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Radio-Diagnosis

A RARE CASE OF ROSAI DORFMAN DISEASE

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ABSTRACT **Introduction:** Rosai-Dorfman disease (RDD) also known as sinus histiocytosis with massive lymphadenopathy is a rare disease of unknown aetiology. It is a non-neoplastic disorder of which the evolution is often self-limiting over a period of few months. It usually presents with painless cervical lymphadenopathy with or without extranodal manifestation. The prognosis of the disease is dependent on the extranodal involvement. **Case report:** A 47 year old male with complaints of pain abdomen, malaise. progressive neck swelling and seven months history of progressive weight loss, cough was not paroxysmal, not barking and no associated difficulty in breathing.There was no history of contact with adult having chronic cough. At the same time the person developed low grade intermittent fever with drenching night sweats but no chills or rigors. **Conclusions:** RDD is rare, requiring knowledge of its clinical manifestations for a rapid and correct diagnosis. In light of the possibility of recurrence, long-term follow-up is needed. Treatment is still controversial. Surgical intervention was done in our patient and follow up visit after 1 month showed no recurrence.

KEYWORDS : Rosai-Dorfman disease, lymphadenopathy, sinus histiocytosis.

INTRODUCTION

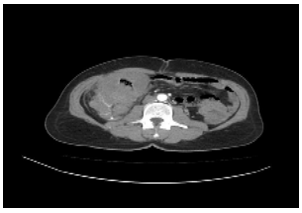
Rosai-Dorfman disease (RDD) is a rare non-Langerhans cell histiocytosis characterized histopathologically by the accumulation of CD68-positive, S100-positive, and CD1a-negative histiocytes with frequent emperipolesis. RDD was first described in 1965 in four African children with lymphadenopathy by Destombes, and was called “adenitis with lipid excess”, owing to the lipid-laden histiocytes in the tissue specimen.1 In 1969, Rosai and Dorfman reported a separate series of four patients with massive cervical lymphadenopathy with specific histopathological features, and called it “sinus histiocytosis with massive lymphadenopathy”.2 Since the original description, further reports, including a summary of 423 cases from an international registry in 1990, described both nodal and extranodal manifestations of the disease.3 In the last decade, the understanding of the biology of related histiocytic disorders such as Erdheim-Chester disease (ECD) and Langerhans cell histiocytosis.

The diagnosis of this condition is challenging due to the wide spectrum of disease manifestations. Multifocal, enlarged lymph nodes in RDD reveal homogeneous enhancement on CT images. The differentiation from lymphoma may not be possible on imaging, as both conditions present with multiple nodes that appear discretely enlarged or with soft-tissue masses. RDD usually follows a benign and self-limiting course with treatment largely targeted at controlling local manifestations (surgical intervention)

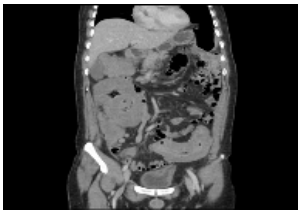
CASE HISTORY

A 47 year old male with complaints of pain abdomen, malaise. progressive neck swelling and seven months history of progressive weight loss, cough was not paroxysmal, not barking and no associated difficulty in breathing.There was no history of contact with adult having chronic cough. At the same time the person developed low grade intermittent fever with drenching night sweats but no chills or rigors. Few days later, he noticed bilateral neck swelling of peanut size which progressively increases to the size at presentation. Neck swelling was painless and there was no associated difficulty in swallowing or noisy breathing. There was significant weight loss, but no history of bleeding diathesis, bone or joint pains. Clinical examination of the patient revealed axillary temperature of 37.2 C, no conjunctival pallor, not jaundiced, acyanosed, not dehydrated, no dysmorphic facie, but with significant and generalized peripheral lymphadenopathy, with the largest measuring 6x4 cm at the right anterior triangle of the neck. The lymph nodes were firm, discrete and mobile, not tender, no differential warmth, no change in colour of overlying skin, no pedal oedema. Abdomen was full, moves with respiration, hernia orifices were intact. There was hepatomegaly 8cm below the right sub-costal margin that is firm in consistency, smooth and not tender. There was no splenomegaly a nd no demonstrable ascites. The other physical examination findings including ear, nose and throat were essentially normal. Haematologic examination revealed microcytic hypochromic anaemia with a haemoglobin concentration of 8.3 g/dl, total leukocyte count of 3 6,600/mm with neutrophil constituting 67%,lymphocytes 3 0 % , and monocytes 3 % , thrombocytosis with a platelet count of 3650,000/mm , red cell morphology showed anisocytosis and spherocytosis, along with few target cells. Blood chemistry, liver and renal function tests were within normal limits, Mantoux test and HIV serology were negative. The Chest X-ray showed multiple lobulated masses of soft tissue density in the mediastinum and both hilar regions compressing both main bronchi. Abdominal ultrasound showed para aortic lymphadenopathy The excisional biopsy of cervical lymph node suggested RDD. The patient was treated with corticosteroid for 6 weeks with a favourable response of weight gain and regression of lymphadenopathy with no evidence of steroid impregnation. The haemoglobin concentration increased from 8.3 to 9.7 g/dl without transfusion or haematenics supplementation. Following clinical and laboratory evidence of improvement, a maintenance dose was continued with gradual dose reduction over six weeks. The patient was eventually lost to follow-up and no phone number was documented in the case file for possible contact, attempt to trace location from the documented address was not successful.

