lymphadenopathy (SHML) is a rare clinic-pathological condition. It is a benign condition which causes significant cervical Lymphadenopathy in children and young adults. The clinical Presentation varies from isolated nodal involvement to significant extra-nodal manifestations. The clinical features are usually mild but rarely life threatening complications can occur in some individuals depending on the site of involvement. Here we present two cases of Rosai-Dorfman Disease, both diagnosed on FNAC and Histopathology and responded well to steroids. One of the patients had extra nodal site involvement in the form of bilateral Nasal mass which is very rare.

Keywords: Rosai-Dorfman Disease, Cervical Lymphadenopathy, Nasal mass.

Introduction

Rosai-Dorfman Disease is a rare histiocytic disorder affecting various groups of lymphnodes in the human body. Rosai and Dorfman in 1969 described this disorder under the term Sinus Histiocytosis with Massive Lymphadenopathy (1). Majority of patients are children or young adults (2). Males are more commonly affected (3). Cervical Lymphnodes are commonly affected; however other lymphnodal groups like axillary, inguinal and mediastinal may also be involved. Extra-nodal involvement is seen in 25-40% cases. Various extranodal sites have been reported including the upper respiratory tract, gastrointestinal tract, Paranasal Sinuses, orbit and even meninges (4). In most of the cases patients are usually asymptomatic except for cervical lymphadenopathy however they may present with symptoms due to extranodal involvement and infiltration of vital organs. The disease usually has a self limiting, benign course and may not need any treatment. Episodes of remissions and exacerbations are characteristic however few patients may die from their disease.

Etiology is not exactly known however like any other histiocytic disorder responds well to systemic steroids. Histologically it is characterised by pericapsular fibrosis with dilated sinuses, heavily infiltrated by large histiocytes, lymphocytes and plasma cells. Emperipolesis is characteristic of lymphnodal involvement. Poor prognosis in the disease is due to wide spread dissemination and involvement of vital organs like kidney and liver or presence of immunological abnormalities. Otherwise the disease has a very stable and benign course.

Cases
Case 1
A 12 yrs old male, presented to us with complaints of slowly progressive bilateral neck swellings since one year. He also complained of bilateral nasal obstruction, insidious in onset and gradually progressive, since last 6 months. It was associated with intermittent nasal bleeds, spontaneous and self limiting. There was no history of fever, pain in throat, difficulty in deglutition, chronic cough, loss of appetite or loss of weight. On examination, there was 3X4 cm swelling present on both sides of neck involving both anterior and posterior triangles. The swellings were firm in consistency, non-tender, non-fluctuant, non matted and smooth in outline. The mass was freely mobile and not fixed to skin or underlying structures. Examination of the nasal cavities revealed bilateral pinkish, insensitive mass which bleed on touching. The nasal mass was more prominent on rt side compared to left side. Routine haematological examination showed anemia with Hb% of 7 gm%. TLC and DLC were within normal limits. ESR was Normal and Montoux Test was negative. USG Neck showed bilateral cervical Lymphadenopathy without caseation or matting. Radiograph of Chest and Ultrasonography of Abdomen were within normal limits. FNAC from the Cervical mass showed lymphocytes, plasma cells and histiocytes showing emperipolesis s/o Rosai Dorfman Disease. Endoscopic biopsy of the nasal mass also showed similar features and was typical of Rosai-Dorfman Disease. S100 stain was done to confirm the diagnosis. The patient was started on Tab. Prednisolone 1mg/kg/day for a period of 2 weeks which was then tapered over next 2 weeks period. The patient was kept on low dose oral steroids for a period of 4 months. He
involvement without lymphadenopathy is seen. Head and 

(4). Rarely, in less than 20% cases, isolated extra-nodal 

system, bones, orbit, central nervous system and breasts 

commonly involved extra-nodal sites include skin, 

mass. Extranodal involvement is seen in 25 – 40% and 

our patients, except for anaemia in first case. This finding 

90% patients. The systemic symptoms were not seen in 

anaemia. The systemic manifestations may be seen in 60-

ESR, hypergammaglobulineamia and sometimes 

systemic symptoms include fever, leucocytosis, increased 

inguinal and retroperitoneal may be involved. Other 

lymphnode groups like axillary, mediastinal, 

swelling, fever, difficulty in respiration, difficulty in 
deglutition, throat pain, loss of appetite or loss of weight. 

