An Unusual Cause of Lymphadenopathy: Rosai Dorfman Disease in a 7-Year-Old Female Zambian Child: Case Report and Literature Review

Clinical Medicine Insights: Case Reports
Volume 17: 1–7

DOI: 10.1177/11795476241265279

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ABSTRACT: Rosai Dorfman disease (RDD) is a rare non-Langerhans histiocytic disorder, which belongs to the R group of the 2016 revised histiocytic classification. It's characterized by the accumulation of activated histiocytes in the sinusoids of lymph nodes and/or extranodal tissues. Herein, we report a 7-year-old female who was initially suspected to have a lymphoma but was later identified as having RDD. She presented with a history of fever, night sweats, and weight loss, and on physical examination had bilateral cervical lymphadenopathy. Histologic examination of the biopsied cervical lymph nodes showed distended sinuses with S100 and CD68 immunoreactive histiocytes demonstrating emperipolesis, confirming a diagnosis of RDD. The condition is known to be self-limiting. However, evidence from literature and our case management shows that medical therapy can hasten remission in pediatric cases.

KEYWORDS: Rosai-Dorfman disease, histiocytosis, massive lymphadenopathy, histopathology, Zambia

RECEIVED: April 5, 2024. **ACCEPTED:** June 11, 2024.

TYPE: Case Report

FUNDING: The author(s) received no financial support for the research, authorship, and/or publication of this article.

DECLARATION OF CONFLICTING INTERESTS: The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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Introduction

Rosai-Dorfman disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy, is a rare non-Langerhans cell histiocytosis characterized by overproduction and accumulation of activated histiocytes within affected tissues, primarily the lymph nodes.[1] Extranodal disease though quite rare can occur and usually affects the skin, orbit, central nervous system, and digestive system.[2,3] The disease was first described in 1965 by Pierre Paul Louis Lucien Destombes in 4 French patients who presented with "adenitis with lipid excess" and was then referred to as Sinus histiocytosis with massive lymphadenopathy. In 1969, Juan Rosai and Ronald Dorfman characterized this condition as a specific clinicopathological entity, and has since been termed the Rosai-Dorfman-Destombes disease.[4,5]

RDD belongs to the R group of the 2016 revised classification of histiocytosis. It is a heterogeneous entity with a wide range of clinical phenotypes occurring in isolation or with autoimmune or malignant diseases.[6] Immunohistochemical studies show that the proliferating cells express histiocytic markers CD163 and CD68 as well as S-100 but are negative for the Langerhans cell markers CD1a, dendritic cell markers CD21 and CD35.[1,2] Some recent studies have found *NRAS, KRAS, MAP2K1*, and *ARAF* mutations in lesional tissues, raising the possibility of a clonal origin in some forms of RDD.[1]

RDD has a worldwide prevalence of 1:200 000, with a slight male preponderance (1.4:1), and is more frequent in individuals of African descent and Caucasians. The disease is more commonly seen in children and young adults.[4] Clinically patients with RDD present with bilateral painless cervical lymphadenopathy, with or without intermittent fevers, night sweats, and weight loss. Extranodal involvement is seen in 25% to 40% of the cases.[7] Laboratory examination reveals an elevated erythrocyte sedimentation rate (ESR), leukocytosis with neutrophilia, hypochromic anemia, elevated ferritin levels, and often hypergammaglobulinemia.[2,3]

The etiology of RDD is not exactly known. Some theories suggest that it occurs due to immune dysregulation and has been associated with autoimmune disorders such as systemic lupus erythematosus (SLE). Infectious agents such as Epstein-Barr Virus (EBV), human herpes virus 6 (HHV-6), and Klebsiella have been implicated. However, this evidence is inconclusive.[3,8] RDD is usually self-limiting and commonly does not require therapy. Therefore, there are no defined therapeutic algorithms for its treatment. Surgery and systemic treatments such as steroids and cytotoxic agents can be used, but since many cases spontaneously regress a "watch and wait" approach is advocated.[8]

We report a case of Rosai-Dorfman disease in a 7-year-old Zambian female diagnosed by lymph node biopsy and

Table 1. Laboratory investigations.

