

Case Report

Intracranial Rosai-Dorfman disease mimicking isolated meningioma: a case report and review of the literature

Lin Xia^{1*}, Li Zhang^{2*}, Yue Xiang³, Liping Liu¹, Zonglin Jing¹

Departments of ¹Radiology, ²Neurology, ³Pathology, Affiliated Hospital of North Sichuan Medical College, Nanchong, Sichuan, China. *Co-first authors.

Received March 3, 2021; Accepted May 19, 2021; Epub July 15, 2021; Published July 30, 2021

Abstract: Rosai-Dorfman disease is a rare malignant infirmity. Here, we present a case of a 57-year-old man with giddiness and unstable gait, as well as blurred vision in the left eye for four months. Radiologically the diagnosis before surgery was meningioma. The patient received a craniotomy, and the histopathologic diagnosis was Rosai-Dorfman disease. We reviewed the diagnosis, mechanism, and treatment of this disease.

Keywords: Rosai-Dorfman disease, meningioma, brain, treatment, immunohistochemistry

Introduction

Rosai-Dorfman disease (RDD) was delineated by Rosai and Dorfman initially in 1969, as a benign histiocytic illness, also referred to as sinus histiocytosis with massive lymphadenopathy [1]. RDD ordinarily affects the lymph nodes of young adults [2]. The majority of extranodal RDD are encountered within the skin, orbit, cavity, cavum sinuses, upper respiratory tract, and bone [3]. To the best of our knowledge, isolated intracranial RDD is very rare [4]. Here, we present a case of RDD in a 57-year-old man, who was misdiagnosed with meningioma radiologically before surgery.

Case report

A 57-year-old man, was admitted to our hospital, with dizziness and unstable gait, as well as blurred vision in the left eye for four months. There was a defect in the temporal visual field of the left eye, which was evident on examination. No lymphadenopathy was found on physical examination, and no abnormalities were found on all routine hematological and biochemical tests. A CT enhancement scan showed a lesion, which was homogeneously contrast-enhancing (2.7 cm × 2.9 cm × 1.2 cm

in size), located in the right temporal region (**Figure 1**). Subsequently, the mass showed slightly low signal intensity on T1-weighted images, fairly low signal intensity on T2-weighted images, and low signal intensity on fluid attenuated inversion recovery images. Mild peritumoral brain edema was also noted. Furthermore, on contrast MRI the lesion showed homogeneous enhancement, and in the sagittal T1-weighted contrast-enhanced MRI, a dural tail could be observed along the right temporal region (**Figure 2**). The mass was misdiagnosed as meningioma radiologically. Other examinations, such as tumor biomarkers, were normal. Histologic examination showed an inflammatory cell infiltrate in the fibrous tissue of the mass, which included histiocytes, lymphocytes, and plasma cells. By immunohistochemistry analysis, the histiocytes showed strong expression of S-100 protein, a strong expression of CD68, and positive for CD163 (**Figure 3**). The pathological manifestation of the mass was consistent with the identification of Rosai-Dorfman disease.

During the operation, the lesion was located in the right temporal region, at the base of the middle cranial fossa, prostrate on the skull base, adherent closely to the brain tissue with

