

# Radiological and clinical findings of isolated meningeal Rosai–Dorfman disease of the central nervous system

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## Abstract

Rosai–Dorfman disease (RDD) with isolated central nervous system (CNS) involvement is an extremely rare disease. Most RDD of the CNS present as dural-based mass mimicking meningioma and other common lesions, which makes preoperative accurate diagnosis of great difficulty. We searched the pathology database in our hospital and 3 cases of RDD with isolated CNS involvement were finally included in our study. Radiological and clinical findings of these three cases were retrospectively analyzed. The lesions of 2 cases were dura-based against the cerebral convexity, presenting as a sheet-shaped thickened dura mater, another case was located just across the cerebral falx, the dural display in the center was intact. The 3 cases showed low signal intensity on T2-weighted image, obviously enhanced, significantly surrounding edema and finger-like protuberance but no invasion of the brain parenchyma or no sign of hyperplasia or sclerosis of the surrounding cranial bones. In conclusion, when we come across a disease that mimicking meningioma, especially when it manifests as the above radiological features, we should considered it might be a kind of proliferative disease of the meninges, such as RDD.

**Abbreviations:** CNS = central nervous system, FOV = field of view, RDD = Rosai–Dorfman disease, T1-WI = T1 weighted image, T2-WI = T2-weighted image, TE = echo time, TR = repetition time.

**Keywords:** central nervous system, magnetic resonance imaging, meningiomas, Rosai–Dorfman disease

## 1. Introduction

Rosai–Dorfman disease (RDD) was first reported by pathologists Juan Rosai and Ronald Dorfman in 1969.<sup>[1]</sup> RDD is a rare lymphoproliferative disease typically characterized with massive painless cervical lymphadenopathy, fever, weight loss, and leukocytosis. RDD can involve any nodal or extranodal site, but most commonly presents as massive cervical lymphadenopathy. The incidence of extranodal manifestations of RDD is 43% as reported and usually involved the skin, upper respiratory system, orbits, bones, and endocrine glands. Central nervous system (CNS) involvement has been reported, but it is relatively uncommon.<sup>[2–4]</sup> RDD of CNS usually manifests as dura-based masses which is difficult for radiologists to make an accurate diagnosis. In this report, we described 3 cases of isolated meningeal RDD with clinical and radiological presentations

mimicking meningiomas, and review literature to raise the awareness of this disease to avoid misdiagnosis of meningiomas.

## 2. Material and methods

We searched the pathology database at The Second Affiliated Hospital of Zhejiang University School of Medicine from January 2011 to August 2018, and a total of 3 patients with RDD diagnosed were included. This study was approved by the institute review board of Zhenjiang University and informed consent was obtained from the subject. All patients underwent conventional brain MR enhancement scan, with plain scan of T1 and T2 sequences in axial position, coronal, sagittal and enhancement. Images were acquired using a 3.0-T scanner (Discovery MR750, GE Medical Systems, USA.) with an 8-channel head coil. Images were acquired using T1-weighted images (T1-WI) (repetition time [TR] = 500 ms, echo time [TE] = 120 ms) and T2-weighted image (T2-WI) (TR = 500 ms, TE = 15 ms, at the same time with a field of view = 260 × 260 mm<sup>2</sup>, matrix size = 256 × 256, slice thickness = 5 mm). All the 3 patients underwent surgery with total or subtotal resection of the masses. All the histologic sections underwent routine pathological and immunohistochemical examination, the pathological results of case 2 and case 3 were sent to University of California at Los Angeles for consultation. Follow-up after the surgery were performed ranging from 2 months to 14 months, recurrences were observed in 1 case during the follow-up in 14 months.

## 3. Results

### 3.1. Clinical findings

Clinical data of the 3 patients were summarized in Table 1. One woman and 2 men ranging from 40 to 54 years (mean 49.3 years)

Editor: Manoj Kumar.

The authors have no conflicts of interest to disclose.