PARAMETERS	FIRST ADMISSION	SECOND ADMISSION	NORMAL VALUES
Hematology			
White blood cell count	**14.57 × 10⁹/l**	6.04 × 10⁹/l	5.00-13.00 × 10⁹/l
Red blood cell count	3.88 × 10⁹/l	5.45 × 10⁹/l	4.00-5.20 × 10⁹/l
Hemoglobin	**7.7 g/dl**	10.6 g/dl	12.1-16.3 g/dl
Mean corpuscular volume	**64.7 fl**	**60.8 fl**	79.1-98.9 fl
Mean corpuscular hemoglobin	**19.8 pg**	**19.4 pg**	27.0-32.0 pg
Platelets	**974 × 10⁹/l**	**746 × 10⁹/l**	150-400
Neutrophils	**77%**	55.4%	2.0-8.00 × 10⁹/l
	11.22× 10⁹/l	3.35 × 10⁹/l	
ESR	**36 mm/h**	**85 mm/h**	0-15 mm/h
CRP	**641.83 mg/l**	**-**	0-6 mg/l
Biochemistry			
Lactate dehydrogenase (LDH)	**322 u/l**		0-250 u/l
Serum iron	86.15 ug/dl		35-145 ug/dl
Total iron binding capacity (TIBC)	312 ug/dl		250-400 ug/dl
Transferrin saturation	28%		20%-50%
Ferritin	**256.0 ng/ml**		10-160
Autoimmune screen			
Parameter	Value	Normal value	Interpretation
Anti-nuclear antibody (ANA)	36.5 AU/ml	0.0-40	Negative
Rheumatoid arthritis-IgM (RA)	**40 IU/l**	<15	**Positive**

The bold figures in Table 1, denote abnormal laboratory values found in our case.

managed medically at the University Teaching Hospitals (UTHs)-Children's Hospital in Lusaka, Zambia. Despite being rare, RDD is a differential diagnosis for cervical lymphadenopathy, that clinicians must be aware of.

Case Presentation

A 7-year-old female presented to the University Teaching Hospital- Children's Hospital on September 19, 2022, with a 12-month history of progressive bilateral neck swelling, fever, night sweats, and weight loss. She was referred from Macha Mission Hospital, a district hospital in the Southern province of Zambia. Her past medical history was unremarkable, with no positive TB contact or family history of similar swelling.

On physical examination, she was alert, moderately pale, and undernourished with a body mass index (BMI) of 10.24 kg/m^2. She had bilateral cervical lymphadenopathy, and her vitals were normal. Local examination revealed multiple bilateral nodes in the submandibular, submental, and posterior regions of the neck. The nodes were rubbery, mobile, and non-tender with normal overlying skin, the largest node was in the right upper posterior triangle measuring 4 × 3 cm. She was noted to have some inguinal lymph nodes as well. There was no hepatosplenomegaly and examination of other systems was unremarkable. The differential diagnoses at this initial admission were lymphoma and tuberculous lymphadenitis.

Routine laboratory tests were performed as shown in Table 1. A peripheral blood smear was done and it revealed hypochromia, anisocytosis, and microcytosis of the red blood cells with rouleaux formation, no blasts were seen, and platelets were increased on the film. Lymph node aspirate and sputum were subjected to gene-Xpert for mycobacterium tuberculosis (MTB) and were negative, lateral flow urine lipoarabinomannan (LF-LAM) for MTB was also negative.

Radiological investigations included a Chest X-ray showing bilateral hilar lymphadenopathy (Figure 1), and CT scans of the neck, chest, and abdomen. CT scan of the neck showed

multiple enlarged lymph nodes in the bilateral submandibular, jugular, and posterior triangle, the lymph nodes showed homogenous contrast enhancement, the largest measuring 2 cm × 2.6 cm, CT scan of the chest showed multiple enlarged nodes in the paratracheal, subcarinal, perivascular and hilar regions and CT scan of the abdomen showed multiple mildly enlarged lymph nodes in the peri gastric, periportal and gastrosplenic region, there was no radiological Extra-nodal manifestations of disease(Figure 2). The patient was initially commenced on anti-tuberculous treatment based on the clinical picture (unconfirmed tuberculosis).

