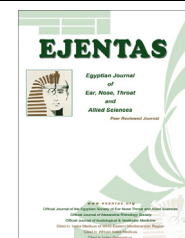




Egyptian Society of Ear, Nose, Throat and Allied Sciences  
**Egyptian Journal of Ear, Nose, Throat and Allied Sciences**

[www.ejentas.com](http://www.ejentas.com)



## CASE REPORT

# Rare case of Rosai Dorfman disease involving paranasal sinuses in paediatric patient: A case report



Gaurav Ashish <sup>a,\*</sup>, Ramanathan Chandrashekharan <sup>a</sup>, Harshad Parmar <sup>b</sup>

<sup>a</sup> Department of ENT, Christian Medical College, Vellore, India

<sup>b</sup> Department of Pathology, Christian Medical College, Vellore, India

Received 17 May 2015; accepted 24 November 2015

### KEYWORDS

The Rosai–Dorfman disease;  
 Histiocytosis;  
 Cervical node;  
 Nasal cavity;  
 Emperipolesis

**Abstract** Rosai Dorfman disease (RDD) is a rare, benign disease of unknown aetiology. It typically presents with massive, painless cervical lymphadenopathy but may have a varied presentation. We report a paediatric case of RDD with initial isolated involvement of nasal cavity and PNS with subsequent involvement of cervical lymph nodes. Endoscopic biopsy confirmed the pathological diagnosis and he was managed successfully with medical therapy. At 20 month follow up there was no evidence of recurrence. The ideal protocol for the treatment is still debated. A long term follow-up is warranted to detect relapses. This case report is illustrated aiming at developing insights into management and diagnosis of such rare clinical entity in a paediatric patient.

© 2015 Egyptian Society of Ear, Nose, Throat and Allied Sciences. Production and hosting by Elsevier B.V. All rights reserved.

## 1. Introduction

Rosai Dorfman syndrome is an immune mediated histoproliferative disease described by Rosai and Dorfman in 1969. Other theories like infectious origin have also been suggested however the immunological theory has been the most accepted one.<sup>1</sup>

It commonly affects children and young adults. It is marked by massive cervical lymphadenopathy with a low grade fever. Detailed investigative workup may show anaemia, leucocytosis and polyclonal hypergammaglobulinemia. The enlarged nodes show the proliferation of sinusoidal histiocytes. Approximately 43% of cases show extra nodal involvement wherein

the possible sites are skin, central nervous system, ear, and orbit and rarely nose and paranasal sinuses.<sup>2</sup>

Usually the treatment of choice is medical therapy using a cocktail of steroid and cytotoxic drugs as in our case steroid and cyclophosphamide were administered. However in refractory cases or in cases where the adenopathy is causing pressure effects, treatment options like radiation and surgery can also be considered. This entity has an indolent course with a possibility of spontaneous remission even after long periods. Hence these patients require a long term follow up.<sup>3</sup>

## 2. Case report

An eight year old child presented to the outpatient department with a history of progressive left sided nasal block for the last 4 months with associated intermittent epistaxis for the last

\* Corresponding author.

Peer review under responsibility of Egyptian Society of Ear, Nose, Throat and Allied Sciences.

<http://dx.doi.org/10.1016/j.ejenta.2015.11.004>

2090-0740 © 2015 Egyptian Society of Ear, Nose, Throat and Allied Sciences. Production and hosting by Elsevier B.V. All rights reserved.

1 month. He also gave a history of bilateral rapidly progressive neck swelling for the last 1 month. He has no other known comorbidities.

Neck examination revealed multiple bilateral cervical lymphadenopathies. Level 2 lymph node on the left side and level 1b on right side were significantly enlarged measuring 5 cm × 5 cm and 1.5 cm × 1.5 cm, respectively. He had no other palpable lymph nodes or organomegaly. Haematological and immunological workups were irrelevant. A diagnostic nasal endoscopy revealed a friable mass involving the left lateral wall and inferior turbinate going up to the choana.