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Medicine (2019) 98:19(e15365)

Received: 25 October 2018 / Received in final form: 1 March 2019 / Accepted: 30 March 2019

<http://dx.doi.org/10.1097/MD.00000000000015365>

**Table 1****Clinical characteristic of patients with intracranial RDD.**

Case	Sex	Age	Location	Clinical presentation	Preoperative diagnosis	Treatment	Follow-up (month)
1	F	54	Right frontal region	Headache	Proliferative disease of the meninges	Resection	12
2	M	40	Left occipital region	Dizziness	Meningioma	Resection	2
3	M	54	Cerebral falx	Epileptic seizure	Meningioma	Resection	14

F=female, M=male, RDD = Rosai-Dorfman disease.

were involved in this study. Case 1 came to our hospital with a symptom of headache, case 2 was dizziness accompanied by aphasia, and case 3 bore repeated left limb convulsions for more than a year but aggravating in the last month. None of the 3 cases suffered cervical lymphadenopathy or the other extranodal involvement. All the 3 cases demonstrated no unusual results of routine laboratory assays.

### 3.2. Neuroimaging and pathological findings

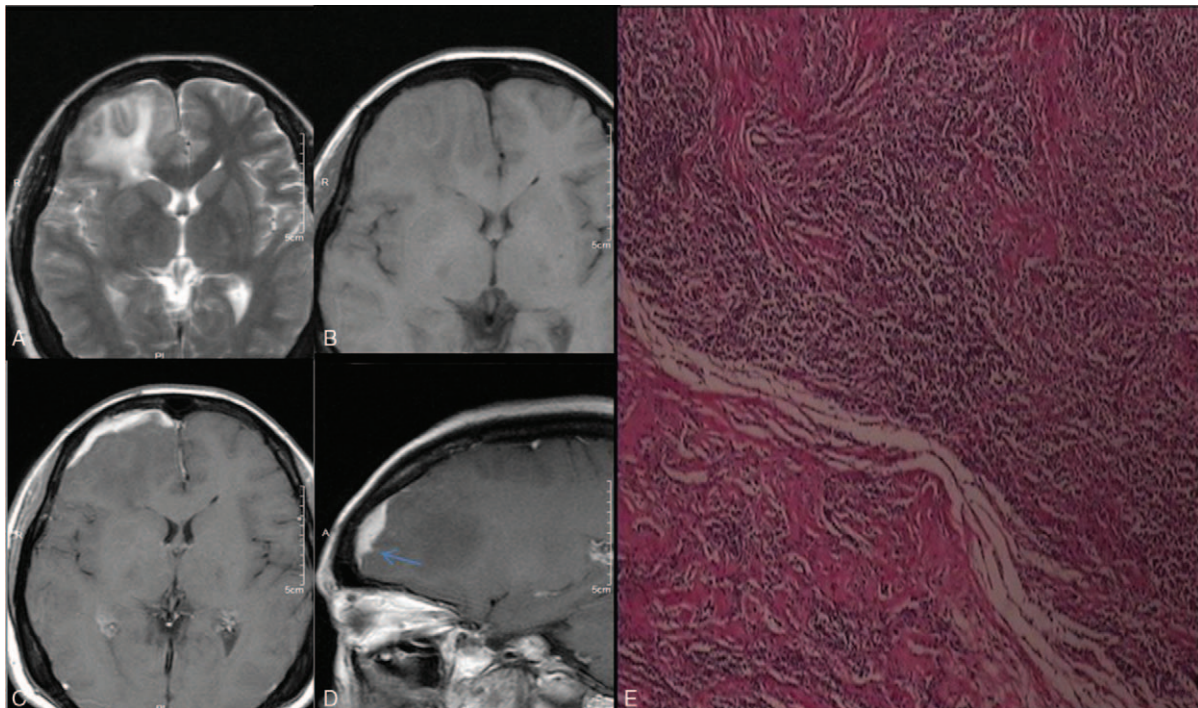
Of the first 2 cases, Brain MRI showed that the lesions were located extraparenchymal and dura-based against the cerebral convexity (Figs. 1 and 2), and case 3 was located just across the cerebral falx (Fig. 3). The thickened meningeal of the first 2 cases on T1-WI and T2-WI was clear, presenting as hypo to isointense on T1-WI, but T2-WI signal of the thickened meninges decreased significantly. On contrast-enhanced T1-WI, the sheet-shaped irregularly thickened meningeal had a

region of a finger-like protuberance (arrow in D in all the 3 cases) extending to the brain parenchyma. The lesion of the case 3 located just across the cerebral falx can be easily identified by T2-WI and T1-WI as a heterogeneous signal, was homogeneously enhanced (Fig. 3). The lesion was symmetrically distributed on the left and the right of the cerebral falx, the dural in the center was intact, but the finger-like protuberance on the left was blurring displayed.