Intracranial rosai-dorfman disease

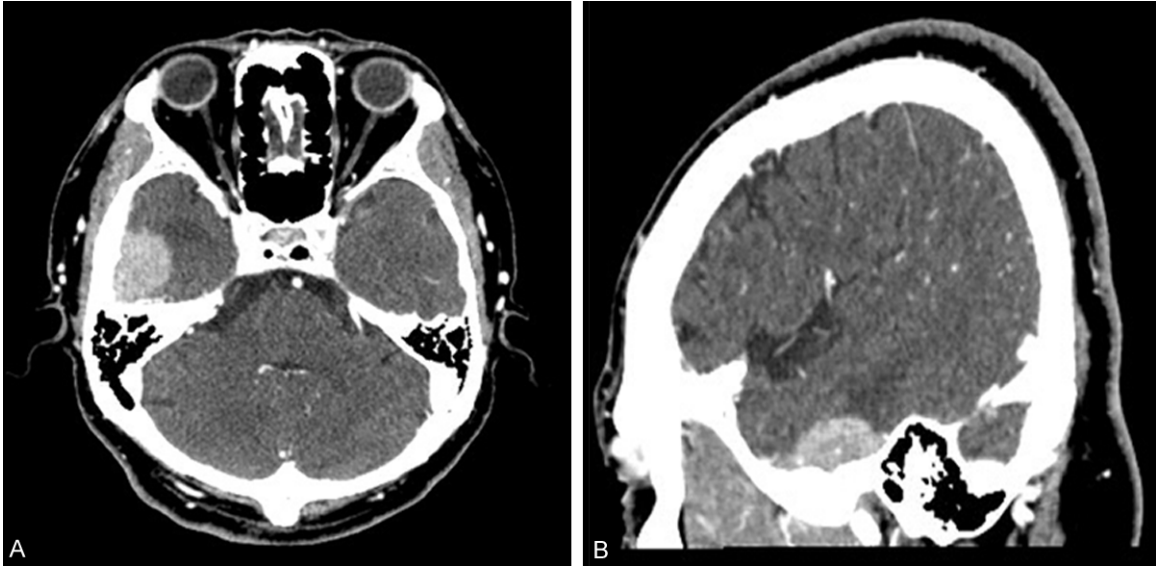
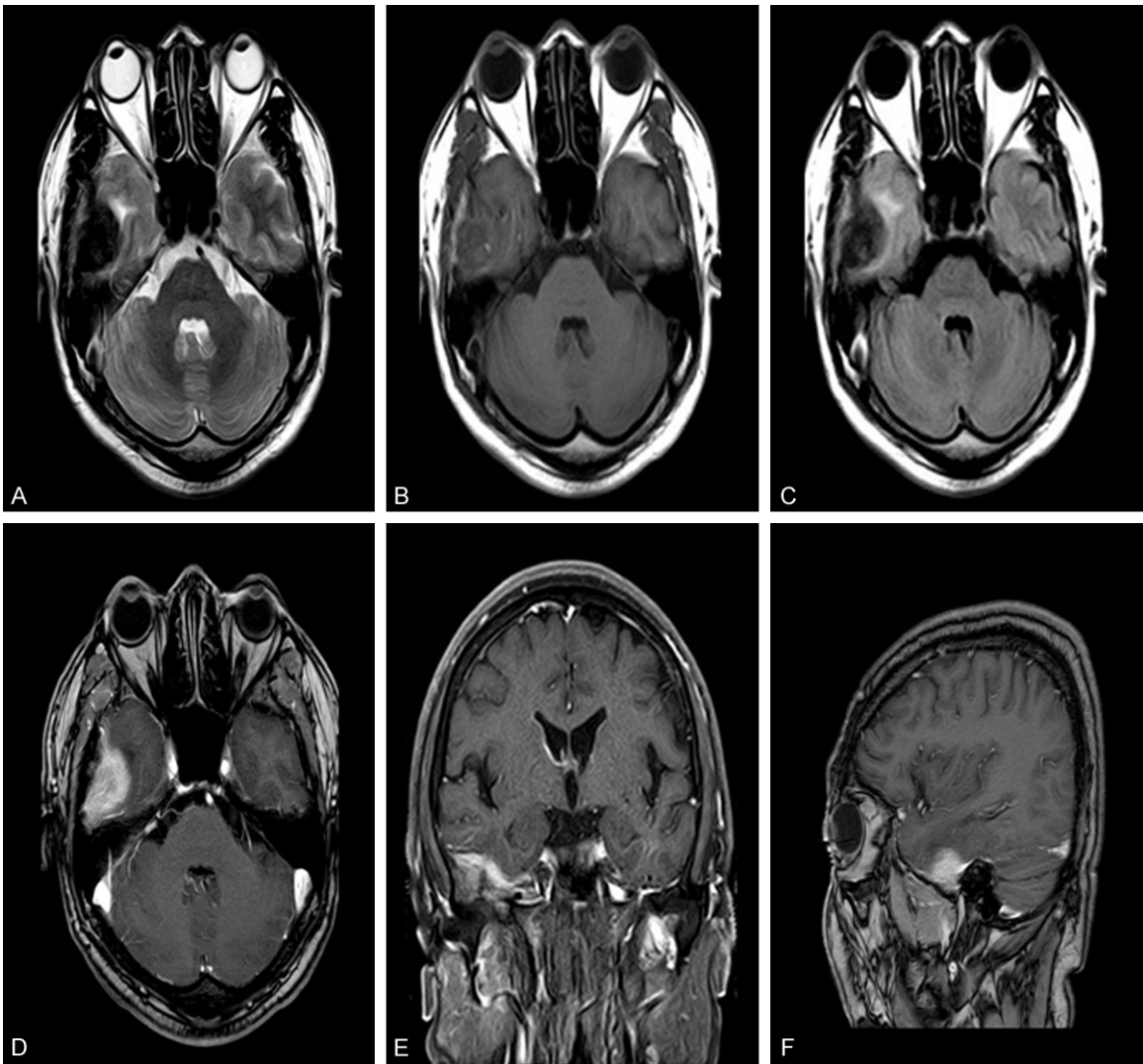


Figure 1. A, B. Axial, sagittal contrast-enhanced CT scan shows a homogeneously enhancing lesion (2.7 cm × 2.9 cm × 1.2 cm in size), with mild peritumoural brain edema, located in the right temporal region (arrow).