MRCT scan T2 images revealed a 3.4 × 2.7 × 2.4 cm sized T2 isointense lesion in the left nasal cavity (Fig. 1) The mass was involving the left inferior turbinate, lateral nasal wall with bowing of the medial wall of the maxillary sinus (Fig. 2) with posterior extension up to the nasopharynx (Fig. 3). Histopathological examination was consistent with extra nodal sinus Histiocytosis (Figs. 4 and 5) with massive lymphadenopathy (Rosai–Dorfman disease). He was treated successfully with oral steroid and low dose cyclophosphamide and at 20 month follow up no evidence of recurrence or residual disease was noted. Repeat nasal endoscopy showed no lesion in the nasal cavity and there were no palpable cervical lymph nodes.

### 3. Discussion

Rosai Dorfman syndrome, well known as Sinus Histiocytosis with Massive Lymphadenopathy (SHML), is a rare benign systemic histio-proliferative disease commonly involving cervical group lymph nodes.<sup>1</sup> It was first illustrated by Azoury and Reed and later by Rosai and Dorfman in 1969.<sup>4</sup>

Presently SHML is classified as one of the non-Juvenile Xanthogranulomas (non-JXG) under the sub-group of the non-Langerhans Cell Histiocytosis (non-LCH).<sup>5</sup>

It commonly affects a younger age group, usually in the first and second decades with a male-to-female preponderance of 1.4:1.<sup>6</sup>

Aetiology is still debated, however immune mediated theory or infections caused by Epstein Barr Virus, brucella or klebsiella have been the two most proposed theories.<sup>2</sup>



Figure 1 CT scan coronal view.

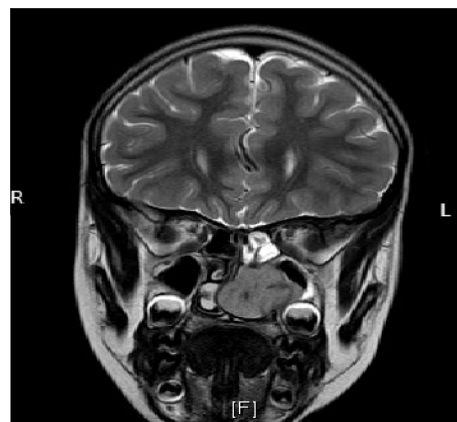


Figure 2 MRI T2 image in coronal view.



Figure 3 MRI T1 sagittal view.

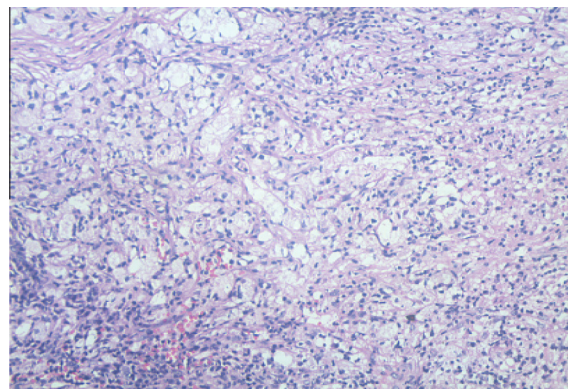
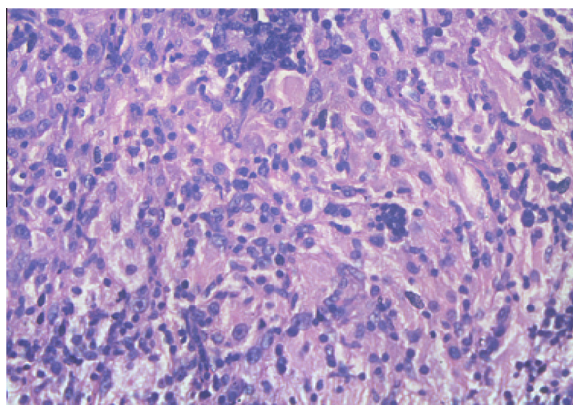


Figure 4 H&E stain – showing cells arranged in solid sheets, with round to oval, irregular, vesicular nuclei, prominent nucleoli and moderate amounts of eosinophilic to vacuolated cytoplasm. The background contains many plasma cells and lymphocytes.

