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ROSAI DORFMAN SYNDROME- A RARE CASE REPORT IN AN ASIAN MALE

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Abstract

Rosai–Dorfman disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy, is a benign, self-limited, idiopathic proliferative histiocytic disorder originally described by Destombes in 1965 and later in 1969 by Rosai and Dorfman. In its typical form the disease is characterized by extensive cervical lymphadenopathy associated with fever, polyclonal gammopathy and leukocytosis with neutrophilia. The skin is the most common site affected. Extranodal manifestations have been reported in 43% of cases. In the present case report a young male from Asian origin diagnosed to be suffering from RDD is discussed.

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INTRODUCTION

RDD is a rare pseudolymphomatous disorder of unknown aetiology which is classified as non-Langerhans cell histiocytosis. The disease can affect any age group but young adult menin the first and second decade are more susceptible to it. It rarely affects people of Asian origin. It has tendency to affect lymph nodes in the head and neck but can also affect extranodal sites like skin, soft tissue, central nervous system and less commonly gastrointestinal system. [1]

CASE REPORT:

A 23 year old young man came to medicine outpatient department with complaints of fever and swelling around the neck for the past 15 days. Fever was intermittent in onset, of moderate degree and no history of chills and rigors, cough, running nose, burning micturition, nausea and vomiting. He was a chronic smoker and alcoholic and was working in cotton industry. His past history revealed that he had swelling around the neck for the past five years and he was suspected to be suffering from extrapulmonary tuberculosis and was on steroids and immunosuppressant.

On examination he was febrile with temperature of 102°F and had bilateral enlargement of cervical and submandibular lymphnodes, no discharge, nontender and no other lymph nodes were palpable. On systemic examination cardiovascular system, respiratory system, central nervous system and per abdomen findings were normal.

Laboratory findings revealed RBC count 4.57 million/cumm, WBC count 6,000 million/cumm, PCV- 38.5%, MCV-84fl, MCH-25.9 pg/L, MCHC-30.8g/dl, platelet count-3.17lakh/cumm. Peripheral smear revealed RBC- normocytic normochromic, WBC- increased on the smear with neutrophilic predominance. Platelets adequate on smear and no parasites

were found. A probable diagnosis of neutrophilic leucocytosis was made.

Biochemical findings revealed Random blood glucose of 120mg/dl, blood urea of 30mg/dl, serum creatinine 0.7mg/dl, total cholesterol 150mg/dl, triglycerides 100mg/dl, HDL 35mg/dl, LDL 95mg/dl, VLDL 20 mg/dl, serum amylase 61U/L.

DISCUSSION:

RDD is a disorder of non malignant histiocytes that infiltrate lymph nodes or extranodal tissues. The cells exhibit emperipolesis, the nondestructive phagocytosis of lymphocytes or erythrocytes. The signs and symptoms vary from one person to another depending on the extent of disorder and organ affected. The skin is the most common extranodal site involved especially of head and neck region.[2] The affected individuals exhibit painless swelling or enlargement of affected lymph nodes mostly of cervical origin and some individuals have nonspecific symptoms like loss of weight, fever, pallor, rhinitis and malaise. In extremely rare cases liver and spleen enlargement is seen. Fine needle aspiration cytology plays a role in diagnosis. Aspirate from affected lesions show proliferation of histiocytes with abundant eosinophilic cytoplasm, vesicular nuclei and lymphophagocytosis or emperipolesis.[3]

As the exact aetiology was not known many conclusions were made like it may be because of immune dysfunction or infections like herpes, parvo virus and Epstein Barr virus infection and in particular HHV-6 expression was seen on histiocytes.RDD was reported in patients with immunoglobulin G4 related disorders but later no evidence was suggestive of it. [4]

The laboratory investigations should include liver function tests, renal function tests, anti-nuclear antibody, rheumatoid factor, screening for HHV6, HHV8, HIV and EBV viruses and contrast CT scan should be done for extra nodal organ involvement. In 90%

patients elevated ESR, neutrophilic leucocytosis and polyclonal gammopathy is seen. The role of bone marrow biopsy is unclear.

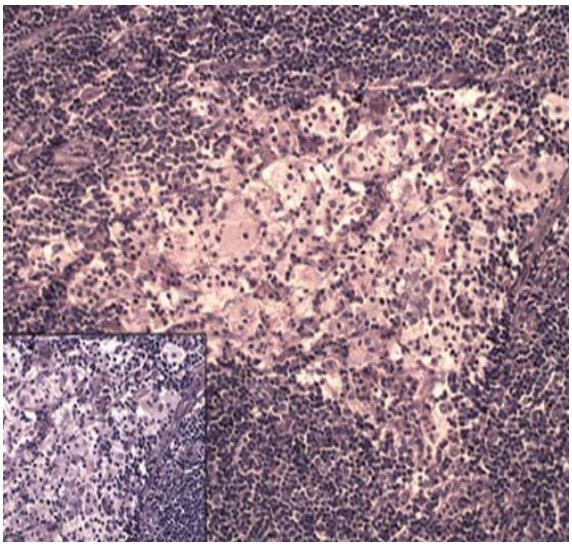

Figure1: Lymph node biopsy: large amount of histiocytes with emperipolesis

The differential diagnosis of RDD includes lymphomas, malignant histiocytosis, disseminated tuberculosis and Langerhans cell histiocytosis and hallmark of RDD is emperipolesis which differentiates it from other disorders. [5] It is a self-limiting disorder in majority of cases, unless there is organ involvement. No precise treatment is available although chemotherapy, radiotherapy, interferon and glucocorticoids have been tried. [6]

CONCLUSION:

RDD or sinus histiocytosisis a self-limiting benign disease with nodal, extranodal and organ involvement. It may mimic a plethora of malignant neoplasms so unnecessary interventions to the patients can be avoided. The present case is rare in that it was seen in Asian male patient with unusual presentation.

CONFLICT OF INTEREST: All the authors declare that they have no competing financial interest.

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