

Isolated Intracranial Rosai-Dorfman Disease: A Case Report of Follow-Up MRI Showing Dynamic Changes of the Lesion

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Abstract

Rosai-dorfman disease (RDD) is a histiocytic proliferative disorder and considered to be extremely rare in the CNS. In this paper, we reported a patient confirmed as isolated intracranial RDD in a 58-year-old woman. MRI imaging without contrast enhancement two years ago found nothing at the onset of the symptoms. Follow-up CT and MRI imaging one year later showed dynamic changes of the lesion from scratch. This report suggests that the symptoms of RDD are earlier than imaging findings. Moreover, we made a systematic literature review on the disease to better understand it.

Keywords: Rosai-Dorfman disease; RDD; Central nervous system

Introduction

RDD, also known as sinus histiocytosis with massive lymphadenopathy, is an uncommon benign histiocytic proliferative disorder of unknown origin with pathognomonic histological and immunohistochemical characteristics [1]. It predominantly affects the lymph nodes but can also be found extranodally in other organs. Primary extranodal sites include the skin, upper respiratory system, orbits, testes, bones, endocrine glands and CNS [2-8]. CNS involvement is unusual and isolated CNS RDD without other body involvement is even more exceptional [8]. We report a case of isolated CNS RDD without other body involvement.

Case Presentation

A 58-year-old Chinese woman with a 2-year history of headache, dizziness, blurred vision and a 3-week history of left facial tics came to our hospital. Neurological examination showed that the patient was with normal mind and without neurological deficit. Systemic lymphadenopathy or cutaneous abnormalities were not identified in physical examination. Cervical and abdomino-pelvic ultrasound and chest X-ray revealed no pathological findings. In routine investigations hematological and biochemical studies were normal.

Follow-up CT and MRI images had been performed for two years every several months. At the onset of these symptoms, cerebral MRI without contrast enhancement showed normal (Figures 1a and 1b). After one year, CT and MRI showed a 2 cm lesion in the right-temporal region, apparently adhering to the dural of the convexity and causing a mass effect and peritumor edema. The lesion appeared isodensity on CT and isointense on T1-weighted and T2-weighted images (Figures 2a-2c). Repeated CT and MRI scans in 6 months and about one year showed the lesion gradually increasing, 2.5 cm and 3.5 cm, respectively.

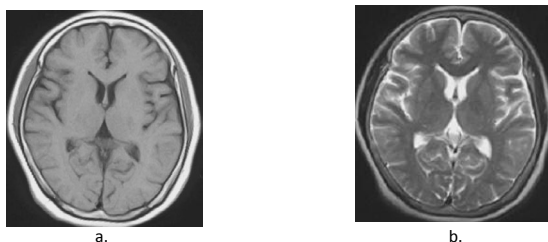


Figure 1: Axial MR images of the head at the onset of the symptoms. T1-weighted (a) and T2-weighted (b) images without contrast enhancement showed normal.

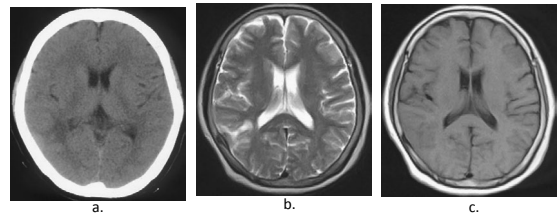


Figure 2: Axial CT and MR images of the head one year later. CT axial image (a) showed an isodensity lesion of 2 cm in diameter in the right-temporal region, apparently adhering to the dural of the convexity and causing a mass effect and peritumor edema. MRI T1-weighted (b) and T2-weighted (c) images showed the lesion appeared isointense.

Contrast-enhanced images on MRI showed diffuse enhancement of the lesion with dural tail (Figures 3a-3d). The appearance of the lesion was better defined to be dural-based by the MRI scan, having the same radiological characteristics with the meningiomas.

The patient underwent surgery and the removal of the right temporal mass was performed by right occipitotemporal craniotomy. The lesion was loosely attached to the dural; however, the lesion was firm and there was no macroscopic tumor infiltration into the adjacent brain.

Microscopically, the tissue was composed of lymphocytes some plasma cells and histiocytes in a background of fibrosis. The histiocytes were aggregated sheets between the lymphocytes and plasma cells. One of the most important findings was the phenomenon of emperipolesis (Figures 4a and 4b). Immunohistochemistry demonstrated the striking positivity of the histiocytes for S100 and CD68 protein while CD1a was negative.

