An Unusual Case of Lymphadenopathy

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ABSTRACT

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Rosai Dorfman Disease is a rare benign lymphoproliferative disease which is an unusual cause for lymphadenopathy. We report a case of 16 year old male who presented with massive bilateral cervical lymphadenopathy of 11/2 months duration. Blood investigations were normal except for raised ESR. Excision biopsy was done. Histological examination revealed mixed cellularity consisting of lymphocytes, plasma cells, numerous histiocytes exhibiting marked emperipolesis; suggestive of Rosai-Dorfman Disease (Sinus histiocytosis with massive lymphadenopathy). He was started on low dose steroids. The patient is being followed up; the case is being reported for its rarity.

Keywords: Rosai Dorfman Disease, Sinus Histiocytosis, Emperipolesis, Cervical Lymphadenopathy

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INTRODUCTION

Rosai-Dorfman disease was first reported in 1969 by Rosai and Dorfman as a distinct entity" Sinus histiocytosis with massive lymphadenopathy".[1,2] It is a rare benign idiopathic proliferative disease of phagocytic histiocytes.[1] Usually seen in children and young adults with male predominance.[3] The most common clinical presentation is painless massive cervical lymphadenopathy with no systemic symptoms.[2,3,4] Extranodal manifestation have been reported to correspond to about 40 % of cases.[2,3] Treatment is not necessary in majority of patients due to spontaneous regression.[5]

CASE REPORT

A 16 year old boy presented with bilateral cervical lymphadenopathy of 11/2 months duration **(figure 1).** He had associated loss of weight and appetite. No history of fever. On examination he had bilateral multiple enlarged cervical lymph nodes, largest measuring 5*4 cm, which was firm and nontender and few enlarged

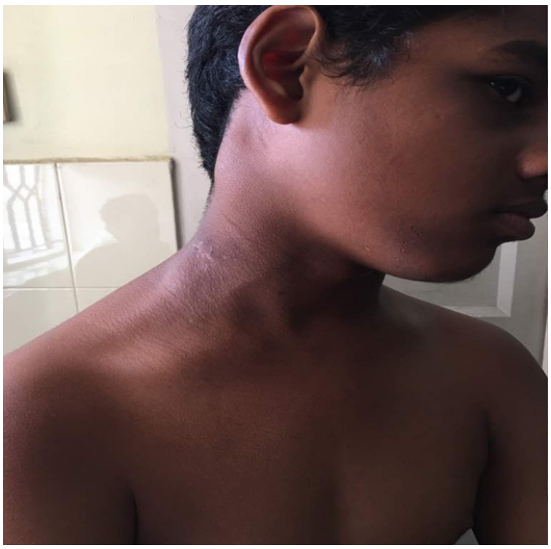

Figure 1. **Cervical Lymphadenopathy**

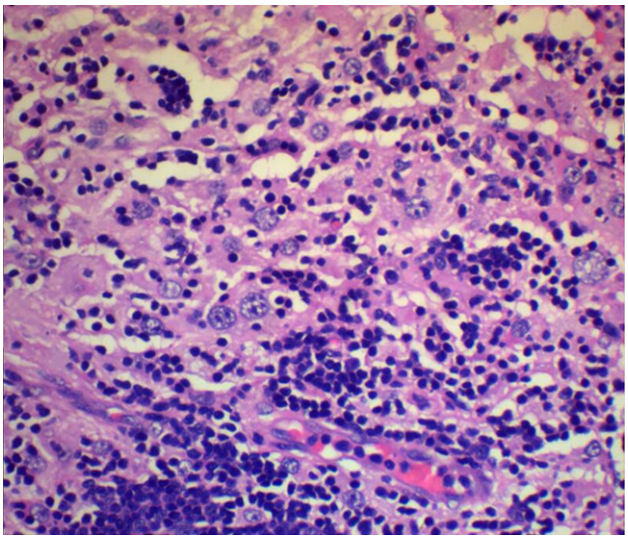

Figure 2. **Histiocytes showing Emperipolesis**

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axillary lymph nodes. Systems were clinically within normal limits. Routine investigations were normal except for raised ESR of 52 mm/1st hour. Peripheral blood smear and ultrasonography of abdomen was normal. Ultrasound of the neck showed noncaseating bilateral cervical lymphadenopathy. The lymph node was biopsied. Histopathological examination revealed architectural disruption with polymorphic infiltrate composed of lymphocytes, plasma cells and numerous histiocytes typically exhibiting emperipolesis suggestive of Rosai Dorfman Disease **(figure 2).**

DIAGNOSIS AND TREATMENT

A final diagnosis of Rosai Dorfman Disease (Sinus Histiocytosis with Massive Lymphadenopathy) was made based on histology showing the characteristic emperipolesis of histiocytes. He was initiated on low dose corticosteroids for the involution of nodes and asked for regular reviews.

DISCUSSION

In 1969, Juan Rosai and Ronald Dorfman described a benign idiopathic histiocytic disorder presenting as massive lymph node enlargement.[1,2] Since 1969 till now around 423 cases have been reported.[1] Usually seen in children and young adults.[3] The mean age at presentation is 20.6 years with slight male predilection, ratio of 1.4:1.[3,4] Etiopathogenesis is still unknown, could be related to viral infections like Herpes virus 6 (HHV-6) and Epstein-Barr virus (EBV) and a disorder of immune regulation.[3,4]

Typical clinical presentation is painless cervical lymphadenopathy seen in 87% of cases.[5] Extranodal disease is typical, with the most common sites being the skin and the central nervous system.[6,7] Other sites include soft tissue sites, genitourinary tract, upper respiratory tract, gastrointestinal tract, thyroid, head and neck region and orbit.[6,7] Cases of isolated RDD have been reported especially cutaneous Rosai Dorfman disease. CNS involvement can present as pachymeningitis or as hypothalamic pituitary axis dysfunction.

Definitive diagnosis is based on histology whose cornerstone of identification is emperipolesis and by Immunohistochemical analysis.[1,3] Emperipolesis refers to the presence of histiocytes containing intact lymphocytes within their cytoplasm; ie lympho phagocytosis. Immunohistochemical staining for S100 is diagnostic; cells will stain positive for CD25, Ki67 and CD68 and negative for CD1a, aids in distinguishing RDD from Langerhans cells.[2,3]

Differential Diagnosis to be considered includes lymphoreticular malignancies like lymphoma, Hodgkin's disease, malignant Histiocytosis and monocytic leukemia.

Presently RDD is considered as a benign clinical entity showing spontaneous regression. Hence majority require no treatment.[1,4] Surgical resection remains the mainstay of treatment for massive lymphadenopathy or extranodal involvement compressing or within vital organs. Corticosteroids can be used for the involution of the nodes.[3] Other treatment options include chemotherapy, radiotherapy and immunomodulation. In 5 % it can progress and rarely can have a fatal course

CONCLUSION

A high degree of suspicion is necessary to diagnose this rare clinical entity. It is a benign condition mostly requiring no treatment. Definitive diagnosis is by histology and immunohistochemistry.

The case is being reported because of its rare occurrence.

END NOTE

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Conflict of Interest: None declared

Editorial Comments: This article is selected for the rarity of the case and unusual presentation. Clinicians need to be familiar with these unusual causes of lymphadenopathy.

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