

# Rosai-Dorfman's Disease Mimicking Nasopharyngeal Carcinoma: A Preliminary Case Report

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**ABSTRACT----** *We report the case of a 28-year-old female Nigerian with Rosai-Dorfman disease mimicking Nasopharyngeal Carcinoma. Patient presented with bilateral cervical lymphadenopathy of 1 year duration, snoring of 3months duration and difficulty in breathing of 3days duration. Developed progressive worsening difficulty in breathing and restlessness. No history of weight loss, fever, or epistaxis. On physical examination, patient was in obvious respiratory distress, with signs of upper airway obstruction. Bilaterally enlarged lymph nodes at level 5 and non-tender. Throat examination showed an obvious postnasal mass on tongue depression with a metallic tongue deppressor. Plain soft tissue neck radiograph revealed narrowed pharyngeal airway caused by a soft tissue mass. Patient was resuscitated and taken to the theatre for emergency tracheostomy, Examination Under Anaesthesia of the postnasal space showed a spherical firm mass which was biopsied. Histology report revealed the mass to be Rosai-Dorfman's disease. Patient was commenced on low dose prednisolone and the mass is regressing.*

**Keywords----** Rosai-Dorfman, disease, nasopharyngeal, carcinoma

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## 1. INTRODUCTION

Rosai-Dorfman's disease is a rare disease characterized by massive lymphadenopathy due to the accumulation of histiocytic cells in the lymph nodes; hence the alternative name of Sinus histiocytosis with massive lymphadenopathy. The disease was reported in 1969 by two Pathologists Rosai J and Dorfman RF as a rare histiocytic disorder [1]. Presentation may be nodal or extra-nodal in nature. In the nodal type, the disease involves primarily the lymph nodes. Most commonly involve are lymph nodes of the cervical region. There may be in addition constitutional symptoms in which the disease may mimic tuberculous lymphadenitis [2].

In the extra-nodal disease, a variety of organs are involved which may include the skin, respiratory system, kidneys and the brain. If the airway is involved, the patient may present with signs and symptoms of upper airway obstruction which may necessitate surgical intervention. Patients presenting with isolated intracranial disease tend to be older and with poorer prognosis [3]. It has been found that bone marrow transplantation as treatment for acute lymphoblastic leukemia or after Hodgkin's and non-Hodgkin's disease may results in the development of the disease [4]. The role of an infective agent such as viral or bacterial infection has not been fully established [5].

## 2. CASE REPORT

We report the case of a 28-year-old female Nigerian with Rosai-Dorfman disease mimicking Nasopharyngeal Carcinoma. Patient presented with bilateral cervical lymphadenopathy of 1 year duration, snoring of 3months duration and difficulty in breathing of 3days duration. Patient was well until a year prior to presentation when she developed bilateral, painless cervical lymphadenopathy, with associated snoring, nasal discharge and progressive nasal obstruction. There was no history of epistaxis, weight loss or fever. Developed difficulty in breathing which was slowly progressive in nature.

Past medical history showed that patient had lymph node excision 2 years prior to presentation and was then treated for tuberculosis. There was no other medical problem and patient was not on any medication prior to presentation.

On physical examination, patient was in obvious respiratory distress, with signs of upper airway obstruction. Plain soft tissue neck radiograph revealed narrowed nasopharyngeal airway by a soft tissue mass in the nasopharynx. Patient was resuscitated and taken to the theatre for emergency tracheostomy and nasopharyngeal mass biopsy (Figure1: At presentation, mass exposed with a metallic tongue depressor and figure 2: same patient 3<sup>rd</sup> day post tracheostomy).



Figure 1: At presentation, mass exposed with a metallic tongue depressor.

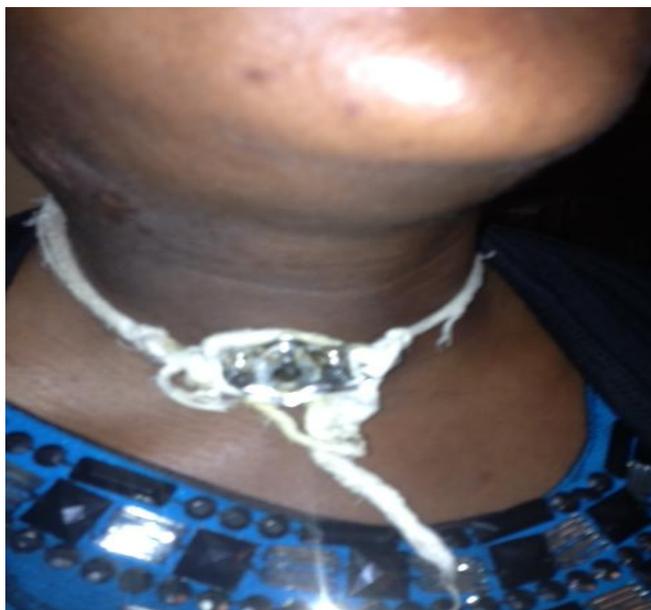


Figure 2: Patient 3rd day post tracheostomy

Histology report showed the mass to be Rosai-Dorfman's disease as shown in figures 3 – 6 (micrographs). Patient was commenced on low dose prednisolone and we are still following up this patient.

