ISSN: 0975-833X

Available online at http://www.journalcra.com

International Journal of Current Research
Vol. 17, Issue, 06, pp.33319-33320, June, 2025
DOI: https://doi.org/10.24941/ijcr.49124.06.2025

INTERNATIONAL JOURNAL OF CURRENT RESEARCH

RESEARCH ARTICLE

ROSAI-DORFMAN DISEASE IN A 16-YEAR-OLD FEMALE: A RARE CASE REPORT

Dr. Mallinath S M[1], Dr. R L Meena[2], Dr. Shailendra Gupta[3], Dr. Rajveer Bunker[4], Dr. Hitesh Sharma[5], Dr. Mohit Gupta[6] and Dr. Shiwangi Chhanwal[7]

[2]Senior Professor and Unit Head, Department of Medicine, RNT Medical College Udaipur
[1,5,6,7]Junior Resident, Department of Medicine, RNT Medical College Udaipur
[3,4]Assistant Professor, Department of Medicine, RNT Medical College Udaipur

ARTICLE INFO

Article History:
Received 09th March, 2025
Received in revised form
21st April, 2025
Accepted 19th May, 2025
Published online 24th June, 2025

Key words:

Rosai-Dorfman Disease, Pediatric Lymphadenopathy, Cervical Swelling, Histiocytosis, Case Report.

Corresponding author:
Dr. Mallinath S M

ABSTRACT

Rosai-Dorfman Disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy, is a rare, benign, self-limiting histiocytic proliferative disorder. We report a case of a 16-year-old female presenting with a one-month history of left-sided cervical swelling. Histopathological examination confirmed the diagnosis of RDD. This case highlights the importance of considering RDD in pediatric lymphadenopathy to avoid misdiagnosis and over treatment.

Citation: Dr. Mallinath S M, Dr. R L Meena, Dr. Shailendra Gupta, Dr. Rajveer Bunker, Dr. Hitesh Sharma, Dr. Mohit Gupta and Dr. Shiwangi Chhanwal. 2025. *"*Rosai-Dorfman Disease in a 16-Year-Old Female: A Rare Case Report*". International Journal of Current Research, 17, (06), 33319-33320.*

INTRODUCTION

Rosai-Dorfman Disease (RDD) is a rare non-Langerhans cell histiocytosis first characterized by Rosai and Dorfman in 1969[1]. It typically presents as painless bilateral cervical lymphadenopathy and affects children and young adults [3] . RDD can mimic other lymphoproliferative or infectious disorders, leading to diagnostic challenges. This case emphasizes the need for awareness of RDD, especially in the pediatric population. The incidence of RDD is estimated to be less than 1% of all benign histiocytic disorders, with no known geographic or ethnic predisposition. The disease is often sporadic, although familial clustering has been occasionally reported, suggesting a possible genetic component[6]. The etiology of RDD remains unclear, though infectious agents like Epstein-Barr virus (EBV) and Human herpesvirus 6 (HHV-6) have been implicated in some cases, suggesting a possible viral trigger[7].

Case Presentation: A 16-year-old female presented with a painless, progressive swelling on the left side of the neck since one month (Fig-1). There was no associated fever, weight loss, or systemic symptoms. Physical examination revealed a firm, mobile, non-tender lymph node approximately 4*3 cm in size. Hematologic and biochemical tests were normal. Imaging confirmed localized cervical lymphadenopathy without extranodal involvement. Excisional biopsy of the lymph node was performed. Histopathology showed dilated sinuses filled with large histiocytes demonstrating emperipolesis. Immunohistochemistry was positive for S100 and CD68, which is consistent with Rosai-Dorfman disease[2.] There were no systemic symptoms such as night sweats, fatigue, or loss of appetite, and the patient's school performance had not been affected. Mantoux test and serology for Epstein-Barr virus, HIV, and toxoplasmosis were negative, helping to rule out the infectious causes of lymphadenopathy[8]. On laboratory investigation complete hemogram, Blood culture and sensitivity were found to be normal.

Ultrasound Neck Region: There is evidence of hypoechoic collection measuring approximately 21mm*15mm with internal echoes seen in left side of the neck. Multiple enlarged conglomerated lymph nodes are seen in left jugular chain region.

