


**CASE REPORT**

# Rosai-Dorfman disease in Malawi

Edwards Kasonkanji<sup>1</sup>  | Ryan Seguin<sup>1</sup> | Bongani Kaimila<sup>1</sup> | Bal M. Dhungel<sup>1</sup> | Matt Painschab<sup>2</sup> | Tamiwe Tomoka<sup>1</sup> | Satish Gopal<sup>1,2,3</sup>

<sup>1</sup>UNC Project-Malawi, Lilongwe, Malawi

<sup>2</sup>University of North Carolina, Chapel Hill, North Carolina

<sup>3</sup>University of Malawi College of Medicine, Blantyre, Malawi

**Correspondence**

Edwards Kasonkanji, UNC Project-Malawi, Lilongwe, Malawi.

Email: ekasonkanji@unclilongwe.org

**Key Clinical Message**

Rosai-Dorfman Disease (RDD) is a rare lymphoproliferative disease with limited cases reported in sub-Saharan Africa, potentially due to a lack of pathological services throughout the region. RDD diagnosis can be difficult, especially in resource-limited setting, as symptoms can be nearly identical to more common causes of lymphadenopathy.

**KEYWORDS**

ear, nose and throat, general surgery, oncology, pediatrics and adolescent medicine

## 1 | INTRODUCTION

Rosai-Dorfman Disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy, is a rare, nonmalignant disorder characterized by high proliferation of the non-Langerhans sinus histiocyte cells within the lymphatic system.<sup>1,2</sup> RDD frequently presents with solitary, painless, and often massive lymphadenopathy, especially cervical.<sup>2</sup> RDD involving extranodal sites accounts for 40% of cases and shows similar pathologic features to its nodal counterpart, although more fibrosis and fewer histiocytes are present and, emperipolesis is less prominent.<sup>3</sup> Laboratory investigations typically demonstrate increased neutrophil count and erythrocyte sedimentation rate. Most cases occur in the first or second decade of life and are usually indolent and non-life-threatening. Spontaneous regression occurs in approximately 20% of cases with a watch-and-wait approach to treatment.<sup>4</sup> The etiology and pathogenesis of RDD are unknown.

## 2 | CASE REPORT

An HIV-negative female adolescent, presented to the clinic with a 7 cm left swollen cervical lymph node, complaining of headache, general malaise, and fatigue. The mass was solitary and embedded within the left sternocleidomastoid

muscle. The patient believed the growth to be part of normal, puberty-related physiological changes, and it was present for at least 6 months prior to presentation. The patient was previously healthy and had no significant social or family history.

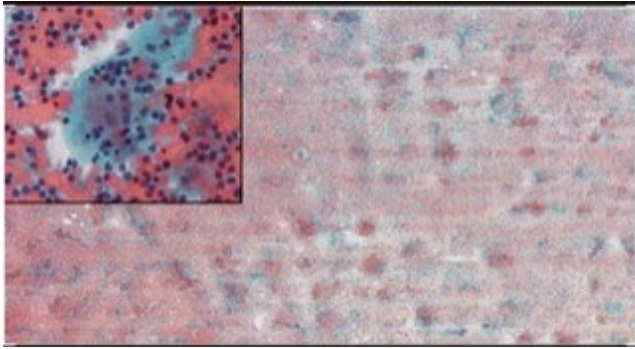
All laboratory findings were within normal limits, including full blood count, chemistries, and lactate dehydrogenase, and Erythrocyte Sedimentation Rate (ESR). Abdominal ultrasound and chest X-ray were also normal.

The patient initially underwent a fine needle aspiration procedure and cytological examination detected numerous histiocytic cells, some showing emperipolesis suggestive of RDD (Figure 1). Immunohistochemistry was not done as the emperipolesis was thought to be pathognomonic for RDD.