Lymph nodes are usually involved but in about 43% of cases, however extra nodal involvement is also well known. Frequently involved extra nodal sites are skin, mucosa, spinal cord, pancreas, nasal cavity and major salivary glands.<sup>7</sup>

Those affected with this disease present with massive, painless, bilateral or unilateral cervical lymphadenopathy with other nodal sites such as axillary, inguinal, para aortic and mediastinal lymph nodes.<sup>8</sup>



**Figure 5** H&E stain – showing evidence of Emperipolesis.

Histopathologically the affected lymph nodes demonstrate extensively distended sinuses, with abundant histiocytes with vesicular nuclei, distinct nucleoli and abundant pale cytoplasm demonstrating Emperipolesis. Emperipolesis was proposed by Humble et al., in 1956 as, “the active penetration of one cell by another which remains intact”. It differs from phagocytosis because the engulfed cell exists within another cell, remains viable, and can exit without disturbing the physiological and morphological aspects.<sup>9</sup> Emperipolesis till date is considered as an important pathognomonic feature of Rosai Hoffman’s disease. This may be less evident in extranodal involvement.<sup>10</sup>

Histological differential diagnosis includes lymphomas, Hodgkin’s disease, Langerhans Cell Histiocytosis X (LCH) monocytic leukaemia and metastatic carcinoma.<sup>11</sup> Characteristic Immunohistochemical features including CD 68 and CD 1a positivity, positive staining for S-100 and anti-chymotrypsin help us to accurately diagnose SHML.<sup>12</sup>

The haematological picture usually demonstrates anaemia, leukocytosis, neutrophilia and increased ESR.<sup>10</sup>

CT and magnetic resonance imaging should be used to assess the territory of the lesion, more frequently if it involves head and neck.<sup>13</sup>

Clinical scenario has generally insidious onset, protracted duration of active disease state and eventual spontaneous remission, but recurrences have also been reported occasionally.<sup>14</sup>

The treatment options include surgery, chemotherapy, and radiotherapy. However surgical option has been proposed to give most promising results by far. RDD in nose and paranasal sinuses can be treated by endoscopic approach or by external approach such as craniofacial resection or midfacial degloving, depending on the extent of the disease.<sup>15</sup>

Medical therapy for RDD includes corticosteroids, cytotoxic agents or both. Promising results have been obtained by a combination of corticosteroid (prednisolone) along with agents like vinca alkaloid (vincristine and vinblastine), 6-mercaptopurine, methotrexate, or alkylating agent (cyclophosphamide).<sup>16</sup>

This clinical entity has an indolent course with approximately 50% of them resolving without any residual disease. However in 1/3rd of patients residual adenopathy is noted and remaining approximately 17% have unresolved symptoms even after 5–10 years.<sup>3</sup> In our case the child showed no residual or recurrence at 20 month follow up.

This disease frequently demonstrates spontaneous remission. Pulsoni et al. demonstrated in his series of 80 patients that 80% of patients had full remission.<sup>17</sup> Surgical treatment becomes important in cases of enlarged tissue with significant symptoms such as compression on vital organs or airway obstruction.<sup>18</sup>

#### 4. Conclusion

Our case demonstrates a rare clinical scenario of isolated involvement of nose and paranasal sinuses with subsequent involvement of bilateral cervical lymph nodes manages successfully via medical therapy. The treating doctor needs to be aware of the pathognomonic clinical features and hallmark diagnostic features of this condition to be able to diagnose it accurately. The disease process undergoes a frequent spontaneous remission indicating that conservative, expectant management is sufficient in most patients, but more aggressive surgical treatment may be necessary in selected cases if warranted, however consensus for treatment is controversial.

