

Case Report

Juan-Ron fever: a rare case report

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ABSTRACT

Juan-Ron fever named after Jaun Rosai and Ronald Dorfman is the fever associated with Rosai Dorfman disease also known as Sinus Histiocytosis with Massive Lymphadenopathy (SHML). It is a rare disorder of unknown etiology that is characterized by abundant histiocytes in the lymph nodes throughout the body. Usually patient presents with painless lymphadenopathy. We present a case of a 45 year old male who presented to us with bilateral cervical lymphadenopathy and fever, later on diagnosed to have SHML.

Keywords: Rosai Dorfman disease, SHML, Histiocytes, Lymphadenopathy

INTRODUCTION

Rosai Dorfman Disease (RDD) or Sinus Histiocytosis with Massive Lymphadenopathy (SHML) is a rare disorder that typically manifests as lymphadenopathy and systemic symptoms.¹ Sometimes systemic symptoms may resemble the B symptoms of lymphomas masquerading malignant lymphomas. The characteristic histopathologic feature of the lymphnode shows lymphophagocytosis or emperipolesis, and immunohistochemistry shows S 100 positivity.

CASE REPORT

A 45 year old male presented with history of intermittent fever since 2 months and progressively increasing painless masses in the neck over a period of 1 month duration. The fever was associated with drenching night sweats. There was no history of any significant weight loss. There was no history of cough, breathlessness, abdominal pain, bone pains, fatigue.

On examination his vitals were stable. He was febrile. Pallor and bilateral cervical and axillary lymphadenopathy was present. The lymphnodes were

firm, non-tender, approximately 2 to 3 cm in size, mobile and discrete. There was no sternal tenderness. Other systems examination was normal.

Hematological investigations revealed Hb - 8 g/dL, TLC - 9600/mm³, DLC - P82, L8, M9, E1, Platelets - 2.16 lacs/mm³, PS - mild anisocytosis with hypochromia, ESR - 98 mm in 1st hour. Blood culture did not grow any organism. Elisa for HIV was negative. Mantoux test was negative. USG abdomen was normal. CT scan of chest and abdomen was normal. Histopathological examination of a left cervical lymph node biopsy displayed dilated sinuses in the parenchymal region with extensive infiltration consisting of histiocytes and chronic inflammatory cells (Figure 1). A large number of lymphocytes could be seen in cytoplasm of histiocytes suggesting lympho-phagocytosis - emperipolesis (Figure 2). Lymphoid follicles were hypertrophied and showed germinal activity.

Immunohistochemical staining of histiocytes was positive for S-100 gene product expression (Figure 3). In view of the above mentioned observations diagnosis of Rosai Dorfman Disease (RDD) or Sinus Histiocytosis with Massive Lymphadenopathy (SHML) was made.

A trial of oral prednisolone in a dose of 40 mg/day was started. There was a dramatic response in form of disappearance of fever and significant decrease in the size of the lymphnodes occurred in 1 week of therapy. The patient was discharged with a steroid taper over next 2 weeks and is awaiting follow-up.

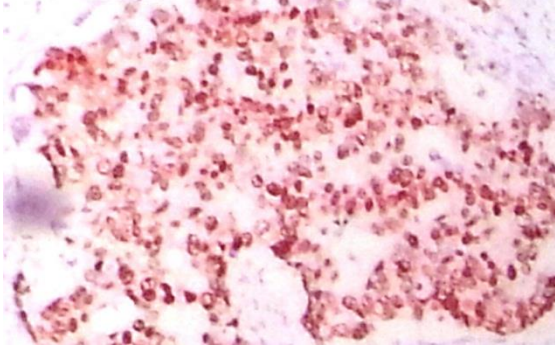


Figure 1: H&E stained slide from cervical lymphnode biopsy showing effaced architecture with diffuse sheets of histiocytes.

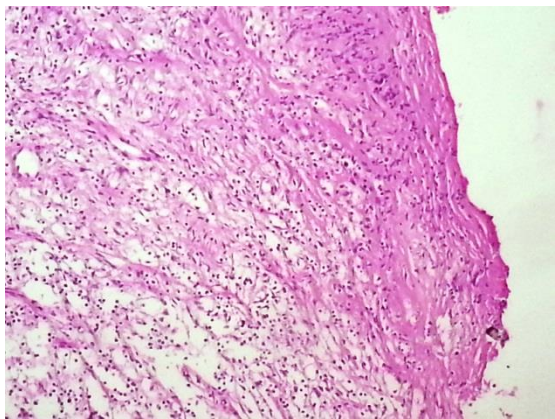


Figure 2: H&E stained slide predominant lymphohistiocytic population with the central area showing lympho-phagocytosis (emperipolesis) in the cytoplasm of histiocyte.