Discussion

RDD was first described by Rosai and Dorfman in 1969 [3]. In 1965,

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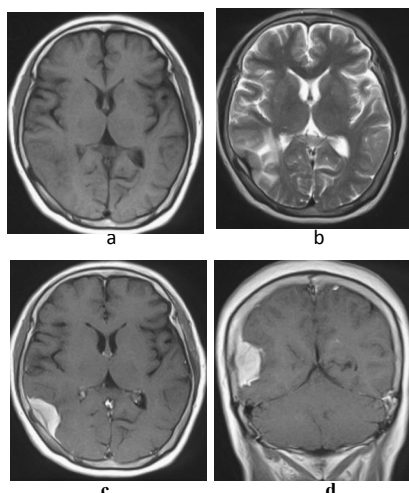


Figure 3: MR images of the head one year later again. Axial T1-weighted (a) and T2-weighted (b) images showed a isointense right-temporal lesion attached to the dural convexity which causes a mass effect and obvious peritumor edema. Axial (c) and coronal (d) T1-weighted contrast-enhanced images revealed a uniform, clear enhancing mass that mimicks meningioma with dural tail.

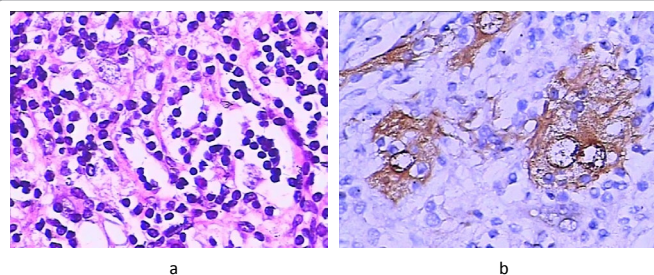


Figure 4: (a) Lesion is composed of attenuated infiltrate of lymphoplasmacytic cells and histiocytic cells which show emperipolesis. (b) Histiocytic cell with emperipolesis is immunopositive for S-100 protein.

Destombes reported four cases of "adenitis with lipid excess" and which got no attention till Rosai and Dorfman four years later characterized the unique pathological features of this rare lymph nodal disorder which they called sinus histiocytosis with massive lymphadenopathy. RDD involves cervical lymph nodes and extranodal sites, which got up to 43% of cases were extranodal. Primary extranodal sites include the skin, upper respiratory system, orbits, testes, bones, endocrine glands and CNS [2]. CNS RDD is considered to be extremely rare, less than 5% of reported cases. However, in this decade, CNS RDD had been a increase [2]. RDD can involve in the spinal and intracranial. 77% of reported cases presented with intracranial, 14% with spine involvement and 9% with both spinal and intracranial disease [5]. Over 90% of CNS RDD occurs in leptomeninges, while rarely found in brain parenchyma [6-9].

The course of RDD is short, about 3-9 months. The clinical manifestations of RDD involving intracranial are similar to Meningioma, which depend on the site of involvement and included cranial nerve deficits, seizures, and increased intracranial pressure [10]. Review of reported intracranial RDD revealed that 70% had no lymphadenopathy and 52% had no associated systemic disease. RDD occurs most commonly in young adult. The mean age of onset with nodal disease is 20.6 years with a male: female ratio of 1.4:1 [7,11]. However, CNS RDD is more common in patients between 40 and 50 years of age, with a male predominance, which the ratio of male to female is 4:1 [12].

Intracranial RDD hasn't characteristic imaging manifestations and normally presents as a meningioma-like, which is most commonly in extraparenchymal, cerebral convexity and the base of skull [2,8,13]. On computed tomography, the lesion may show a homogeneous, lobulated, hyperattenuating mass and moderate-to-marked peritumor edema. Moreover, there may be associated with bone erosion and appear obvious contrast enhancement. However, calcifications are not seen in the intracranial RDD. On T1-weighted magnetic resonance, the lesion may appear homogeneous, lobulated and isointense to the brain parenchyma, with strong homogeneous enhancement after contrast enhancement. On T2-weighted, the lesion may appear heterogeneously hypo-to-isointense, with intensity similar to adjacent dura. The differential diagnosis of intracranial RDD includes non-specific inflammatory disease of the brain parenchyma and meninges, lymphoproliferative disease, intracranial plasma cell granuloma and Langerhans cell histiocytosis [12-16].

Conclusion

In this case, there had no lymphadenopathy and associated systemic disease except headache, dizziness, blurred vision. At the beginning of these symptoms, CT and MRI examination appear normal. After about one-year, repeated CT and MRI images showed an isolated lesion in the right temporal region, which progressively increased. In the course of two years, we documented imaging data of the entire process of occurrence and development of the lesion. From the case, it suggests that imaging examination may appear normal at the onset of some RDD symptoms. For patients with persistent symptoms, the imaging examinations should be enhanced and undergone repeatedly every several months. Intracranial RDD may appear progressive increase, so an operation should be made as early as possible. Intracranial RDD hasn't characteristic imaging appearance and it is difficult to differentiate meningioma and another tumor. The secure diagnosis of intracranial RDD is entirely based on histopathology and immunohistochemistry [7,13,17].

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