Histopathological examination showed chronic inflammatory cells emperipolesis but without the destructive phagocytosis of plasma cells or lymphocytes. Immunohistochemical results with an expression of CD68 and S-100 protein but CD1a negative confirmed that it was RDD.

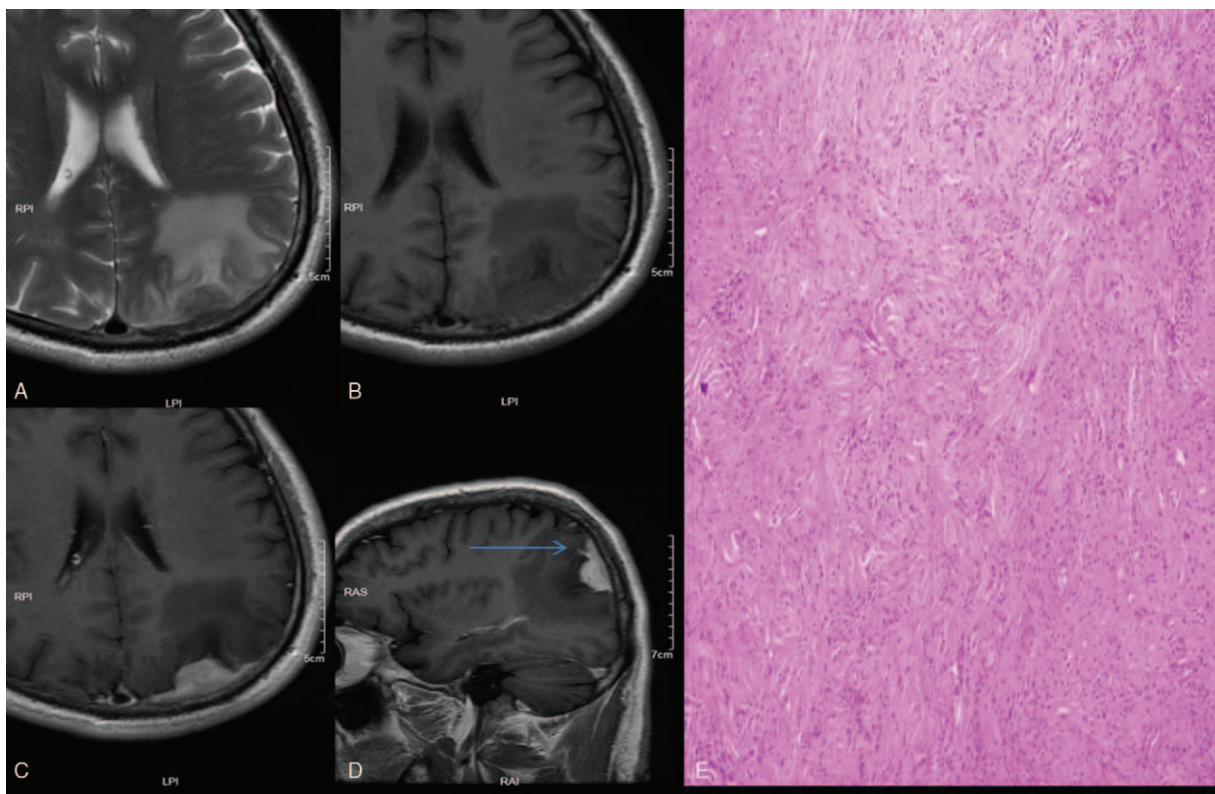
### 4. Discussion

Extranodal RDD occurs in 43% of all the cases.<sup>[2]</sup> The rate of RDD isolated to the CNS is less than 5% as reported.<sup>[4-7]</sup> Most of