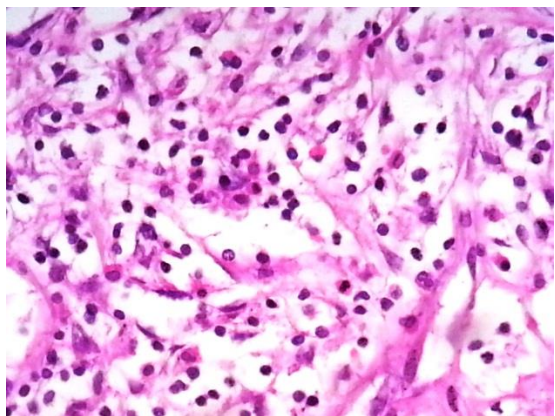


Figure 3: IHC staining (Dako kit) 40x view showing S-100 positive histiocytes (predominantly nuclear with occasional cytoplasmic positivity).

DISCUSSION

Although RDD may occur in any age group, it is most frequently seen in children and young adults.² RDD is rare in Indian population with less than a dozen cases being reported in children and none in adults.³ This is a rarest case of RDD which manifested in an adult. Males are commonly affected.⁴ RDD has been reported following bone marrow transplant for precursor-B acute lymphoblastic leukemia and concurrently or after Hodgkin's and non-Hodgkin's lymphoma.⁵ It is stated that various conditions ranging from autoimmune diseases, hematological malignancies and viral infections especially with Epstein-Barr Virus (EBV) trigger a cytokine-mediated migration of monocytes leading to histiocytes accumulation and activation. However, there is no strong evidence for this at the moment.^{6,7}

The most frequent clinical presentation of RDD is a massive bilateral and painless cervical lymphadenopathy with fever, night sweats and weight loss. Mediastinal, inguinal and retroperitoneal nodes may also be involved. Extranodal involvement by RDD has been documented in 43% of cases with the most frequent sites being skin, soft tissue, upper respiratory tract, multifocal bone, eye and retro-orbital tissue with lymphadenopathy or as an isolated initial manifestation of disease. Other sites include urogenital tract, breast, gastrointestinal tract, liver, pancreas and lungs. Head and neck involvement has been reported in 22% of cases, most commonly the nasal cavity followed by the parotid gland. As in our case conditions like lymphoma, leukemia, tuberculosis, sarcoidosis, HIV, infectious mononucleosis that presents with fever and lymphadenopathy have to be ruled out.

Biopsy in such cases is the next best step. The differential diagnosis of a chronic inflammatory infiltrate containing numerous large histiocytes includes granulomatous diseases such as Wegener's granulomatosis, sarcoidosis, Hodgkin's disease, and Langerhans' Cell Histiocytosis (LCH). The classical finding of emperipolesis (active penetration by a smaller cell into or through a larger cell) differentiates it from other diseases.⁸ SHML cells constantly express the S-100 protein which differentiates it from lymphomas and Langerhans Cell Histiocytosis (LCH).

ESR is elevated in 88.5% of cases of RDD and was raised in this case too.

RDD patients can be subdivided into three categories:

- 1) Patients with only lymph nodes that enlarge suddenly with spontaneous regression and without any further recurrences.
- 2) Patients with immunologic abnormalities at presentation who have a more widespread nodal disease and a higher fatality rate.⁹

- 3) Patients with several extranodal site involvement, multinodal disease and a protracted clinical course with multiple relapses and remissions for years. In these cases, the severity of disease depends on the type and number of extranodal sites.¹⁰

Active management of RDD is not required unless the disorder becomes organ or life threatening since it is mostly self-limiting. There is sometimes a dramatic response to oral short course prednisolone in steroid responsive RDD as in this case.¹¹ If required, in multimodal and extra nodal systemic disease vinca alkaloid in conjunction with alkylating agent and corticosteroid provide the most efficient chemotherapeutic regimen besides radiation therapy which is also used.¹²

In conclusion, RDD indeed is a rare finding, especially in Indian subcontinent. Physicians need to have a high degree of suspicion in patients presenting with generalized lymphadenopathy, with or without extranodal involvement, to diagnose RDD. Indolent course, presence of histiocytes with emperipolesis in inflammatory exudates and S100 positive immunohistochemical staining clinch the diagnosis in favour of Rosai Dorfman disease or sinus histiocytosis.

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