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

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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
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).
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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-
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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 immunochemical examination.

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Disclosure of conflict of interest

None.

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References