#### Conflict of interest

None.

#### References

1. Snehalatha R, Senthilnathan P, Ramani P, Herald SJ. The Rosai Dorfman syndrome-the report of a rare case. *J Clin Diagn Res (JCDR)*. 2013;7(7):1519–1521.
2. Kare M, Dang S, Dang A. Rosai Dorfman syndrome with sinonasal mucosa and intraocular involvement. *J Assoc Physicians India*. 2007;55:448–450.
3. Raveenthiran V, Dhanlaxmi M, Hayavadana Rao PV, Vishwanathan P. Rosai Dorfman disease: report of a 3 year old girl with review of treatment options. *Eur J Paediatr Surg*. 2003;13:350–354.
4. Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy. A newly recognized benign clinicopathological entity. *Arch Pathol*. 1969;87(1):63–70.
5. Weitzman S, Jaffe R. Uncommon histiocytic disorders: the non-Langerhans cell histiocytoses. *Pediatr Blood Cancer*. 2005;45(3):256–264.
6. Goodnight JW, Wang MB, Sercarz JA, Fu YS. Extranodal Rosai–Dorfman disease of the head and neck. *Laryngoscope*. 1996;106(3 Pt 1):253–256.
7. Azoury FJ, Reed RJ. Histiocytosis. Report of an unusual case. *N Engl J Med*. 1966;274(17):928–930.
8. Sacchi S, Artusi T, Torelli U, Emilia G. Sinus histiocytosis with massive lymphadenopathy. *Leuk Lymphoma*. 1992;7(3):189–194.
9. Humble JG, Jayne WH, Pulvertaft RJ. Biological interaction between lymphocytes and other cells. *Br J Haematol*. 1956;2(3):283–294.
10. Foucar E, Rosai J, Dorfman R. Sinus histiocytosis with massive lymphadenopathy (Rosai–Dorfman disease): review of the entity. *Semin Diagn Pathol*. 1990;7(1):19–73.
11. NDJ 2011 – 68374 [Internet]. <<http://www.ajol.info/index.php/ndj/article/viewFile/77978/68374>>; 2013 cited 13.09.13.
12. Paulli M, Bergamaschi G, Tonon L, et al. Evidence for a polyclonal nature of the cell infiltrate in sinus histiocytosis with massive lymphadenopathy (Rosai–Dorfman disease). *Br J Haematol*. 1995;91(2):415–418.
13. Kumar B, Karki S, Paudyal P. Diagnosis of sinus histiocytosis with massive lymphadenopathy (Rosai–Dorfman disease) by fine needle aspiration cytology. *Diagn Cytopathol*. 2008;36(10):691–695.

14. Parida PK, Panda NK, Sharma A, Mahesha V, Das A. Varied manifestations of Rosai–Dorfman disease and its management – a case report and review of literature. *Indian J Otolaryngol Head Neck Surg Off Publ Assoc Otolaryngol India*. 2008;60(4):365–368.
15. Annessi G, Giannetti A. Purely cutaneous Rosai–Dorfman disease. *Br J Dermatol*. 1996;134(4):749–753.
16. Jabali Y, Smrcka V, Pradna J. Rosai–Dorfman disease: successful long-term results by combination chemotherapy with prednisone, 6-mercaptopurine, methotrexate, and vinblastine: a case report. *Int J Surg Pathol*. 2005;13(3):285–289.
17. Pulsoni A, Anghel G, Falcucci P, et al. Treatment of sinus histiocytosis with massive lymphadenopathy (Rosai–Dorfman disease): report of a case and literature review. *Am J Hematol*. 2002;69(1):67–71.
18. Komp DM. The treatment of sinus histiocytosis with massive lymphadenopathy (Rosai–Dorfman disease). *Semin Diagn Pathol*. 1990;7(1):83–86.