To rule out a possibility of leukemia, a bone marrow aspirate was done and it showed a hypercellular bone marrow, activated monocytes, presence of a few phagocytes with vacuolated cytoplasm, activated lymphocytes where noted, and increased plasma cells. To make a definitive diagnosis, a cervical lymph node biopsy was done, which comprised one encapsulated tan mass of tissue measuring 18 mm × 16 mm × 13 mm.

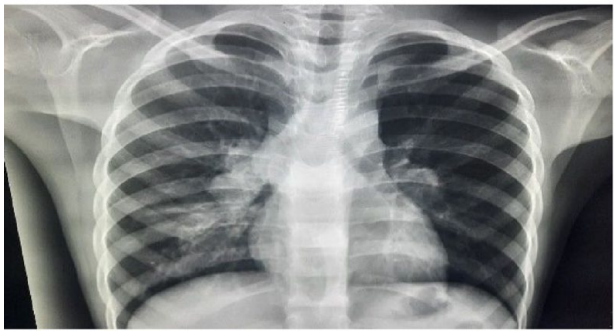

Figure 1. Chest X-ray showing perihilar lymphadenopathy.

Transection of the mass showed an encapsulated firm tissue mass with a nodular architecture. Microscopic examination of hematoxylin and eosin (H&E) stained slides revealed an intact lymph node with a nodular architecture composed of expanded pale areas (sinuses) containing numerous histiocytes (Figure 3A and B). The sinus histiocytes had large vesicular nuclei with abundant clear to eosinophilic cytoplasm containing numerous intact lymphocytes and occasional neutrophils, a feature designated as emperipolesis or lymphocytophagocytosis (Figure 3C and D). There were focal areas of dense intra- nodal fibrosis. This prompted the use of immunohistochemistry with antibodies against S100 and CD68, which demonstrated positive cytoplasmic immunoreactivity of the sinus histiocytes with both markers (Figure 4) thereby confirming the diagnosis of Rosai-Dorfman disease.

ATT was stopped and the patient was commenced on steroids (prednisolone 20mg once daily) for 6 weeks, with a resolution of fever but no reduction of the lymph node size. Nevertheless, the patient's father requested to go home due to social and financial challenges. She was discharged on a slow taper-off dose of prednisolone. She was reviewed 6 months later and was restarted on prednisolone at a higher dose of 40 mg per day(2 mg/kg/day) this was because of the increase in the nodal size compared to the first admission. There was a noted reduction in the size of the nodes at this higher dose as shown by (Figure 5).

Discussion

Rosai-Dorfman-Destombes disease is a rare disorder characterized by the overproduction and accumulation of histiocytes in lymph nodes sinuses and extranodal sites. It can affect