FNAC and Biopsy: Microscopic Examination: Stained smear (Fig. 2) shows numerous histiocytes with abundant pale blue vacuolated cytoplasm and few histiocytes showing intracytoplasmic lymphocytes and plasma cells against the

Fig. 1. Showing left sided neck swelling

background of lymphocytes, neutrophils, plasma cells and RBCs. At some places granuloma like clusters of histiocytes with epithelioid features also seen. Picture suggestive of Sinus Histiocytosis.

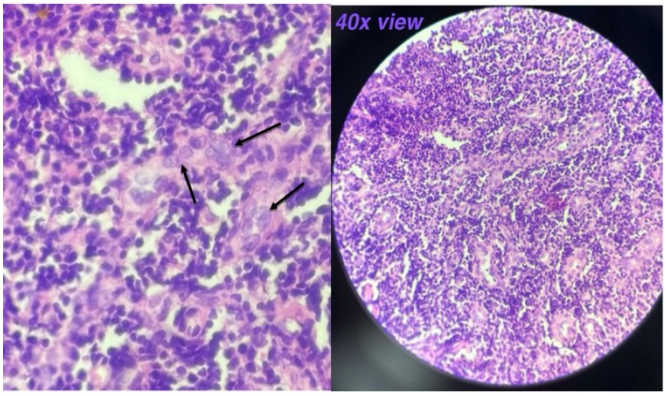

Fig. 2. Histopathology of lymph node showing cluster of histiocytes (arrow marked)

(Lab ref no. – 4673, Hospital reg no. – 10947242, Dept. of Pathology, RNT Medical College Udaipur)

DISCUSSION

RDD is a benign condition, but its presentation can mimic serious pathologies such as lymphoma or tuberculosis. Emperipolesis and immunohistochemical profile are key diagnostic features[2]. While the etiology remains uncertain, infectious and immune mechanisms are hypothesized. Most cases are self-limiting, but treatment may be warranted for symptomatic or systemic disease[4]. This case reinforces the importance of histopathological confirmation in persistent pediatric lymphadenopathy [5].

RDD must be differentiated from other causes of lymphadenopathy, such as tuberculosis, infectious mononucleosis, and malignancies including Hodgkin and non-Hodgkin lymphoma[9]. In some cases, RDD can coexist with autoimmune disorders like systemic lupus erythematosus and rheumatoid arthritis, indicating a potential role of immune dysregulation [10].

CONCLUSION

Rosai-Dorfman Disease is a rare cause of pediatric lymphadenopathy. A high index of suspicion and histopathological confirmation are essential for accurate diagnosis. Early recognition can prevent unnecessary investigations and treatments.

Patient Consent: Written informed consent was obtained from the patient for the publication of this case report.

Conflict of Interest: The authors declare no conflicts of interest.

Funding Statement: No funding was received for this study.

REFERENCES

1. Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy. Arch Pathol. 1969;87(1):63–70.
2. Foucar E, Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease): review of the entity. *Semin Diagn Pathol*., 1990;7(1):19–73.
3. Komp DM. Sinus histiocytosis with massive lymphadenopathy. *Pediatr Clin North Am*., 1985;32(4):901–910.
4. Dalia S, Sagatys E, Sokol L, Kubal T. Rosai-Dorfman disease: tumor biology, clinical features, pathology, and treatment. Cancer Control. 2014;21(4):322–327.
5. Chikuie T, *et al*. Pediatric Rosai–Dorfman disease: a diagnostic challenge. *J Pediatr Hematol Oncol*. 2021;43(3):e394–e397.
6. Komp DM. Sinus histiocytosis with massive lymphadenopathy. *Pediatr Clin North Am*. 1985;32(4):901–910.
7. Levine PH, *et al*. Detection of human herpesvirus 6 in nodal and extranodal Rosai-Dorfman disease. *J Infect Dis*., 1992;166(2):291–295.
8. Abla O, *et al*. Consensus recommendations for the diagnosis and clinical management of Rosai-Dorfman-Destombes disease. *Blood*. 2018;131(26):2877–2890.
9. Pulsoni A, *et al*. Treatment of Rosai-Dorfman disease. *Haematologica*. 2000;85(10):1097–1100.
10. Gaitonde S. Multifocal, extranodal sinus histiocytosis with massive lymphadenopathy: a review. *Arch Pathol Lab Med*., 2007;131(7):1117–1121.

****