Intracranial rosai-dorfman disease

Figure 2. (A) Axial T2-weighted MRI and (C) axial fluid-attenuated inversion recovery MRI reveals a hypointense mass with a mild peritumoural brain edema, in the right temporal region. (B) Axial T1-weighted MRI shows a slightly hypointense mass. (D-F) Axial, coronal, and sagittal T1-weighted contrast-enhanced MRI demonstrate the lesion had homogeneous enhancement. A dural tail was observed along the right temporal region (arrow).

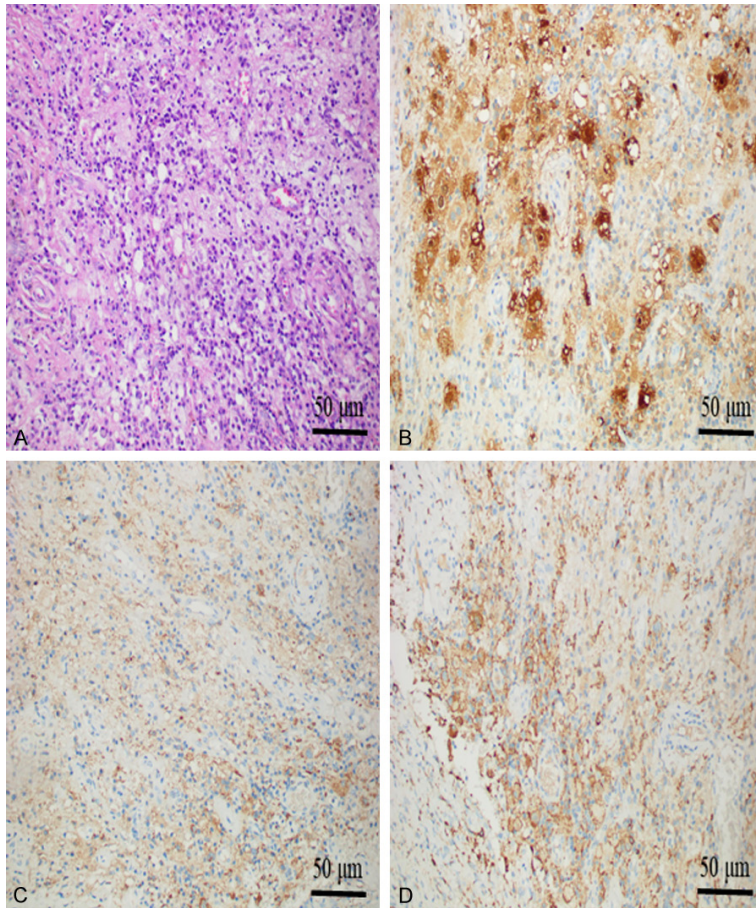


Figure 3. (A) Histologic sections demonstrating fibrous tissue with an infiltrate of inflammatory cells, including histiocytes, lymphocytes, and plasma cells (hematoxylin and eosin stain, $\times 200$). Immunohistochemically, positive reactivity for S-100 protein (B) ($\times 200$) and CD68 (C) ($\times 200$), and positive for CD163 (D) ($\times 200$).

no obvious boundary, and with abundant blood supply. The patient's symptoms improved, and he recovered well after the operation.

Discussion

Rosai-Dorfman disease (RDD) initially was described by Rosai and Dorfman in 1969 [1].

RDD usually involves cervical lymph nodes. It is reported that up to 43% of RDD is involved in the extranodal sinus, orbit, spine, skin, and upper respiratory tract [5]. Neurological involvement is very rare, and less than 5% occur in

RDD patients [2]. Males are dominant, and the ratio of males to females is 1.8:1. Most of cases of CNS manifestation have been located intracranially. An extremely few cases have been seen in the spinal cord. Few patients have suffered both intracranial and spinal RDD lesions [6]. As far as it is known, more than 230 patients with CNS RDD have been reported in the literature [6-10].

Currently, the pathogenesis is not clearly understood. There are several different theories about the etiology of RDD. Jiang et al. think that many factors may lead to the occurrence of RDD, such as immunodeficiency, causes of infection, autoimmune illness, and a neoplastic process [11]. Some researchers have suggested that viruses may be associated with the onset of RDD, such as Epstein-Barr virus, and the virus of human herpesvirus [12]. Other factors, such as gene mutations and immune disorders may be possible factors [13, 14].

The radiologic diagnosis of RDD is full of challenges preoperatively. Due to the characteristic of meningeal masses with uniform contrast enhancement on T1-weighted images, it is highly similar to intracranial meningioma. However, imaging study is very important in the localization of the disease preoperatively. Intracranial RDD typically shows a homogeneous mass with equal signal intensity to soft tissue on CT scans. On MRI scans, it shows a low signal or equal signal on T1-weighted images and high signal on T2-weighted images [11], but the lesion in our case showed hypointense on T2-weighted images. We believed that the increase in fibro-

sis is one of the main factors leading to the decrease of T2WI signal.