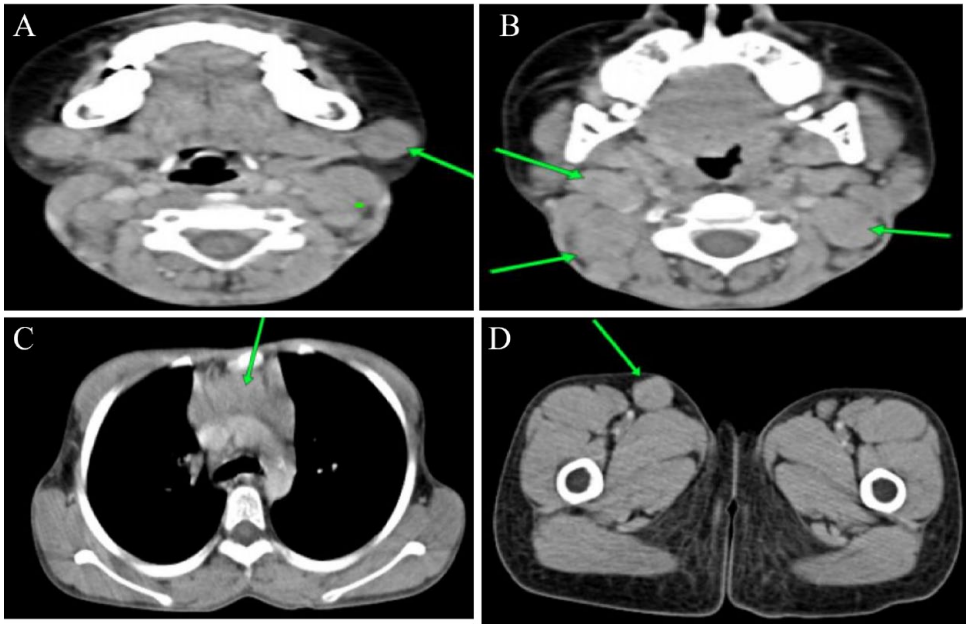

Figure 2. CT images showing generalized lymphadenopathy in the mediastinal, bilateral cervical, and right inguinal regions, (A) Submandibular lymph nodes largest is 1.4 cm, (B) Right anterior and bilateral upper posterior jugular lymph nodes largest is 1.6 cm. (C)Pre-vascular lymphadenopathy, matted nodes measuring $d = 2.7$ cm × 2.2 cm, and (D) Right inguinal nodes, measures $d = 1.7$ cm.

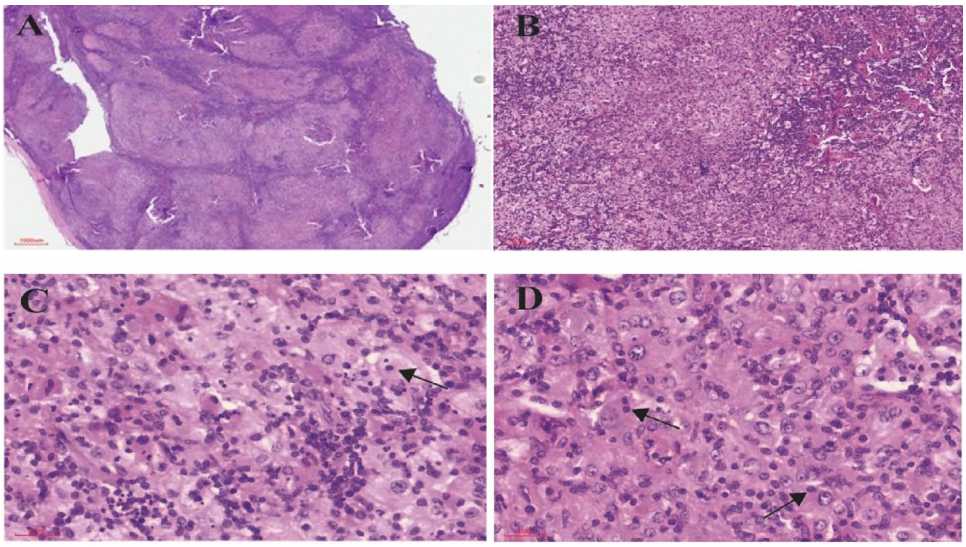

Figure 3. H&E histomorphologic findings of the cervical lymph node. (A) Low power view showing a lymph node with expansion of the sinuses (pale areas) and compression of other compartments (×40 magnification). (B) Pale areas demonstrate numerous sinus histiocytes with areas of lymphoid aggregates (×100 magnification). (C and D) High power view showing numerous sinus histiocytes with abundant eosinophilic cytoplasm and emperipolesis—arrows. Engulfed lymphocytes are evident in this image (×400 magnification).

