



Mini Review

Volume 6 Issue 2 - September 2021  
DOI: 10.19080/JETR.2021.06.555685

J Endocrinol Thyroid Res

Copyright © All rights are reserved by Samasuk Thammachantha

# Rosai-Dorfman Disease with Pituitary Compression: Mini-review



**Samasuk Thammachantha\***

Department of Pathology, Neurological Institute of Thailand(NIT), Bangkok, Thailand

Submission: July 16, 2021; Published: September 15, 2021

\*Corresponding author: Samasuk Thammachantha, Department of Pathology, Neurological Institute of Thailand(NIT), 312, Ratchawithi Rd, Thung Phaya Thai, Ratchathewi, Bangkok, 10400, Thailand

**Abstract**

Rosai-Dorfman disease is group of histiocytic disorder. This disorder is idiopathic and presented with nodule/mass. Intracranial lesion of RDD usually manifest with compression effect. Pituitary lesion can affect nearby pituitary structures. Surgery is the treatment of choice. Other etiologies must be excluded, such as infection, meningioma and IgG4-related disorder.

**Keywords:** Histiocytes; Pituitary gland; Rosai-Dorfman disease

**Introduction**

Rosai-Dorfman disease(RDD) is one of histiocytic disorder. Intracranial RDD has been rarely reported and usually manifested as dural-based mass. Nearly all articles described about mass effect. However, this disorder possibly presented with pituitary hormonal dysfunction. The author reviews this disorder and discuss about histological diagnosis and management.

Emile et al, a member of the Histiocyte Society (HS), divided this disorder into 5 groups; that is L (Langerhans), C (cutaneous and mucocutaneous), M (malignant), R (Rosai-Dorfman), and H (hemophagocytic). The RDD is categorized in R group [2]. The classic (nodal) RDD presented with lymphadenopathy, meanwhile the extranodal type manifested elsewhere. Most frequent sites of extranodal RDD are skin (16%), nasal cavity (16%), eye and adnexa (11%), bone (11%), salivary gland (7%), and central nervous system (7%) [3]. Intracranial RDD usually occurs without extracranial lesion, and most are attached with the dura [4] (Table 1).

**Discussion**

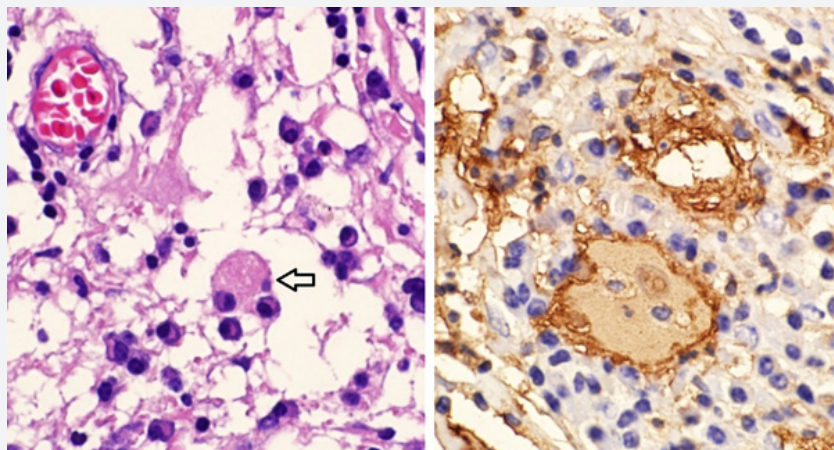
RDD was discovered in 1969, characterized by reactive bilateral lymphadenopathy with fever, hence it was named "Sinus histiocytosis with massive lymphadenopathy (SHML)" [1]. Recently,

**Table 1:** Summary of histiocytic disorder.

| Disorder                      | Clinical Features   | Localization                                       | Histology   | Special Studies   |
|-------------------------------|---|--|---|---|
| Langerhans cell histiocytosis | Child (often <10 years old); DI                           | Skull tumor with secondary CNS spread Hypothalamus | Nuclear grooves or folds Eosinophil-rich infiltrate | CD1a <sup>+</sup> , S-100 <sup>+</sup> ; BRAF V600E <sup>+</sup> in subset; Birbeck granules on electron microscopy |
| Rosai-Dorfman disease         | Mimics meningioma Systemic or CNS alone                   | Dural-based  | Emperipolesis Plasma cell-rich infiltrate           | S-100 <sup>+</sup> , CD1a <sup>-</sup> ; mutations of KRAS or MAP2K1 in subset                                      |
| Juvenile xanthogranuloma      | Infant/young child May have cutaneous or systemic disease | Meningeal, ventricular, parenchymal                | Touton giant cells Spindled to foamy cells          | Factor XIIIa <sup>+</sup> , S-100 <sup>+</sup> , CD1a <sup>-</sup>  |

