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Case Report

Rosai Dorfman Disease with Autoimmune Hemolytic Anemia

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Abstract

Rosai-Dorfman disease is a rare histiocytic disorder with massive lymphadenopathy. Approximately 80% of patients present with painless massive cervical lymphadenopathy. Isolated extranodal involvement is relatively uncommon [1]. We present a case of 18 months old Sudanese boy who presented with yellowish discoloration of skin, sclera and pruritic skin rash with dark colored urine associated with fever and poor weight gain diagnosed with histopathology and immunochemistry studies as a case of Rosai-Dorfman disease. Patient got treated with low dose corticosteroid.

Keywords: *Rosai-Dorfman disease; Lymphadenopathy; Jaundice; Autoimmune hemolytic anemia*

Introduction

Rosai-Dorfman disease (RDD) characterized by non-neoplastic proliferation of histocytes/phagocytes in dilated sinusoids of lymph nodes and in extranodal tissues. Initially described as a separate entity in 1969 by Rosai and Dorfman [2]. RDD can occur at any age group, most frequently affects children and young adults [3]. It is more common in males and in individuals of African descent [4]. It is a self-limiting pseudolymphomatous disorder. The causes of RDD are not known. Usually presenting with cervical lymphadenopathy, fever, elevated ESR and hematological abnormalities [5]. Most often the RDD takes a benign course and treatment is not necessary [6,7].

Case Report

18 months old Sudanese boy not known to have any medical illness before, on 23rd September 2015, he referred from other hospital. He was admitted there for 1 month and referred to us on 24th November 2015 for further workup and treatment. Patient 3 months prior to presentation developed low grade fever undocumented for 1 week then became high grade documented 39 constant all the day, decreased partially by antipyretic, associated with night sweating, poor weight gain, loss of appetite, decrease activity, abdominal distension and pruritic skin rash Yellowish discoloration noticed by the mother in the sclera then progressed to all over the body associated with Cola like colour urine and hypopigmentation skin lesion in all limbs specially upper limb and forehead with history pruritic rash 3 to 4 months back, NO history of abnormal movement, NO Cough, Shortness of breath or Chest pain, NO Abdominal pain, Diarrhoea, vomiting, constipation or change colour of stool or limitation of movement, No bleeding from any site, No Joint pain, swelling, limitation of movement. He is a product of Full Term, Spontaneous vaginal delivery, NO NICU admission, he was admitted in a private hospital for 2 days then referred where he's admitted for 1 month, received blood transfusion many time during Hospitalization and given vancomycin and ceftazidime. He was on exclusive breast feeding till 14 months of age. Vaccination: up to date. Development: up to age. He is 1st baby in the family, with 1st degree consanguinity. No history of contact with sick patient, No history of contact with T.B patient, No history of haematological disease, malignancy or hereditary disorder, No History of recent travel to other country. They live in rural area, Low socioeconomic status.

The father work as accountant, the mother housewife. On Examination: Patient looks conscious, alert, irritable, pale, deeply icteric (green olive), not distress or cyanosed, not dysmorphic.

Vital sign: Temperature 36.9°C, pulse 131, blood pressure 101 /58 (66), spo2 = 100% in room air. Height: 80 cm 50%, Weight: 10 kg 25%. Chest and cardiovascular: were normal. Abdomen: distended, soft, lax, with hepatomegaly 4 cm and splenomegaly 3 cm below costal margin with no tenderness. Lymph node: discrete, mobile, non-tender lymph node different size largest one was 2 * 1.5 cm at Bilateral cervical, Axillary and inguinal area. Head and neck: clear.