CECT image



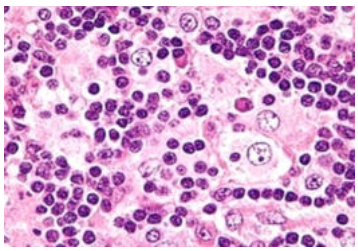
Axial and coronal



Lon
Long segment homogeneously enhancing polypoidal circumferential thickening in ascending, transverse, descending and rectosigmoid junction.

Two possible differential diagnoses:
1. Multifoal colonic and duodenal lymphoma.
2. Colonic xanthomatosis.

Histopathology report
Studies sections showed histiocytic disease involving entire colon, mesentery and regional lymph nodes. Lymph nodes showed effaced architecture by plenty of histiocytes showing emperipolesis. polypoidal circumferential thickening in ascending, transverse, descending and rectosigmoid junction with skip areas in between causing luminal narrowing and loss of mural stratification. Homogeneous enhancement is seen on arterial and venous phases. Multiple enlarged pericolic lymph nodes in RIF and root of mesentery, predominantly along ascending colon, conglomerating with each other.



Diagnosis of extranodal sinus histiocytosis with massive lymphadenopathy (Rosai Dorfman disease) was made.

DISCUSSION
Rosai-Dorfman Disease is a rare chronic disease characterized by benign self-limiting histiocytic proliferation of non-Langerhans histiocytosis3cells. Its aetiology remains obscure, even though its association with infectious nature is often reported, including infections with EBV, CMV, measles, rubella and toxoplasmosis which are still common in our setting, but at the same time disorders of the immune system with abnormal histiocytes reaction 4have also being reported. This rare disorder was first reported by Juan Rosai 2 and Ronald F Dorfman in 1969, in 1990, Rosai 1 reported 423 documented cases. Since then several hundreds of isolated cases of RDD have been reported worldwide, only a few cases were1,5-8 reported from Africa. There was only a case 9 reported from Nigeria. Extranodal involvement is 10,11 found commonly in the head and neck regions. Our patient had hepatomegaly which may suggest hepatic involvement, which is even more rare and in consistent with the case reported by Maheshwari 12 et al. Fever and weight loss may also be presenting symptoms of this disease as was the case for our patient which are in consistent with the two case 1reported by Atoumane Faye et al. Other rare presenting features include tonsillitis, nasal obstruction which were not seen in our patient. The disease may occur at any age,in our case, the patient is aged 47 years. Lymphadenopathy constitute a classical mode of presentation of the disease, and are chronic, non-inflammatory often localized to the cervical region but may rarely involve other nodal regions as was the case with our patient where there were axillary, inguinal and deep mediastenal and hilarlymphadenopathy clinically 1consistent with reported cases in the literature. The masses are typically voluminous cervical lymphadenopathy that can measure up to 7cm in 13,14 their long axis. More often bilateral, asymmetric, firm in consistency, painless without necrosis or tendency to suppuration or fistula formation as the case with our patient. In our observation, the most voluminous lymphadenopathy was 5cm of long axis with the same characteristics described as in the index case. The lymphadenopathy can last for several months or years. Our case had hepatomegaly at variant with many reported cases having both liver and spleen been consistently 2,5,8,9 normal. This long standing hepatomegaly may be from various aetiologies commonly implicated particularly in tropics including hepatic15,16 schistosomiasis and visceral leishmaniasis. Histology was suggestive of RDD using Haematoxylene and Eosin in our case, although other mimics such as tuberculosis was rule out by Mantoux negative tes t and genexpert. For our patients immunohistochemistry was not done because of its unavailability in our centre, however, if done it can assist in ruling out Langerhans histiocytosis and lymphoproliferative disorders especially in atypical RDD. To date, there is no consensus on the initiation of 2,5 systemic treatment of the disease, except in rarecases where the life is threatened such as airway compression. Our patient presented, low grade intermittent fever, along with painless non-tender bilateral cervical, mediastenal and hila lymphadenopathy, and hepatomegaly was observed. However, corticosteroids remain important in the management of RDD, Promising results were obtained with prednisone or a combination with vinblastine or chlorambucil. We used prednisolone at a dose of 1mg/kg/day for 8 weeks with remarkable clinical and laboratory improvement without steroid impregnation in this index case. It was difficult to assess the long-term effect of the corticosteroid treatment, keeping in mind the self-limiting possibility of RDD, and therefore, assessment of corticosteroid therapeutic effect remains difficult.

CONCLUSION
RDD is rare, requiring knowledge of its clinical manifestations for a rapid and correct diagnosis. In light of the possibility of recurrence, long-term follow-up is needed. Treatment is still controversial. Surgical intervention was done in our patient and follow up visit after 1 month showed no recurrence.

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