The histological findings of RDD demonstrated mixed inflammatory infiltrates of small lymphocytes and plasma cells. There are large foamy histiocytes, which have large vesicular nuclei and abundant pale cytoplasm. These histiocytes engulf lymphocytes or plasma cells (Emperipolesis). By immunohistochemistry, the histiocytes are positive for CD68, S100 protein, but negative for

CD1a (Figure 1). According to the literature, there has been article showing the RDD with pituitary dysfunction. The patient firstly presented with headache and visual disturbance. Later, the patient revealed the sign and symptom of inappropriate ADH secretion. The mass located at posterior pituitary gland and was resected.



**Figure 1:** Large foamy histiocytic cell engulfs plasma cells (Emperipolesis) (A: Hematoxylin & Eosin, 600x magnification; B: S100, 600x magnification).

Radiotherapy was added and the patient developed diabetes insipidus later 3 years [5]. Another paper mentioned the incidental finding of RDD in autopsy case. The mass situated in posterior pituitary, pars tuberalis, pituitary stalk and the adjacent dura. In addition, the authors found PRL cell hyperplasia which developed by “stalk effect” [6]. Surgery is first-line treatment of choice of RDD cases. Most of patients have good prognosis without recurrence. Additional radiotherapy is still controversial. Recent paper in 2014, recommended to exclude IgG4-related disorder. Especially, RDD case presenting with large numbers of IgG4+ plasma cells [7].

### Conclusion

RDD has been found increasingly nowadays. This lesion can occur anywhere in intracranium. Endocrinologists and neurosurgeon need to know this entity and must exclude infectious etiology firstly. Finally, meningioma and IgG4-related disorder must be rule out also.

### References

1. Rosai J, Dorfman RF (1969) Sinus histiocytosis with massive lymphadenopathy. A newly recognized benign clinicopathological entity. *Arch Pathol* 87(1): 63-70.
2. Emile J-F, Ablu O, Fraitag S, Annacarin Horne, Julien Haroche, et al. (2016) Revised classification of histiocytoses and neoplasms of the macrophage-dendritic cell lineages. *Blood* 127(22): 2672-2681.
3. Shukla E, Nicholson A, Agrawal A, Darshana Rathod (2016) Extra Nodal Rosai-Dorfman Disease (Sinus Histiocytosis with Massive Lymphadenopathy) Presenting as Asymmetric Bilateral Optic Atrophy. *Head Neck Pathol* 10(3): 414-417.
4. Andriko JA, Morrison A, Colegial CH, Davis BJ, Jones RV (2001) Rosai-Dorfman disease isolated to the central nervous system: a report of 11 cases. *Mod Pathol* 14(3):172-178.
5. Kelly WF, Bradey N, Scoones D (1999) Rosai-Dorfman disease presenting as a pituitary tumour. *Clin Endocrinol (Oxf)* 50(1): 133-137.
6. Rotondo F, Munoz DG, Hegele RG, Bruce Gray, Nasima Khatun, et al. (2010) Rosai-Dorfman disease involving the neurohypophysis. *Pituitary* 13(3): 256-259.
7. Menon MP, Evbuomwan MO, Rosai J, Jaffe ES, Pittaluga S (2014) A subset of Rosai-Dorfman disease cases show increased IgG4-positive plasma cells: another red herring or a true association with IgG4-related disease? *Histopathology* 64(3): 455-459.



This work is licensed under Creative Commons Attribution 4.0 License  
DOI: [10.19080/JETR.2021.06.555685](https://doi.org/10.19080/JETR.2021.06.555685)

**Your next submission with Juniper Publishers  
will reach you the below assets**

- Quality Editorial service
- Swift Peer Review
- Reprints availability
- E-prints Service
- Manuscript Podcast for convenient understanding
- Global attainment for your research
- Manuscript accessibility in different formats

**( Pdf, E-pub, Full Text, Audio)**

- Unceasing customer service

**Track the below URL for one-step submission**

<https://juniperpublishers.com/online-submission.php>