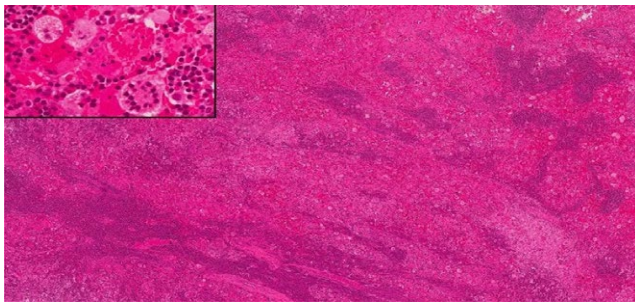
Partial surgical excision was done, due to the tumor's anatomical position. Following incomplete resection of the tumor and continued symptoms of malaise post-operatively, the patient was treated with prednisolone at 0.6 mg/kg body weight for 1 week then tapered off over 1 month.<sup>5</sup>

After 48 months of follow-up, the patient remains asymptomatic without signs of recurrence. The tumor has decreased substantially to a size less than a centimeter, and her full blood picture, chemistries, lactate dehydrogenase, and ESR all remain within normal limits.

The left swollen 7 cm cervical tumor was subtotally, surgically excised. Pathological examination demonstrated numerous histiocytes with abundant clear to vacuolated cytoplasm



**FIGURE 1** Papanicolaou stain of fine needle aspiration smear from patient demonstrating numerous scattered Histiocytic cells, some showing emperipolesis



**FIGURE 2** Hematoxylin and Eosin stain from excised lymph node demonstrating numerous histiocytes with abundant clear to vacuolated cytoplasm displaying emperipolesis. The sinuses are dilated and packed with these histiocytes

displaying emperipolesis (Figure 2). The sinuses were dilated and packed with histiocytes.

### 3 | DISCUSSION

Rosai-Dorfman Disease is rarely progressive and typically follows a disease course of alternating periods of exacerbations and resolutions. However, patients may require long-term follow-up due to the variable clinical course.<sup>6,7</sup>

Our case currently indicates features of complete remission following surgical excision and tapering steroid doses, in line with most reported cases of RDD.<sup>5,6,8</sup>

Although our case presented with singular lymph node disease, common extranodal sites are skin, central nervous system, orbit and eyelid, upper respiratory tract, and gastrointestinal tract.<sup>3,9,10</sup> Other, less common extranodal sites include the thyroid gland, breast, meninges, and spinal cord, with most spinal cord involvement presenting as focal extradural masses although intramedullary and intradural masses have been reported.<sup>11</sup>

There is insufficient evidence to support immunodeficiency, autoimmune disease, or a neoplastic process as the etiology. Molecular studies of RDD have found no evidence of

clonal rearrangement, implying a reactive or non-neoplastic condition. Associations with Epstein-Barr virus, cytomegalovirus, brucella, klebsiella, and human herpes virus 6 have been suggested but not proven.<sup>3</sup>

Diagnosing RDD in limited resource settings can be difficult, as its symptoms can be nearly identical to more common causes of lymphadenopathy, such as lymphoma and other histiocytic conditions, including Erdheim-Chester disease, Langerhans cell histiocytosis, reactive histiocytic proliferation, and juvenile xanthogranuloma can also mimic RDD symptoms.<sup>9</sup>

A familial version of RDD exists with mutations in the *SLC29A3* gene, which encodes an intracellular human equilibrative nucleoside transporter, though the pathogenesis of how this mutation specifically leads to RDD is yet to be elucidated.<sup>12</sup> Given the relative absence of proven risk factors and clinical symptoms that are often indistinguishable from other, more common diseases, RDD may be underrecognized in sub-Saharan Africa.

To our knowledge, there are no other published reports from sub-Saharan Africa describing this disease, potentially due to a lack of pathological services throughout the region.

Increasing clinician and pathologist familiarity with RDD and improving pathology services throughout sub-Saharan Africa are important steps toward accurate differential diagnosis and recognition of the disease. Additional published reports from the region are needed to further guide evidence-based treatment.

### CONFLICT OF INTEREST

The authors have no conflict of interests to declare.

### AUTHOR CONTRIBUTIONS

EK: Conception, designing, and case report writing. RS: peer reviewed and helped in case report drafting and design. TT: pathological diagnosis and interpretation the tissue biopsy slides. SG: Provided the administrative support, reviewed, and provided mentorship throughout the writing process. BG, TZ, MC, BT, AM, SC: data collection and management.

### ORCID

Edwards Kasonkanji  <http://orcid.org/0000-0002-1543-814X>

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**How to cite this article:** Kasonkanji E, Seguin R, Kaimila B, et al. Rosai-Dorfman disease in Malawi. *Clin Case Rep.* 2018;6:2313–2315. <https://doi.org/10.1002/ccr3.1820>