BULLETIN OF STOMATOLOGY AND MAXILLOFACIAL SURGERY
Volume 21, Issue 11

DOI: 10.58240/1829006X-2025.21.11-114

ORIGINAL ARTICALE

EXTRANODAL ORBITAL HISTIOCYTOSIS IN ROSAI-DORFMAN DISEASE: A CASE REPORT AND LITERATURE REVIEW

Heba Elmetwally Abdallah Farahat[1],[2]*, Sara Ali Albishi[3], Abeer Ali El-Sherbiny Ateya[4], Sally Daoud Mohamed[5] , Abdul Qadeer Memon[6], Maha M Abdul-Latif[7]

[1]*Assistant Professor, Department of Radiology and Medical Imaging,College of Applied Medical Sciences , Prince Sattam bin Abdulaziz University, Al-Kharj, Saudi Arabia.

[2]*Department of Radiology, Mansoura Specialized Hospital, Mansoura, Egypt.Email: Heba.elmetwally@yahoo.com , h.farahat@psau.edu.sa, ORCID ID: https://orcid.org/0009-0003-7949-0929

[3]PhD in Nutritional Science, Assistant Professor, Clinical Nutrition Department, College of Applied Medical Science, University of Hafr Al-Batin, Hafr Al-Batin, Saudi Arabia.Email: saalbishi@uhb.edu.sa, ORCID ID: https://orcid.org/0009-0001-7930-2743

[4]Assistant Professor, Department of Medical Laboratory, College of Applied Medical Sciences, Prince Sattam bin Abdul-AzizUniversity,Al-Kharj,SaudiArabia.Emails: abeer_alielsherbiny@yahoo.com; a.ateya@psau.edu.sa, ORCID ID: https://orcid.org/0009-0006-8788-3652

[5]Lecturer, Department of Radiology and Medical Imaging,College of Applied Medical Sciences, Prince Sattam bin Abdulaziz University, Alkharj, Saudi Arabia, Email: S.daoud@psau.edu.sa, ORCID ID: https://orcid.org/0009-0002-1618-0758

[6]Assistant Professor, Department of Surgery, College of Medicine, King Faisal University, Saudi Arabia
Emails: drqadeermemon@yahoo.com; amuhammad@kfu.edu.sa, ORCID ID: https://orcid.org/0000-0002-7071-9489

[7]Assistant Professor, Ophthalmology Surgery Department, Faculty of Medicine, Northern Border University, Saudi Arabia.
Email: maha.m.latif79@gmail.com, ORCID ID: https://orcid.org/0000-0002-3865-5817

*Corresponds Author: Heba Elmetwally Abdallah Farahat, [1]*Assistant Professor, Department of Radiology and Medical Imaging, College of Applied Medical Sciences, Prince Sattam bin Abdulaziz University, Al-Kharj, Saudi Arabia
[2]*Department of Radiology, Mansoura Specialized Hospital, Mansoura, Egypt.Email: Heba.elmetwally@yahoo.com , h.farahat@psau.edu.sa, ORCID ID: https://orcid.org/0009-0003-7949-0929

Received:*Oct 29. 2025;* ***Accepted:*** *Nov 27, 2025;* ***Published:*** *Dec 5, 2025*

ABSTRACT

Rosai-Dorfman disease (RDD) is a rare disorder characterized by sinus histiocytosis with massive lymphadenopathy. It most commonly affects young males. Most patients were presented with enlarged cervical lymph nodes, pyrexia and fatigue. It primarily happened in the lymph nodes. Extranodal disorder can be present in eye, salivary gland, respiratory tract, skin, skeleton, brain layers and testis. A 14-year old male presented with proptosis associated with throbbing pain. MRI and DWI MRI,were done. MRI and DWI revealed a diffusely infiltrative orbital soft tissue mass with eyelid swelling.

Keywords**:** Rosai-Dorfman disease, MRI, DWI MRI.