**Figure 1.** A 54-year-old woman (case 1) A and B: T2-WI and T1-WI showed the proliferative disease of the meninges on the right frontal region and with significant edema of right frontal lobe, the bone was neither hyperosteo-geny nor sclerosis, the thickened meninges showed obvious low T2 signal. C and D: Axial and Sagittal postcontrast T1-WI revealed an irregularly sheet-shaped thickened dura mater and significantly enhancing, there was a finger-like protuberance (arrow in D) extending to the brain parenchyma. E Histologic section showed the lesion marked by the proliferation of collagen fibers and the infiltration of lymphocytes and plasma cells, accompanied by multiple tissue cells, immunohistochemical examination showed an expression of CD68 and S-100 protein but CD1a negative. T1-WI=T1-weighted image, T2-WI=T2-weighted image.





**Figure 2.** A 40-year-old man (case 2) A and B: T2-WI and T1-WI showed the proliferative disease of the meninges on the left occipital region and with significant edema of left occipital lobe, the bone surrounding the lesion did not cause hyperosteoecy or sclerosis. The thickened meninges showed obvious low T2 signal. C and D: axial and sagittal postcontrast T1-WI showed uneven thickened and significantly enhancing meningeal. The thickened meningeal also had a finger-like protuberance (arrow in D) extending to the brain parenchyma. E: Pathologic studies in local and University of California at Los Angeles showed that S-100 positive tissue cells were mainly mixed infiltration with plasma cells and a phagocytosis of lymphocytes known as emperipolesis, a small amount of the adjacent neural parenchyma was being invaded. T1-WI=T1-weighted image, T2-WI=T2-weighted image.

the RDD of CNS are dural-based mimicking meningioma, which makes it a challenge for accurate diagnosis.<sup>[2]</sup> The hallmark of RDD is the characteristic emperipolesis of lymphatic plasma cells with the background of many plasma cells and lymphocytes, the other characteristic is with an expression of CD68 and S-100 protein but CD1a negative. Immunohistochemical stains are generally sufficient to differentiate RDD.<sup>[8,9]</sup>

As far as we know, there are few articles about imaging to differentiate from meningioma. The lesions of the 3 cases were hypo to isointense on T1-WI and low on T2-WI, possibly reflect their process of inflammatory process. After enhancement, the thickened meningeal is sheet-like in shape with a finger-like protuberance (Figs. 1–3, arrow in D) extending to the brain parenchyma. The lesion of the case 1 and case 2 did not cause cranial hyperplastic sclerosis or bone invasion. We have carefully contrasted the images provided by Huang et al,<sup>[10]</sup> Zhu et al,<sup>[11]</sup> the finger-like protuberance also appeared in their cases, this performance may be directly related to the significant edema around the lesion or possible lead to postoperative recurrence. The dura mater in case 3 shared the same imaging features as the case Forest et al<sup>[12]</sup> reported, which indicates that RDD is a kind of neoplastic state, rarely involving the parenchyma.

RDD and meningioma could show the similar signal intensity on T1 and T2, but when the lesions were inhomogeneous, such

as necrotic, cystic, or calcified, most of this chance, it is likely to be meningioma. Most meningiomas are slow-growing benign tumors always board-based to the dura mater with an enhancing “dural tail.” RDD often resembles an en plaque meningioma, which is a rare type of meningioma characterized by diffuse and extensive dural involvement, and at times invading the bone. In addition, hyperosteoecy and sclerosis at the site are common and meningioma is well known to invade the bone.<sup>[13]</sup> Note that bone invasion is more often seen in en plaque meningioma. Apart from meningioma, the differential diagnosis of RDD includes lymphoma, metastasis, and some other dura-based diseases.

In conclusion, when we come across a disease manifesting as dura-based mass, sheet-like in shape, showed a finger-like protuberance, with a significant edema but without invasion of the brain parenchyma or bone invasion, we should consider it might be a kind of proliferative disease of the meninges, such as RDD.

