



Mini Review

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Rosai-Dorfman Disease with Pituitary Compression: Mini-review



Samasuk Thammachantha*

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

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*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.

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,

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).

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 ⁺ , S-100 ⁺ ; BRAF V600E ⁺ 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 ⁺ , CD1a ⁻ ; 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 ⁺ , S-100 ⁺ , CD1a ⁻

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.

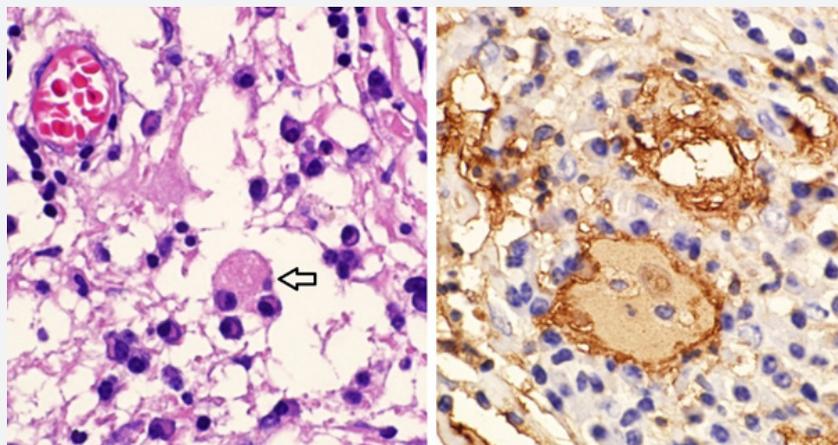


Figure 1: Large foamy histiocytic cell engulfs plasma cells (Emperipolesis) (A: Hematoxylin & Eosin, 600x magnification; B: S100, 600x magnification).

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.

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