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# Sinus Histocytosis with Massive Lymphadenopathy (Rosai-Drofman Disease) in Seven Years Old Child in Sudan with Hepatomegaly Case Report

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**Abstract** **Case Report**

Sinus Histiocytosis with massive lymphadenopathy (SHML) Rosai-Dorfman Disease is a rare, idiopathic, node-based histiocytic proliferative disorder affecting different age group, 10% of the incidence in children less than 10 years old and 80% in young age group less than 20 years old [1]. Higher incidence of the disease was reported in African and West Indian descent [1]. The disease usually presents with massive painless cervical lymphadenopathy as part of a generalized process involving lymph nodes and may involve extranodal sites independently of lymph node status. The head and neck region represents one of more common extranodal areas, particularly the Sinonasal tract [1]. Here we report the first presentation of 7 years old child with unusual presentation of sinus histiocytosis (Rosai –dorfman disease) and hepatospleenomegly in Sudan.

**Key words: RDD** Rosai-dorfman disease, hisitocytosis, lymphadenopathycervical lymph node swelling, atypical, mycobacterium.



## INTRODUCTION

Sinus histiocytosis with massive lymphadenopathy (SHML) Rosai-Dorfman is a rare disease having reported prevalence of 1:200.000 [5]. The disease was first recognized as a specific pathological entity in 1969 by Juan Rosai and Dorfman. The disease is more in male than female, more in young group than children.

SHML is a form of class II histiocytosis typically characterized by massive bilateral and painless cervical lymphadenopathy associated with systemic symptoms such as fever and weight loss this typical presentation referred as the systemic form or "classical "of the disease, the cutaneous form sharing the same epidemiological characteristi**c** with no involvement of tissue other than the skin[3, 4].

Cervical lymph nodes involved in 87% of case. Multiple lymph nodes maybe involved, and it manifests as enlarged, painless, frequently matted masses [2].

Extra nodal manifestations represent 25-40%, involving skin, upper respiratory tract bone and CNS.(2) It's also associated with fever, leukocytosis, anemia, elevated ESR, polyclonal hypgammaglobulinemia. It may cause death due to amyloidosis involvement of a vital organ or immunologic abnormalities [2].

## CASE PRESENTATION

7 years old male child present to pediatric department at Omdurman military hospital with progressive painless bilateral cervical lymphadenopathy and high grade fever for one month.

His parents reported that the bilateral anterior neck swelling has been increasing in size for the last one month; the neck swelling was not associated with any pressure symptoms, shortness of breath, cough, chest, bone or abdominal pain.

On /E the child was febrile (Temp is 39c, not pale. There was Small bilateral sup maxillary cervical lymphadenopathy, 5-1c in size, not tender, not matted and no skin change over it. Inflamed throat. Investigation shows: Normal CBC. So considered as simple pharyngitis, reserved oral antibiotic for 10 days Amoxiclv. Two weeks later the child represent again to the pediatric department with non-resolving high grade fever and painless multiple neck swelling. The fever not

subsided with the antibiotic and the masses number increased. They deny any pressure symptoms, shortness of breath, cough, night sweating, Wight loss, abdominal pain or anorexia.

There was no history of similar condition, atopy,asthma, eczema, or any evidence of autoimmune disease, or repeated hospital admission. The parent denies any family history of similar condition.

On /E: the child was febrile temperature was 39c, ill, not pale. All groups of the cervical lymph node was enlarged, oracular, occipital, and supraclavicular.No organomegally detected.

**Note:** Axillary group was speared.Investigation showed:
*Complete blood count showed high total white B cells and high lymphocyte count.
*Blood culture showed no growth.
*Fine needle aspiration (FNA) showed reactive granulomatous formation, Advised for executional biopsy.
**considered as atypical mycobacterium received macrolide oral antibiotic erythromycin for 10 days.