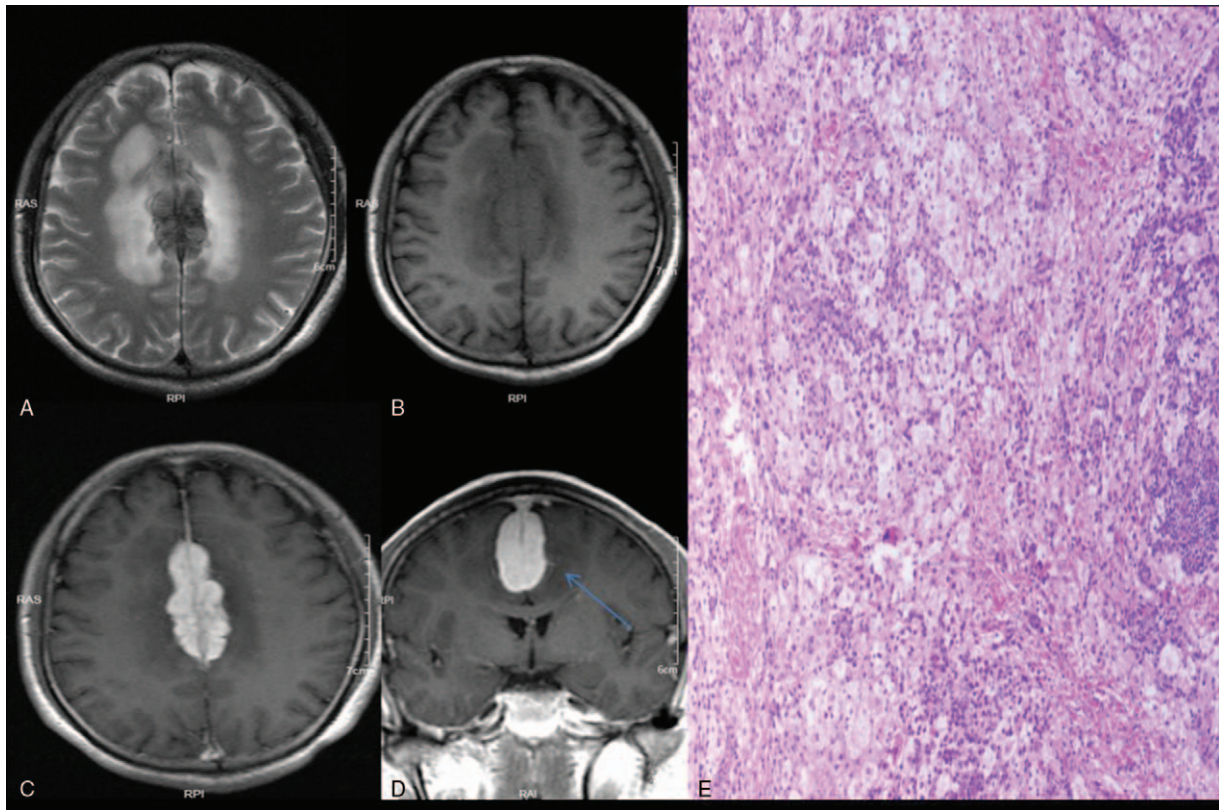
#### Author contributions

**Data curation:** Xiao-juan He.

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**Supervision:** Jie Min.

**Writing – original draft:** Jia-hua Wen.



**Figure 3.** A 54-year-old man (case 3) A–D: axial T2-WI, T1-WI, axial and coronal enhanced T1-WI. A significantly dural-based lesion with an obvious low T2 signal, located right across the cerebral falx, accompanied with significant edema of peripheral brain parenchyma. The lesion was marked enhanced and the dural display in the center was intact. The finger-like protuberance on the left was lightly displayed. E: Histopathological examination showed chronic inflammatory cells and a small amount of neutrophil infiltration. Foam-like cells were rich in cytoplasm, in cytoplasm neutrophil, mononuclear and multi-core cells visible occasionally. T1-WI=T1-weighted image, T2-WI=T2-weighted image.

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