Doi: 10.32677/IJCR.2015.v01.i02.003 **Case Report**

Rosai Dorfman Disease – A case report

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ABSTRACT

Rosai Dorfman disease or Sinus histiocytosis with massive lymphadenopathy (SHML) is a benign, self-limiting disorder that commonly presents as massive bilateral and painless cervical lymphadenopathy. We present a case of a 6 year old male child who presented with cervical lymphadenopathy with no other complains and was diagnosed on fine needle aspiration cytology as a case of Rosai Dorfman disease. Patient was given low dose steroids to which patient responded well and became asymptomatic in 15 days.

Keywords: *Lymphadenopathy, Rosai Dorfman disease, Sinus Histiocytosis, Steroids*

Rosai - Dorfman disease (RDD) is a rare histiocytic disorder initially described as a separate entity in 1969 by Rosai and Dorfman under the term sinus histiocytosis with massive lymphadenopathy (SHML) [1]. SHML is a rare self-limited pseudolymphomatous disorder of unknown etiology usually presenting with cervical lymphadenopathy, fever, elevated ESR and hematological abnormalities [2]. Besides lymph nodes, the disease can also involve extra nodal sites such as skin, pancreas, thyroid, and kidney [3-6]. The cause of RDD is not fully understood, and treatment strategies can be different according to the severity or vital organ involvement.

CASE REPORT

A 6 year old male child, second by birth order, presented with history of multiple swellings in left side of the neck for 2 months which was painless and slowly increasing in size. There was no history of fever, cough, weight loss, bone pains or night sweats. There was no history of contact with tuberculosis. On examination, patient was well-nourished, afebrile, conscious and well oriented with stable vitals. There were multiple lymph nodes in the left parotid and submandibular area, largest measuring 2x2 cm and 3x3 cm respectively. All the lymph nodes were mobile, not fixed to underlying structures and were non-tender (Fig 1). No other lymph nodes were palpable.

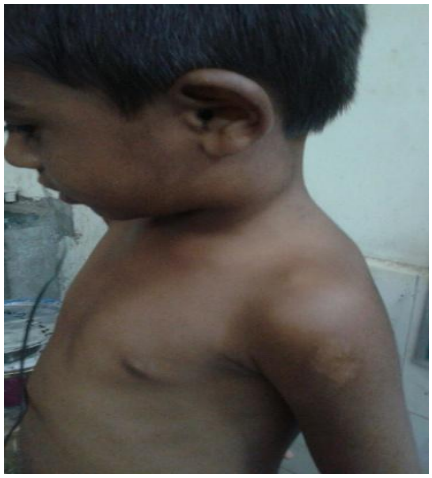

Figure 1 - Child with cervical lymphadenopathy

On laboratory investigations, his haemoglobin was 11.4gm%, total leucocyte count was 13,200/mm^3 and platelet count was 250,000/mm^3. His ESR was 26 mm after 1 hour and Mantoux test was negative. Fine needle

aspiration cytology (FNAC) of two lymph nodes was performed which revealed presence of large number of histiocytes throughout the smears. Histiocytes had multiple nuclei, but no nuclear atypia or nuclear grooving was seen. These histiocytes had abundant clear cytoplasm and exhibited numerous intact lymphocytes (emperipolesis) and plasma cells. Background had mature lymphocytes, plasma cells, neutrophils, and macrophages (Fig 2).

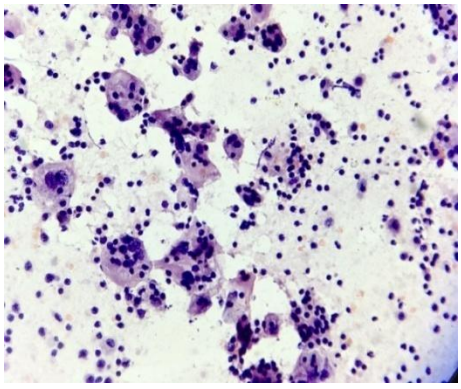

Figure 2 – FNAC of lymphnodes showing large number of histiocytes

Based on this characteristic cytomorphology, diagnosis of Rosai-Dorfman was made. Both CT brain and HRCT thorax, done to rule out involvement of any other site, were normal. Patient was started on low dose steroids (prednisolone 5 mg) twice daily to which patient responded very well. Size of the swelling decreased significantly and after 15 days was not visible on inspection (Fig 3).

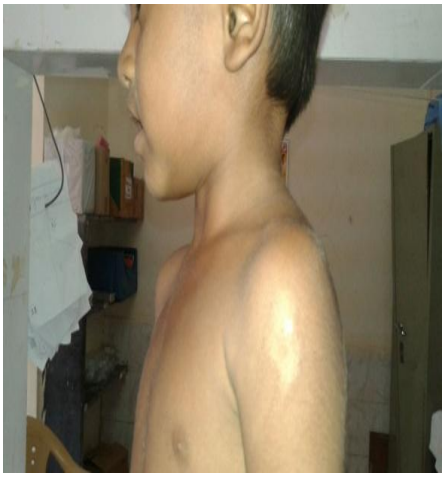

Figure 3 – Child after steroid therapy

Patient was instructed to continue steroids for 1 month and after 1 month of follow up, only 3 sub-centimetre lymph nodes were palpable. Steroids were stopped after one month and patient was followed for six months which did not show any recurrence.

DISCUSSION

Rosai - Dorfman disease is a benign condition usually affecting individuals in the first two decades of life with a slight male predominance. Exact pathogenesis of this condition is not known, but many factors were shown to be associated with the disease such as infection with Human Herpes Virus 6 [7] and Epstein Barr Virus [8]. RDD has been reported following bone marrow transplant for precursor-B acute lymphoblastic leukemia [9], and concurrently or after Hodgkin's and non-Hodgkin's lymphoma [10]. The clinical features described in the early reports were fever, weight loss, night sweats and massive bilateral cervical lymphadenopathy; however, later involvement of extra nodal sites such as skin, head, and kidney, thyroid, liver, pancreas, and lungs have also been reported [3-6].

Clinical course of RDD is unpredictable with episodes of exacerbation and remissions that could last many years. The disease is often self-limiting with very good outcome; nevertheless, 5-11% of the patients die from their disease. Various treatment strategies have been suggested for RDD such as low dose corticosteroids, methotrexate, 6-mercaptopurine, and thalidomide depending on the site involved [11-13]. For disease limited to lymph nodes only, low dose corticosteroids are found to be very effective [11]. Our patient was having only lymphadenopathy and no other site was involved, so we started low dose steroids and patient responded well. Other modalities of treatment are usually reserved for patients not responding to low dose steroids.

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