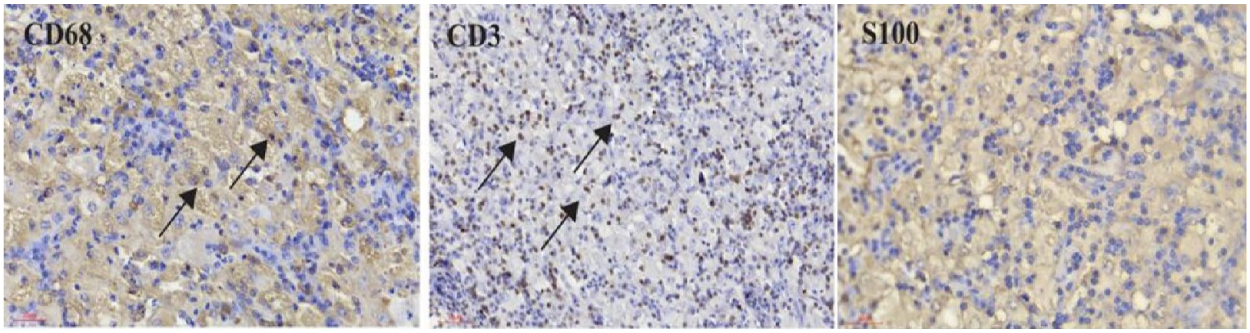

Figure 4. Immunohistochemistry staining patterns using antibodies against CD68, CD3, and S100. The sinus histiocytes demonstrate cytoplasmic immunoreactivity for CD68 and S100 while some lymphocytes, immunoreactive for CD3, are seen within the cytoplasm of the histiocytes -arrows (×400 magnification).

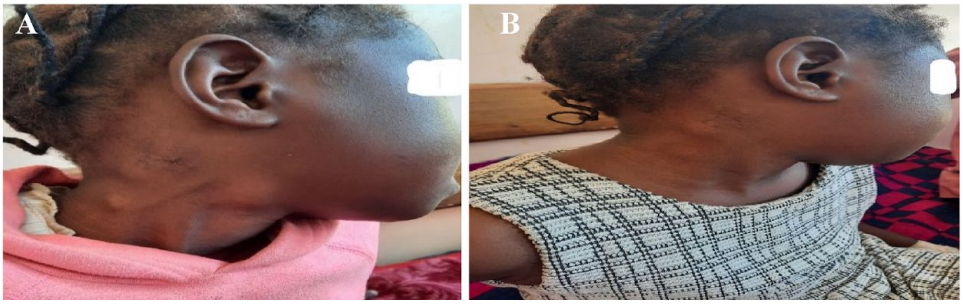

Figure 5. (A) The patient with cervical lymphadenopathy before steroid treatment, (B) a reduction in cervical nodal size after 3 weeks of steroid therapy (prednisolone at 2 mg/kg/day).

people of all ages, but it is commonly seen in children and young adults, it terms of gender, males are more commonly affected than females.[9,10] In this case, we presented a 7-year-old Zambian female with classic features of this rare disease. The etiology of RDD is unknown, Some studies have indicated that the disease might be related to viral infections such as CMV, HIV, EBV, human herpes virus 6 (HHV- 6), HHV-8, parvo B19, and some bacterial infections have also been implicated which include Klebsiella, Brucella., however, they has been no conclusive evidence to confirm this theory.[2,8,11,12]HIV infection is a possible contributor to the pathogenesis of RDD, therefore HIV testing in patients from areas of high HIV endemicity should be undertaken.[13]RDD associated with HIV infection was described as coincidental in the past, but some

evidence supports the hypothesis of a relation between RDD and HIV infection, viral infections such as HIV can create an immunologic environment that will result in activation of histiocyte-microphage system.[14] In our patient, we tested for HIV, hepatitis B, and hepatitis C which were negative. We were unable to test for the other viral etiologies due to limited resources.

RDD is associated with multiple diseases, it can be familial, neoplasia-associated, immune-related, and Ig-4-related.[1] Familial RDD is caused by germline mutations in SLC29A3, which involves faisalable histiocytosis, H syndrome, and pigmented hypertrichotic dermatosis with insulin-dependent diabetes. Neoplasia-associated RDD either proceeds or appears after lymphoma or myelodysplastic syndrome. Immune-related RDD, which constitutes 10% of RDD cases is associated with systemic lupus erythematosus, autoimmune hemolytic anemia, and idiopathic juvenile arthritis. Ig-4-related RDD is common in extranodal RDD such as those involving the liver, lungs, or colon.[1,11] The Rheumatoid Arthritis-IgM (RA) was positive in our patient but the Anti-Nuclear Antibody (ANA) was negative, therefore we are inclined to think that our patient has immune-related RDD, however apart from the generalized lymphadenopathy and hematological abnormalities, she had no other features of juvenile idiopathic arthritis (JIA).