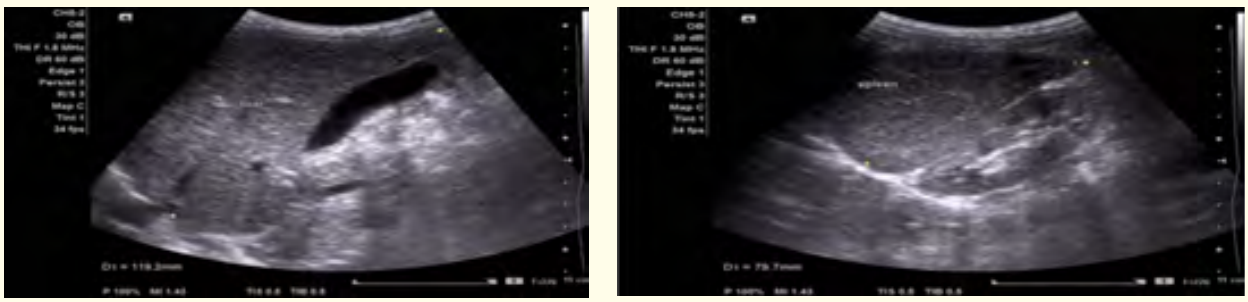
Investigation

Complete blood count	
WBC	33.9 10^3/µL
Neutrophil %	72 %
Lymphocyte %	18 %
RBC	3.219 10^6/µL
HgB	4.3 g/L
Platelet	219 10^9 /L
Reticulocyte %	11 %
Liver profile	
Bilirubin Total	473 mg/dl
Bilirubin Direct	400 mg/dl
Albumin	22 g/dl
aspartate aminotransferase	63 U/L
Alanine aminotransferase	22 U/L
Alkaline Phosphatase	102 U/L
Gamma-glutamyltransferase	20U/L
Coagulation profile	
Prothrombin time	13.7 second
Partial thromboplastin time	27.2 second
Iron study	
Iron	18.6 µg/dl
Total iron binding capacity	43.5 µg/dl
Ferritin	286.5 ng/mL

Immune electrophoresis			
IgG	27.26 mg/dl		
IgM	.80 mg/dl		
IgA	3.55 mg/dl		
Hemoglobin electrophoresis			
Hb A	96.2 %		
Hb A2	3.2 %		
Hb F	0.7 %		
Sickle test	-ve		
Direct combos test	+ve		
Cold agglutination test	+ve		
C-Reactive Protein	-ve		mg/dl
Lactic Dehydrogenase	722 U/L		
Erythrocyte sedimentation rate	150	122	120 mm/hr
Peripheral blood smear	Aniocytosis with some RBC agglutination, many polychromatic cell and NRBCs are seen. Some spherocyte and target cell are noted. Mild left shifted granulocytic cell. Monocytes are prominent. Some atypical lymphocytes are seen.		

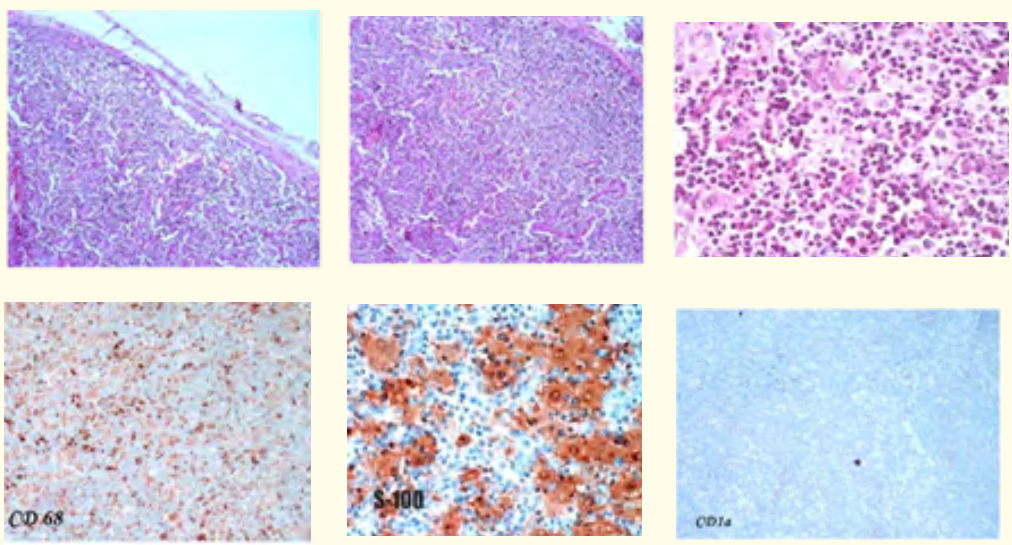
Other laboratory investigation: Mono spot test, HBsAg, HCAb, HIV, *Brucella*, *Leishmania*, *Mycoplasma*, and Malaria were all negative except for Paul Panel test was positive.