1-INTRODUCTION

Rosai-Dorfman disease (RDD),first described as a unique pathological disorder commonly seen in male pediatric group and young adults.[1]

This disease is characterized by relapsing course. Patients presented with fever, leukocytosis, and painless enlarged cervical lymph nodes.[2]

RDD shares similarities with many orbital disorders as inflammatory pseudo tumors, both benign and malignant infiltrative orbital tumors, optic nerve lesions and lymphoma.[3,4]

Aim of the work: MRI and DWI MRI are highly recommended in diffuse infiltrating orbital masses to narrow the differential diagnosis.

2-Case presentation

A 14- year old male presented to our hospital by proptosis associated with throbbing pain and soft tissue swelling.The Patient had no history of virus infection. Visual acuity was 20/20.An ophthalmological examination showed left inter orbital soft masses, with slight tenderness. Physical examination revealed bilateral cervical lymph nodes enlargement.

Pre and post contrast MRI and DWI were done ,that revealed soft tissue lesion in left intra conal ,extra conal spaces infiltrating retro bulbar fat, rectus muscles and eye lid ,the mass showed low SI on T1WI and mixed hypo intense SI on T2WI ,after contrast injection the mass showed diffuse heterogeneous enhancement.

The diffusion images revealed free diffusion.In our department, we diagnosed the lesion as diffuse inflammatory pseudotumor as clinical and radiology support our diagnosis ,we also exclude lymphoma from our diagnosis as lymphoma mostly associated with restricted diffusion due to high cellularity. The patient received antibiotics and steroids with partial improvement, followed by surgery, as curative and histopathology was done that confirm the diagnosis of RDD.Then patient advised to take short course of chemotherapy for six months.

3- Patient methods and results

Patient was prepared for MRI examinations with multiple sequences were applied; slice thicknesses (3 mm) and magnetic field strengths (3-T Signa GE Healthcare).

Pre and post contrast MRI were performed,and diffusion weighted images were done by acquiring conventional T2WI with addition of strong diffusion gradients using a multisession single shot spin echo EPI sequence with TR/TE/NEX: 5800/139 ms/1 with b value of 0, 500, and 1000 mm²/sec.

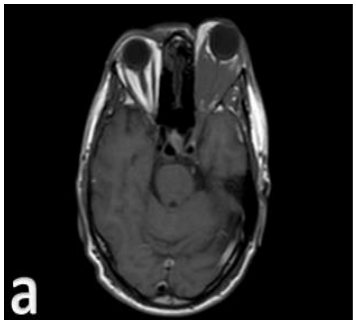
MRI revealed left orbital enhancing diffused infiltrative multicompartmental soft tissue lesion affected intra conal, extraconal spaces and orbital muscles.It measures 2x2.5x3 cm (Ap x Tr x height respectively).It displayed low SI on T1WI and mixed hypo intense SI on T2WI , the mass showed heterogeneous enhancement after contrast injection (Fig 1&2). The mass showed free diffusion, ADC ratio measure 1.9×10−3mm2/s (Fig3).

4-Histopathology

A histopathological analysis was conducted.There were patches of necrosis and fibrosis in the removed bulk. Histological analysis revealed a mixture of intense histiocytic growth and phagocytosis (emperipolesis) of inflammatory cells, primarily plasma cells and lymphocytes. Large histiocytes aggregated with different types of plasma cells, lymphocytes, and uncommon neutrophils, according to H&E staining. (Fig 4).The specimen showed S-100 protein positive.

5-Sequels

Surgical excision was done, and chemotherapy was advised with follow up observations.



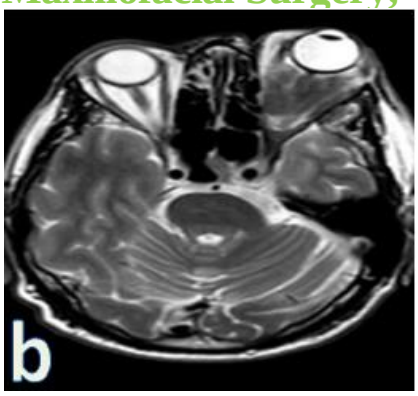
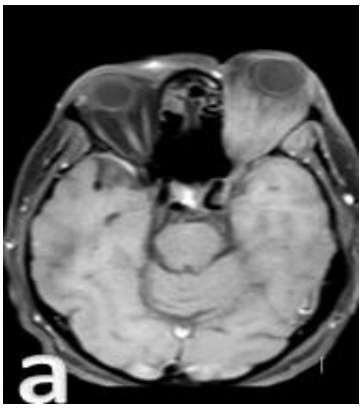


Figure 1. (a&b): A case of 14-year-old boy with painful proptosis:MRI orbit was done , axial T1WI and T2WI images (a&b) respectively show diffuse infiltrative soft tissue orbital mass affecting intraconal,extraconal spaces, retro bulbar fat and orbital muscles. It measures about 2x2.5x3cm respectively (Ap x Tr x H) respectively It displayed low SI on T1W1 and mixed SI on T2WI.