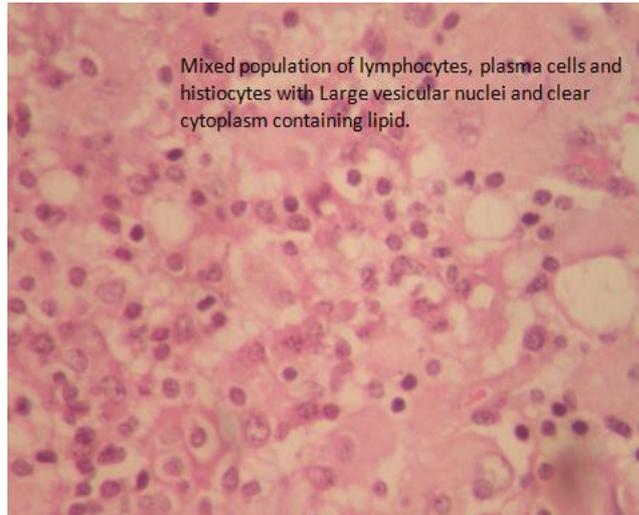


Figure 3: Mixed population of lymphocytes, plasma cells and histiocytes with large vesicular nuclei and clear cytoplasm containing lipid.

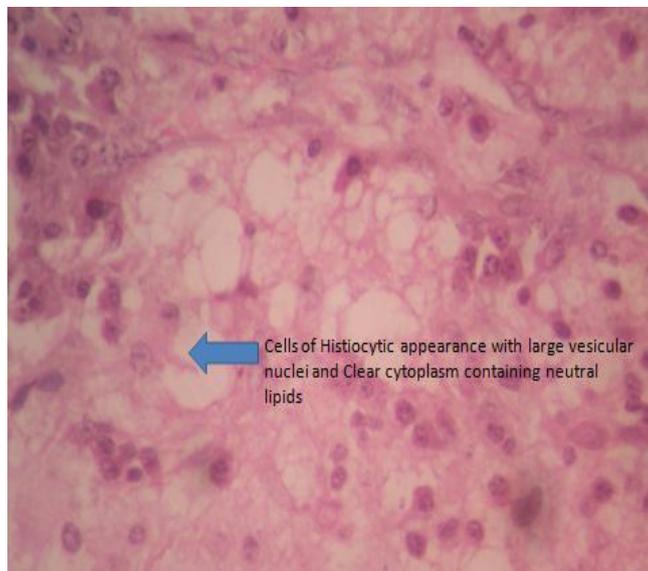


Figure 4: Cells of histiocytic appearance with large vesicular nuclei and clear cytoplasm containing neutral lipids.

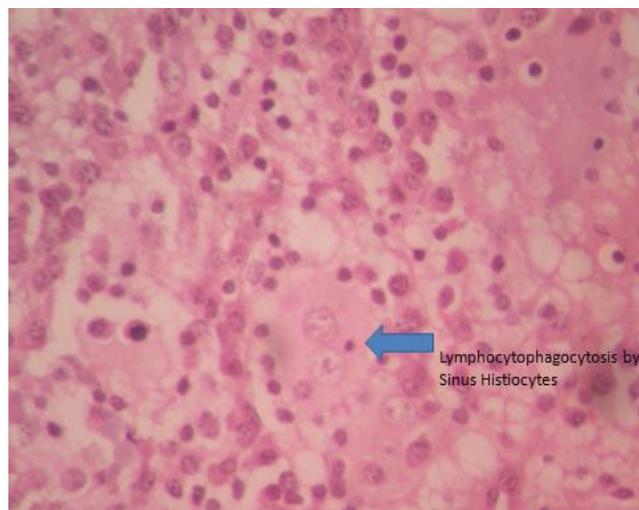


Figure 5: Lymphocytaphagocytosis by sinus histiocytes.

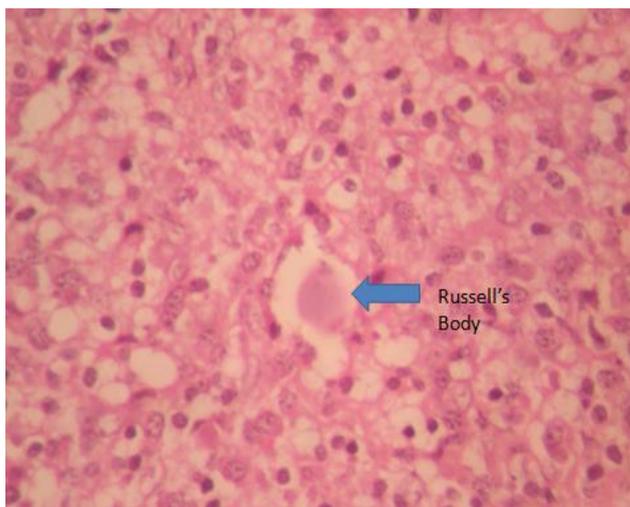


Figure 6: Russell's body

The diagnosis is based on the histology of the biopsied tissue showing lymphocytes and other inflammatory cells residing within the cytoplasm of the histiocytes and a positive staining for CD68 and S-100 as confirmatory.

### 3. TREATMENT

Treatment depends on the types, presentation, severity and disease complications, if any. Most times the disease runs a benign course and so treatment may therefore be unnecessary.

However, treatment is warranted in patients with severe extra-nodal disease involving vital organs with life-threatening complications as in the index case[6].

Surgery may be needed for lymph node biopsy to make a histological diagnosis and to relieve upper airway obstruction as in this case[6]. In cases where complete resection of the mass may not be feasible, then partial resection to relieve immediate symptoms may be aimed at. In some cases adjuvant chemotherapy has been found to be successful in achieving remission[7]. Radiotherapy has been shown to have limited efficacy while systemic corticosteroids have been shown to be beneficial in decreasing nodal size and symptoms despite their side effects[8].

### 4. CONCLUSION

This report is to draw the attention of Head and Neck Surgeons to how closely this rare disease can mimic a nasopharyngeal carcinoma and the need to have an open mind when treating patients with cervical lymph node enlargement associated with postnasal mass.

### 5. REFERENCES

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