Chest X-Ray: was unremarkable. Abdominal Ultrasound: shows Liver mild enlarged of 12 cm, normal shape and echo pattern of the liver with no focal lesions seen. Normal portal vein and common bile duct. Spleen mild enlarged in size 7.9 cm, no focal lesion seen. Gallbladder, pancreas is normal. No ascites defined. Pan CT with contrast (not done in our hospital) report showed: No focal lesion, hepatosplenomegaly, Multiple enlarged Peri and Paraaortic, iliac mesenteric abdominal Lymph node, Bilateral enlarged inguinal Lymph node, Clear both lung field, Enlarged mediastinal Lymph node, Bilateral enlarged Axillary Lymph node.



Bone marrow biopsy: cellular bone marrow aspirate for age, showing active trilineage hematopoiesis with prominent erythropoiesis. Flowytometry, shows 28% T-lymphocyte with no phenotype evidence of aberrancy in the available monoclonal markers shows negative for CD1a, S-100, CD68, TdT, CD34, CD19, CD3, CD5, CD20 and Glycophorin-A were not available.

Excisional Biopsy: In pathology, Lymph node with capsular fibrosis an packing of sinuses by histocytes that some time multinucleated with many plasma cells and few eosinophil. The histocytes having central vesicular nuclei with abundant clear cytoplasm, showing lymphophagocytosis (Emperipolesis), marked plasmatic infiltrate is noted in connective tissue between sinuses. ImmunoHistochemical stain, Histiocytes are positive for S-100, CD68, Negative for CD1a, T-Lymphocyte positive for CD4, CD8.



Management: Patient started prednisone 2 mg/kg divided dose. Rapidly had achieved remission hematologically and clinically with disappearance of enlarged LNs and regression of hepatosplenomegaly but he required to be on low maintenance dose ever since as he has relapsed once shortly after interruption of steroid. Currently he completed 14 mo since the time of diagnosis and start of steroid on low dose < 0.5 mg /kg alternative days. No features of steroid toxicity so far, stable course of illness with no relapses. Not clear plan when to stop prednisone, yet watchful waiting for self-remission of the disease is possibly the appropriate approach as the course of the disease is unexpected and may take months to years for self-resolution and on low dose prednisone < 0.5 mg/kg.

Conclusion

Rosai–Dorfman disease is a nonmalignant histiocytic disorder that classically presents with massive, painless cervical lymphadenopathy, fever, and an elevated erythrocyte sedimentation rate. A high degree of clinical suspicion is needed to make the diagnosis because the differential diagnosis includes both malignancy and other histiocytic disorders [8]. Initially we suspect the patient has Autoimmune Lymphoproliferative syndrome Vs. Lymphoma so we did further workup investigation, included excisional biopsy to distinguish Lymphoma and other histiocytic disorder .In Autoimmune Lymphoproliferative syndromes massive lymphadenopathy with immune cytopenia but in our patient double negative CD4 CD8 T lymphocytes was within normal percentage. Lymphoma was excluded by morphological and immunophenotyping of the lymph nodes. Langerhans cell histiocytosis (LCH): CD1a was negative in our patient made LCH is less likely that lead us to our final diagnosis Rosi-Dorfman disease with autoimmune hemolytic anemia in remission.

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