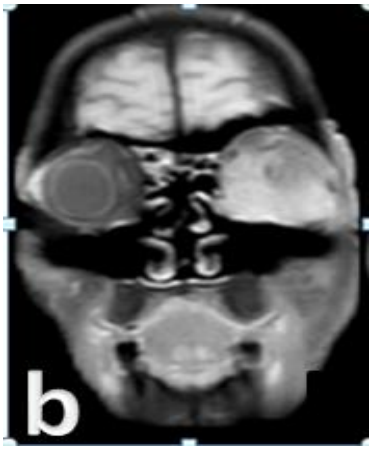


Fig2 (a&b): post contrast MRI revealed heterogeneous enhancement of the pervious described mass in axial and coronal images respectively.

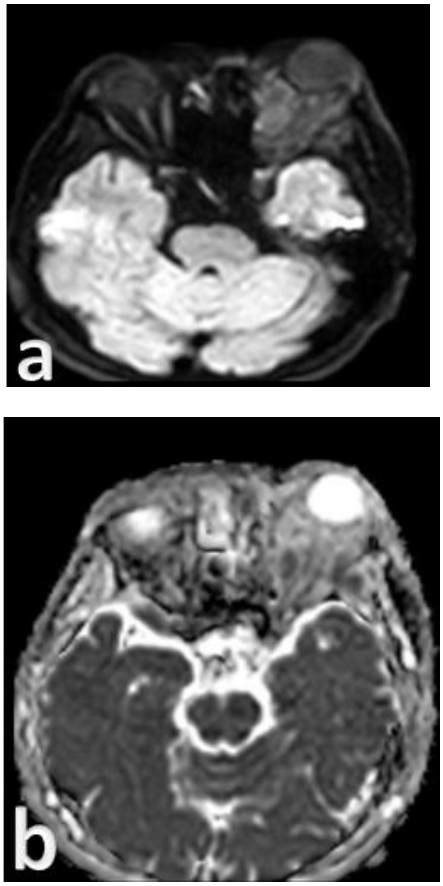


Figure 3(a& b): Diffusion MRI displayed low intense SI on DWI and high intense SI on ADC (Free diffusion). ADC ratio measures 1.9×10−3mm2/s.

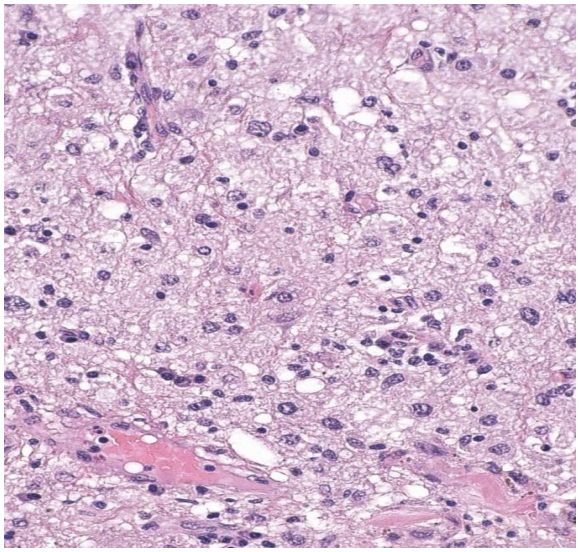


Figure 4. Histopathology demonstrated histiocytes with abundant clear cytoplasm With fine vacuoles and reactive background lymphocytes and plasma cells.