Local examination revealed bilateral 6X4cm swellings 

involving the anterior triangle of the neck. The lesion was 

firm in consistency, non-tender, non-matted, smooth and 

freely mobile. Ultrasonography of neck revealed bilateral 

cervical Lymphadenopathy with matting. Radiograph of 

chest and Ultrasonography of abdomen were non-

significant. CT neck was done and it showed bilateral, 

non-caseating cervical lymphadenopathy involving upper 

depth cervical nodes. Routine haematological 

investigations were normal. Montoux Test was negative. 

FNAC from cervical Lymphnode was highly suggestive 

of Rosai-Dorfman Disease; however biopsy was advised 

for confirmation. After ruling out Tuberculosis, Cervical 

Lymphnode Biopsy was done under General Anaesthesia 

and histopathology was diagnostic of Rosai-Dorfman 

Disease. Patient was started of systemic oral steroids 

1mg/kg/day for a period of one month and then low dose 

oral steroids for further 4 months. She responded well to 

treatment with drastic reduction in size of the swelling. 

She is under regular followup and doing well.

Discussion

Rosai-Dorfman Disease is a distinct benign histiocytic 

disorder, which presents in younger age group with 

massive cervical lymphadenopathy. The disease is 

worldwide in distribution and males are commonly 

affected with male to female ration of approximately 2:1. 

Other lymphnode groups like axillary, mediastinal, 

inguinal and retroperitoneal may be involved. Other 

systemic symptoms include fever, leucocytosis, increased 

ESR, hypergammaglobulineamia and sometimes anaemia. The systemic manifestations may be seen in 60-

90% patients. The systemic symptoms were not seen in 

our patients, except for anaemia in first case. This finding 

may be secondary to recurrent epistaxis due to nasal 

mass. Extranodal involvement is seen in 25 – 40% and 

commonly involved extra-nodal sites include skin, 

subcutaneous tissue, respiratory system, genitor-urinary 

system, bones, orbit, central nervous system and breasts 

(4). Rarely, in less than 20% cases, isolated extra-nodal 

involvement without lymphadenopathy is seen. Head and 

neck region is involved in 22% cases, nasal cavity being 

most common (2). Our patient had extra-nodal 

involvement in the form of bilateral nasal mass and nodal 

involvement in the form of bilateral cervical 

Lymphadenopathy. Diagnosis of Rosai-Dorfman 

disease is based on clinical suspicion and 

histopathological confirmation. Histologically there is 

infiltration of the tissue by lymphocytes, histiocytes and 

plasma cells. Demointration of emperipolesis i.e. 

engulfment of lymphocytes and erythrocytes by 

histiocytes, is usually diagnostic of Rosai-Dorfman 

Disease. Immunohistochemistry is usually necessary for 

confirmation of diagnosis. Characteristically S100 is 

always positive. Also some other markers like CD68, 

CD163, α1 antichymotrypsin and α1 antitrypsin may also 

be positive (2). Systemic symptoms in this disease may 

be related to enhanced production of such cytokines (5). 

In general the disease has a benign course and is self 

limiting. However massive lymphadenopathy and 

multisystemic involvement, especially vital organs like 

CNS, Liver, Kidney and lungs is usually associated with 

poorer prognosis. The treatment in Rosai-Dorfman 

Disease is non-specific and depends on the site of 

involvement. Isolated Lymphadenopathy may not be 

treated at all except for cosmetic reasons. However if any 

vital organs are involved or if the lesion is causing some 

obstructive symptoms or pressure symptoms, aggressive 

treatment may be indicated. The medical treatment 

includes corticosteroids, chemotherapy, low dose 

interferon, antibiotics and radiation therapy. But response 

to treatment is highly variable with repeated remission 

and exacerbation episodes. Surgical treatment may be in 

the form of partial or total resections. However surgery is 

usually limited to biopsy for confirmation. Debulking or 

excision may be reserved for compressive symptoms 

involving the upper respiratory tract, orbit or CNS. 

However the best treatment for Rosai-Dorfman disease is 

yet to be established. In our patients, surgery was limited 

to diagnostic biopsy and both our patients responded very 

well to systemic steroids.

Conclusion

Rosai-Dorfman Disease should be kept as a differential 

diagnosis in young patients presenting with massive 

cervical lymphadenopathy. High degree of clinical 

suspicion with typical histopathological features and 

immunohistochemistry are diagnostic. These patients 

respond well to systemic steroids and aggressive surgical 

treatment is needed only in life threatening complications. 

However it is necessary that both clinicians and 

pathologist keep this entity in their list of differential 

diagnosis for massive lymphadenopathy.
References