Only through pathologic examination can the final diagnosis of RDD be confirmed. This must include histologic and immunohistochemical examinations [6, 7, 10]. Microscopic examination revealed chronic inflammation with infiltration of large amounts of histiocytes, lymphocytes, and plasma cells [9].

There are many treatment choices for intracranial RDD, such as surgery, systemic chemotherapy, radiotherapy, and corticosteroid treatment [2, 4-6].

Conclusion

Although isolated intracranial RDD is a rare disease, the differential diagnosis should include meningioma, lymphoma, and other dural-based masses. Image examination can reveal the extent of the lesion, but definitive diagnosis depends on histopathological and immunohistochemical examination.

Acknowledgements

This study was funded by Scientific Research Development Program of North Sichuan Medical College in 2020 (No. CBY20-QA-Y08). The present study was approved by the Ethics Committee of the Affiliated Hospital of North Sichuan Medical College (Nanchong, China). Written informed consent was obtained from the patient.

Disclosure of conflict of interest

None.

Address correspondence to: Zonglin Jing, Department of Radiology, Affiliated Hospital of North Sichuan Medical College, Nanchong, Sichuan, China. E-mail: jzl325@163.com

References

- [1] Rosai J and Dorfman RF. Sinus histiocytosis with massive lymphadenopathy. A newly recognized benign clinicopathological entity. *Arch Pathol* 1969; 87: 63-70.
- [2] Wan S, Teng X, Zhan R, Yu J, Gu J and Zhang K. Isolated intracranial Rosai-Dorfman disease mimicking suprasellar meningioma: case report with review of the literature. *J Int Med Res* 2008; 36: 1134-1139.
- [3] Buchino JJ, Byrd RP and Kmetz DR. Disseminated sinus histiocytosis with massive lymphadenopathy: its pathologic aspects. *Arch Pathol Lab Med* 1982; 106: 13-16.
- [4] Tang Z, Chen F, Shao Y, Li C and Zhang J. Intracranial Rosai-Dorfman disease mimicking melanoma: a case report and review of the literature. *Int J Clin Exp Pathol* 2020; 13: 1463-1467.
- [5] Gupta K, Bagdi N, Sunitha P and Ghosal N. Isolated intracranial Rosai-Dorfman disease mimicking meningioma in a child: a case report and review of the literature. *Br J Radiol* 2011; 84: e138-141.
- [6] Yang X, Liu J, Ren Y, Richard SA and Zhang Y. Isolated intracranial Rosai-Dorfman disease mimicking petroclival meningioma in a child: Case report and review of the literature. *Medicine (Baltimore)* 2017; 96: e8754.
- [7] Adeleye AO, Amir G, Fraifeld S, Shoshan Y, Umansky F and Spektor S. Diagnosis and management of Rosai-Dorfman disease involving the central nervous system. *Neurol Res* 2010; 32: 572-578.
- [8] Sandoval-Sus JD, Sandoval-Leon AC, Chapman JR, Velazquez-Vega J, Borja MJ, Rosenberg S, Lossos A and Lossos IS. Rosai-Dorfman disease of the central nervous system: report of 6 cases and review of the literature. *Medicine (Baltimore)* 2014; 93: 165-175.
- [9] Tian Y, Wang J, Li M, Lin S, Wang G, Wu Z, Ge M and Pirotte BJ. Rosai-Dorfman disease involving the central nervous system: seven cases from one institute. *Acta Neurochir (Wien)* 2015; 157: 1565-1571.
- [10] Tian Y, Wang J, Ge J, Ma Z and Ge M. Intracranial Rosai-Dorfman disease mimicking multiple meningiomas in a child: a case report and review of the literature. *Childs Nerv Syst* 2015; 31: 317-323.
- [11] Jiang Y and Jiang S. Intracranial meningeal rosai-dorfman disease mimicking multiple meningiomas: 3 case reports and a literature review. *World Neurosurg* 2018; 120: 382-390.
- [12] Huang BY, Zong M, Zong WJ, Sun YH, Zhang H and Zhang HB. Intracranial Rosai-Dorfman disease. *J Clin Neurosci* 2016; 32: 133-136.
- [13] Melki I, Lambot K, Jonard L, Couloigner V, Quartier P, Neven B and Bader-Meunier B. Mutation in the SLC29A3 gene: a new cause of a monogenic, autoinflammatory condition. *Pediatrics* 2013; 131: e1308-1313.
- [14] Breiner A, Dubinski W, Gray B and Munoz DG. A 63 year old woman with white matter lesions and pachymeningeal inflammation. *Brain Pathol* 2013; 23: 225-228.