DISCUSSION

Demographic data

Rosai–Dorfman disease (RDD) is a rare and self-limiting benign disease process that presents most commonly in male young patients. It is marked by more histiocytes in lymph nodes, However, extranodal disorder is rare.[5] In RDD, orbital involvement has seldom been documented on its own; it is typically unilateral and infrequently bilateral.[6] and this is agree with our study that reported a young age male of unilateral left orbital soft tissue swelling .

Etiology

The actual cause of RDD remains unclear. Infection by human herpes virus-6, Epstein–Barr virus (EBV),and Parvovirus B19 may be one of the causes. Multiple genetic alternations including , mutations have also been reported.[7]

In the current study laboratory investigations (CBC, virology and immunological tests) were done, and revealed free results.

Classifications and clinical features

RDD can be classified into 3 groups lymph node, extra nodal, and mixed types according to the site of the lesion, However, the extra nodal type and mixed type are rare.[8] In our study extranodal orbital RDD was reported.

Clinical manifestations as pyrexia, leukocytosis, weight loss, and cervical lymph nodes enlargement in individuals with RDD may be similar to multiple diseases as LCH, lymphoma and diffuse optic nerve infiltrations.[9,10]

Histopathology is mandatory for confirming the diagnosis. The disease tends to be multifocal associated with painless lymphadenopathy.[11,12]

Diagnosis

MRI and DWI play a role in detection of the lesion and characterization of it as it appeared of low SI on TWI and mixed hypo intense SI on T2WI with definite boundary with homogenous enhancement without detection of orbital osteolytic lesions in our study and this is agree with Najafi-Sani et al[3],that noted that SI of most orbital RDD of low to intermediate SI on T2WI .

RDD is mostly diagnosed by histopathology, which is defined by histiocyte expansion and proliferation linked to a high number of mature plasma cells, lymphocytes, and normal phagocytic lymphocytes in the cytoplasm. Although it is present in only 70% of cases, Castellano-Sanchez[13] observed that Emperipolesis, which indicates the phagocytosis of lymphocytes, is a feature of Rosai-Dorfman illness. This is consistent with our investigation following the H,E stain, which showed larger histiocytes phagocytes lymphocytes.

Differential Diagnosis

In our study the patient had a diffuse infiltrative orbital mass that has broad differential diagnosis, as lymphoma,

pseudo tumor, lymphoid hyperplasia and IG related diseases (Table 1).

Table1. Differential Diagnosis of (Rosai-Dorfman disease)

Inflammatory disease (orbital inflammatory pseudo tumor)
Thyroid orbitopathy
Sarcoidosis
Lymphoma
Metastasis

In our case diagnosis of Lymphoma was excluded ,as free diffusion was detected but pseudo tumor was the first possibility in our diagnosis as a patient complaint of painful orbital swelling associated with proptosis and infiltrative soft tissue mass.

Najafi-Sani et al. [3]reported that Orbital histiocytosis differ than RDD as an orbital histiocytosis characterized by diffuse infiltrative orbital lesion associated with osteolytic bone lesion but RDD affects soft tissue of orbit only .This opinion coincides with our study as no obvious bone lesions was detected, only soft tissue orbital infiltrations were detected.

Prognosis and Strategy of treatment

A biopsy is safe, essential for verifying the diagnosis of cancers, and effective for making an accurate diagnosis, as demonstrated by Bijlsma et al[14].

Extranodal RDD typically manifests as a benign condition with a favorable prognosis.

The primary treatment for RDD is surgical resection, which should be augmented with chemotherapy, radiation, or steroids, following a biopsy for a definitive diagnosis. If masses are physically accessible, surgical removal has been demonstrated to be the most successful treatment strategy for orbital RDD. For severe instances of RDD, chemotherapy and systemic corticosteroid therapy are recommended [15].

CONCLUSION

Rosai-Dorfman disease was suspected in children and young adults with a unilateral or bilateral slowly progressive proptosis manifesting with a rubbery firm mass, with or without cervical lymph nodes enlargement. A biopsy is mandatory to confirm the diagnosis.

Learning point

1- Orbital RDD usually presents as soft tissue orbital swelling which can be associated with exophthalmos or proptosis.

2- Orbital RDD can be mistaken for peri-orbital cellulitis or diffuse infiltrative orbital masses, both benign and malignant tumors, lymphoma, orbital histiocytosis and optic nerve lesions.