Clinically RDD is a heterogeneous entity with a wide range of phenotypes, from limited and self-resolving to life-threatening forms. it has 2 clinical forms, nodal and extranodal disease. Nodal disease classically presents with bilateral massive and painless cervical lymphadenopathy with or without intermittent fever, weight loss, and night sweats. Cervical lymphadenopathy is present in 90% of patients affected with RDD, however, other lymph node groups can be affected which include axillary, inguinal, mediastinal, and hilar lymphadenopathy.[5,12,15,16] Extra nodal RDD accounts for approximately 25% to 40% of all cases, the most commonly affected sites are the upper respiratory tract (nasal cavities and paranasal sinuses) skin, eye and retro-orbital tissues and bone tissue. Salivary glands and the central nervous system are less frequently involved. Localization of RDD in lungs, urogenital and gastrointestinal tract, breast, thyroid, and even heart have also been reported.[4,5,17] Our case presented with the classic clinical picture of cervical lymphadenopathy, fevers, weight loss, and night sweats for 1 year. She had no features of extranodal disease. In our setting, the commonest differential diagnoses for the above clinical presentation are tuberculous lymphadenitis and malignancies such as lymphomas. Zambia has been ranked amongst the 30 high TB burden countries according to the Global Tuberculosis Report of 2022,[18] which is why the attending pediatrician found it prudent to start anti-tuberculous treatment (ATT) while waiting for the results of the lymph node biopsy. Despite being on the ATT for 3 weeks, the patient's symptoms did not improve, and ATT was eventually stopped after histopathology and immunohistochemistry confirmed the diagnosis of RDD.

Laboratory findings are distinct in patients with RDD, on a full blood cell count, the most common anomaly is leukocytosis with neutrophilia, a mild normochromic or microcytic anemia, and rarely hemolytic anemia. ESR is elevated, and the ferritin levels are usually normal or elevated. Increased IgG and Rheumatoid Factor(RF) levels and positive antinuclear antibody (ANA) tests have also been reported.[2,3,16] Our patient had a mild leukocytosis with neutrophilia and the hemoglobin (HB 7.7 g/dl) was low at initial presentation, the mean corpuscular volume and mean hemoglobin concentration were low at all the hospital visits. Our patient had microcytic hypochromic anemia, which is not very common in patients with RDD, with most cases presenting with normochromic anemia. ESR, CRP, and ferritin were elevated as expected. The clotting profile and lipid profile were within normal limits in our patient. In terms of the autoimmune screen, ANA was negative but the Rheumatoid Arthritis-IgM was positive, showing an element of immune dysregulation in our patient.

Definitive diagnosis of RDD is confirmed by histopathological examination of the biopsied tissue, it is characterized by the presence of histiocytes with emperipolesis, a feature in which intact lymphocytes are engulfed within the cytoplasm of the histiocytes, this feature differentiates it from other histiocytic disorders. The diagnosis of RDD can further be confirmed by immunohistochemistry, with positive staining for S100, CD 68, and CD163, but are negative for the Langerhans cell markers CD1a, dendritic cell markers CD21 and CD35.[10] Histologic examination of the biopsied cervical lymph node in our patient, showed distended sinuses with S100 and CD68 immunoreactive histiocytes demonstrating emperipolesis, confirming the diagnosis of RDD