10 days later the child represents again with non-resolving high grade fever, weight loss, profuse sweating. On /E: the child was febrile temperature was 39c, ill, not pale, his weight dropped from 19 kg to 17 kg .and generalized lymphadenopathy.

On GIT examination abdomen was soft, no superficial or deep masses, liver enlarged 12-14 cm on size, slightly palpable spleen .other systemic review was insignificant to any abnormality in respiratory, cardiovascular, genitourinary, Central nervous system or musculoskeletal.

*CBC: showed high total b cells, and high lymphocyte count
*Abdominal ultrasound scan showed liver enlargement, smooth 12-14 cm in size. Mild Spleen enlargement 1-2cm and para aortic lymph node.
*blood cultureshowed no growth.
*L ymph node biopsy showed

Microscopically shows the lymph node is distended by aggregated of histocytes admixed with lymphocyte, plasma cells, and neutrophils, thereis no evidence of granuloma or malignancy in section of biopsy submitted. The feature is consistent with sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease).

## DISCUSSION

Sinus histiocytosis with massive lymphadenopathy Rosai- Dorfman disease (RDD) is an uncommon, benign disease of unknown etiology, the recent studies indicate that it may be caused by the development and expansion of abnormal Langerhans cells that subsequently lead to accumulation of the other cells of the immune system [6].

Sinus histocytosis has two form of presentation, the" classic" systemic form involving the cervical lymph node commonly and other exrnodal system, and the cutaneous form involving only the skin. Cervical lymphadenopathy is present in over 90 % of patients affected by RDD. Typically, it is painless, bilateral and frequently massive, with single nodal elements measuring up to 6 centimeters [16].

However, any group of lymph nodes cans beinvolved. Axillary and inguinallymph nodes are enlarged in 38%and 44 % of cases, respectively**.**

Mediastinal and hilar nodes are involved in approximately 30 % of patients. In a minority of cases, retroperitoneal lymph node localizations have also been described [7]. However in our case the cervical group and the parotic lymph node is involved extranodal involvement has been documented in 43 % of cases [8].

The most commonly affected sites are the upper respiratory tract, skin, eyes and retro-orbital tissue and bone tissue. Central nervous system is less frequently involved. Localizations of RDD in lungs, urogenital andgastrointestinal tract, breast, thyroid, and even heart have also been reported [6, 9, 10, 11].

The onset of the disease is generally subtle, with an average of 3–6 months between the beginning of signs and symptoms and the diagnosis.

Different symptoms can be present, including fever, malaise, weight loss, and night sweats [12]. In case of extranodal localizations, the clinical picture will depend on the affected organ or apparatus. Laboratory findings in RDD are non-specific include leukocytosis, elevated ESR and CRP levels, and polyclonal gammopathy. Normochromic normocytic anemia and elevated serum ferritin levels have also been reported. Less common positivity for rheumatoid factor and antinuclear antibodies, and a reversal of the CD4/CD8 ratio in peripheral lymphocytes [13].

There are also reports on RDD complicated by the development of autoimmune hemolytic anemia, especially in children, for which the pathogenesis mechanism is not known [14]. Ultrasound can also be helpful. Definitive diagnosis can only be made by histological analysis of affected lymph nodes or tissue.

Clinical course of RDD is generally benign, with spontaneous complete resolution in most cases, especially if the disease affects mainly lymph nodes. However, locoregional recurrence of RDD and even dissemination can be possible, particularly in forms with extranodal involvement. There have also been

reports of deaths directly caused by RDD, even in children, especially in severe extranodal involvement. RDD is often self-limiting [6].

However, in case of significant extranodal disease and/or compression of vital organs by massive lymphadenopathy, prompt therapy may be indicated [6].

Medical therapeutic options include corticosteroids, antibiotics, antiviral agents, chemotherapy, and radiotherapy. However, no universally accepted treatment guidelines exist forms, with involvement of CNS, kidneys or respiratory tract [16]. In our case, the presentation was mild, self-limiting, and no any of the therapeutic options needed.

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