3- Orbital RDD may be similar in pattern with Langerhans and non-Langerhans histiocytosis as the growth of LCH causes diffuse inflammatory reactions, However in LCH mostly associated with osteolyic bony lesion but RDD affected soft tissue of orbit only.

4- Orbital lesions associated with an intracranial extension are regarded as CNS-risk lesions and need systemic chemotherapy.

5- Post contrast MRI and diffusion weighted image play a great role in limit the differential diagnosis of orbital RDD, however biopsy is the best method confirming the diagnosis.

6- There is no selected therapy for RDD, however treatment has included surgery, radiotherapy, and chemo therapy.

DECLARATIONS

Ethics approval and consent to participate

Not applicable

Conflicts Of Interests

None

Funding

None

Informed consent statement

Oral informed consent to publish this case report including images was obtained from patient's parents

REFERENCES

1. Raslan O, Schillinger D, Gregory N, Fuller GN, Ketonen LM. Rosai-Dorfman Disease in Neuroradiology: Imaging Findings in a Series of 10 Patients. AJR Am J Roentgenol; 2011;196(2): W187-93.
2. Ha H, Kim KH, Ahn YJ, Kim JH, Kim JE, Yoon SS. A rare case of Rosai-Dorfman disease without lymphadenopathy. Korean J Intern Med (Engl Ed) 2016; 31(4):802-804.
3. Najafi-Sani M, Saneian H, Mahjoub F. Rosai-Dorfman Disease with nodal and extranodal involvements: a case report. J Res Med Sci. 2011;16(9):1251–1256.
4. Yoon AJ, Parisien M, Feldman F, Young-In Lee F. Extranodal Rosai-Dorfman disease of bone, subcutaneous tissue and paranasal sinus mucosa with a review of its pathogenesis. Skeletal Radiol 2005;34(10):653-657
5. Mehraein Y, Wagner M, Remberger K, Füzesi L, Middel P, Kaptur S, et al. Parvovirus B19 detected in Rosai-Dorfman disease in nodal and extranodal manifestations. J Clin Pathol 2006;59(12):1320-1326.
6. Al-Maghrabi B, Elnaggar T, Alamri O, et al. Orbital

Rosai-Dorfman disease: a case report and literature review. J Microsc Ultrastruct 2019; 7: 50–52.
7-Garces S, Medeiros LJ, Patel KP, Li S, Pina-Oviedo S, Li J, et al. Mutually exclusive recurrent KRAS and MAP2K1 mutations in Rosai-Dorfman disease. Mod Pathol; 2017;30(10)1367-1377.
8-Gaitonde S. Multifocal, extranodal sinus histiocytosis with massive lymphadenopathy: an overview Arch Pathol Lab Med; 2007;131(7):1117-1121.
9-Su X, Zhang L. Orbital Rosai–Dorfman disease: a case report and literature review. J Int Med Res; 2019; 47(11):5891–5895.
10-Iyengar S, Nishanth S. Michelle W, Hill T, Tang K, Gardner S. Orbital Rosai-Dorfman disease initially diagnosed as IgG4-related disease: a case report. Acta Neuropathol Commun; 2020; 8(1):113.
11-Xu Q , Fu L , Liu C. Multimodality imaging-based evaluation of Rosai-Dorfman disease in the head and neck: A retrospective observational study. Medicine (Baltimore); 2018;96(51): e9372.
12-Deen I U, Chittal A, Badro N, Jones R, Haas C. Extranodal Rosai-Dorfman Disease- a Review of Diagnostic Testing and Management. J Community Hosp Intern Med Perspect; 2022; 12(2):18–22.
13-Castellano-Sanchez AA, Brat DJ. 57-year-old-woman with acute loss of strength in her right upper extremity and slurred speech. Brain Pathol 2003;13:641-2,645.
14-Bijlsma WR, Elbert NJ, Kalmann R. The role of biopsy in diagnosing patients suspected of idiopathic orbital inflammation. Curr Eye Res 2012; 37: 251–253.
15-McClain KL, Natkunam Y, Swerdlow SH. Atypical cellular disorders. Hematology Am Soc Hematol Educ Program; 2004; 283-296.