There is no unified therapeutic protocol for the treatment of this disease due to its rarity and self-limiting nature. As a general principle, treatment is best tailored to the individual clinical circumstances. Treatment options include expectant monitoring, steroids, chemotherapy, and surgical debulking.[4,5,8] After the diagnosis of RDD is made, expectant management is reasonable in many cases, this approach is suitable for patients with uncomplicated lymphadenopathy or asymptomatic cutaneous RDD because 20% to 50% of patients with nodal/cutaneous disease will have spontaneous remission. Surgery is usually limited to biopsy, but resection can be curative for unifocal disease, surgical debulking may be warranted for upper airway obstruction, spinal cord compression, or large lesions causing end-organ compromise and it's the most effective treatment for cutaneous RDD. Corticosteroids are usually helpful in reducing nodal size and symptoms, although responses have been variable. The optimal corticosteroid (prednisolone and dexamethasone) dose and duration are not clearly defined. Prednisolone (40-70 mg per day) has produced complete or partial response in some cases.[1,8]

Sirolimus (Mammalian target of rapamycin inhibitor) and prednisolone induced objective responses and disease stabilization in some patients with RDD and Erdheim-Chester disease. Lastly, chemotherapeutic agents have shown mixed results. Although chemotherapy is generally reserved for refractory or relapse cases, sometimes it is used as initial therapy in disseminated or life-threatening disease.[1,8] In our case, initially we decided to give prednisolone at a low dose of 20 mg per day(1 mg/kg/day) for 4 weeks and tapered down in the next 2 weeks, but there was no response at this dose. On the second hospital admission, the patient was restarted on prednisolone at a higher dose of 40 mg per day(2 mg/kg/day) with a noticeable reduction in the size of the nodes. We opted to use steroids in our case instead of expectant management because literature has shown that steroids hasten remission and are good for nodal reduction.[4] It is recommended that a clinical examination and laboratory test be performed every 3 to 6 months during the first 2 years following diagnosis, then every year. This is important because in some cases RDD has been found to precede a lymphoma.[2,11]

Recently, somatic mutations in the mitogen-activated protein kinase (MAPK) pathway have been found in lesional tissue in a subset of RDD cases[15] some literature has reported genetic mutations of BRAF-V600E in some cases of RDD which is a mutation associated with another non-Langerhans cell histiocytosis named Erdheim-Chester disease. Other mutations such as SLC29A3, ENT3, KRAS, NRAS, ARAF, and MAP2K1 were also reported, which suggest a clonal nature in some RDD patients, this may open new channels for targeted therapy, particularly in refractory cases.[11,15]

In terms of prognosis, in 2011 the histiocyte society divided RDD patients into 3 major groups, the first group is comprised of patients with sudden enlargement of lymph nodes, this group has the best prognosis because spontaneous regression without further recurrences is expected. The Second group consists of patients with immunological abnormalities in which the lymphadenopathy is generalized, the prognosis is worse. The Third group, these are patients with extranodal site involvement and/or multimodal disease with relapses and remission for years, the prognosis depends on the type and number of extranodal sites.[2] Our case falls in the second category and will therefore need frequent hospital reviews to monitor the progression of the disease.

Raising awareness about RDD is important, because of its variable clinical presentation, it can be easily misdiagnosed as a malignancy or an infectious disease like Tuberculosis in our setting. A thorough clinical evaluation coupled with the right diagnostic tests is important to exclude other diagnoses. Improved awareness of this condition will lead to timely intervention and good patient outcomes.[10]

Conclusion

Rosai Dorfman disease, although rare should be considered as a differential diagnosis in patients with bilateral cervical lymphadenopathy especially in our setting where Tuberculous lymphadenitis and lymphoma are the commonest causes of cervical lymphadenopathy. Diagnosis is confirmed via histological examination of the biopsied tissue revealing characteristic histiocytic infiltrate with emperipolesis and immunohistochemical staining positive for CD163, CD68, and S-100, while negative for CD1a. In terms of management, a watch and wait approach is advocated, however Surgical excision, corticosteroids, and chemotherapy may be employed in cases requiring intervention. In our case, we used steroids which have been shown to hasten remission and reduce nodal size. Regular monitoring is essential to assess disease progression and treatment response.

Acknowledgements

The authors would like to acknowledge The University Teaching Hospital-Children's Hospital management, the medical and nursing team that provided medical care to the patient, and the guardians for being cooperative.

Author Contributions

All authors made a significant contribution to this manuscript whether that is in patient care, processing of the biopsy specimens, and follow-up. All authors contributed to drafting, revising, and approving the manuscript's publication.

Consent

The father of the patient provided written informed consent for publication of this case.

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REFERENCES

1. Abla O, Jacobsen E, Picarsic J, et al. Consensus recommendations for the diagnosis and clinical management of Rosai-Dorfman-Destombes disease. *Blood*. 2018;131:2877-2890.
2. Miękus A, Stefanowicz J, Kobierska-Gulida G, Adamkiewicz-Drożyńska E. Rosai-Dorfman disease as a rare cause of cervical lymphadenopathy – case report and literature review. *Cent J Immunol*. 2018;43:341-345.
3. Giri K, Baral A, Tiwari N, Sharma KS. Rosai-Dorfman disease in 6-year-old child: presentation, diagnosis, and treatment. *Clin Case Rep*. 2021;9:e04795-e04796.
4. Amoako E, Danso KA, Akuaku RS, Ulzen-Appiah K. A report of Rosai–Dorfman disease in an adolescent. *Case Rep Pediatr*. 2022;2022:1-5.
5. di Dio F, Mariotti I, Coccolini E, et al. Unusual presentation of Rosai-Dorfman disease in a 14-month-old Italian child: a case report and review of the literature. *BMC Pediatr*. 2016;16:62-67.
6. Emile JF, Abla O, Fraitag S, et al. Revised classification of histiocytoses and neoplasms of the macrophage-dendritic cell lineages. *Blood*. 2016;127:2672-2681.
7. Anand A, Lanjewar K, Ekhar VR, et al. Rosai-Dorfman disease: a rare cause of cervical lymphadenopathy. *Int J Head Neck Surg*. 2014;5:152-154.

8. Amiji I, Mohammed U, Salum H, et al. Rosai Dorfman disease -a rare case of cervical lymphadenopathy. *Clin Case Rep*. 2020;8:2712-2715.
9. miceli A, Cleaver NS. Roai Dorfman disease cutis. *Dermatopathol Diagn Discuss*. 2015;95:3-6.
10. Dhakal S, Katwal S, Ghimire A, Bhusal A, Yogi TN. Uncommon presentation of Rosai-Dorfman disease: nasal and nasopharyngeal involvement: a case report and discussion. *Radiol Case Rep*. 2024;19:956-960.
11. Magableh HM, Jaber HD, Magableh AM, et al. Rosai-Dorfman disease: case series and literature review. *Cureus*. 2023;15:e35193-e35199.
12. Al-Maghrabi JA. Rosai-Dorfman disease: a pathological experience from 2 tertiary care hospitals in western Saudi Arabia. *Saudi Med J*. 2022;43:341-347.
13. Laudin GE, Lakha AB, Dullabh N, et al. A meta-analysis of cases of Rosai Dorfman disease reported on the African continent and a description of two cases from a tertiary academic hospital in Johannesburg, South Africa. *Pan Afr Med J*. 2023;45:130.
14. Beg M, Saleem T, Zubari A, et al. Maxxillofacial Rosai Dorfman disease in a newly diagnosed HIV-infected patient. *Int J Infect Dis*. 2008;12:218-219.
15. Nasany RA, Reiner AS, Francis JH, et al. Rosai-Dorfman-Destombes disease of the nervous system: a systematic literature review. *Orphanet J Rare Dis*. 2022;17:92-11.
16. Hariz H, Marzouk S, Salah RB, et al. Rosai-Dorfman disease in a 23-year-old patient. *J Clin Case Rep*. 2013;03:8-10.
17. Alrabiah K, Aractingi S, Sohier P. Clinical and diagnostic characteristics of Rosai Dorfman disease: a case series. *Int J Innov Res Med Sci*. 2022;7: 291-297.
18. WHO. *WHO Global Tuberculosis Report 2022*. WHO; 2022.