

CASE REPORT

Carotid Artery Occlusion in a Patient with Intracranial Rosai-Dorfman Disease

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ABSTRACT

Intracranial involvement of Rosai-Dorfman disease is rare. Central nervous system involvement occurs in less than 5% of patients. To date, there have only been 52 patients with this condition reported in 41 articles. This report is of a patient with a 13-year history of intracranial Rosai-Dorfman disease experiencing occlusion of the internal carotid artery. The clinical, pathological, and radiological features of the disease are discussed, together with a review of the literature.

Key Words: Central nervous system, Internal carotid artery, Occlusion, Rosai-Dorfman disease

INTRODUCTION

Rosai-Dorfman disease (sinus histiocytosis with massive lymphadenopathy) was first described by Rosai and Dorfman in 1969.¹ Since then, more than 650 cases have been reported. Nevertheless, the cause of the disease remains uncertain. Although an extranodal lesion has been recognised, intracranial involvement of Rosai-Dorfman disease is rare. This report is of a patient with intracranial Rosai-Dorfman disease with occlusion of the internal carotid artery. The arterial occlusion was incidentally shown at follow-up magnetic resonance imaging (MRI). The patient was totally asymptomatic.

CASE REPORT

A 35-year-old Chinese man sustained a head injury in 1989. Neurological examination was unremarkable. However, he experienced 2 episodes of tonic-clonic convulsions 2 years later. MRI of the brain in 1992 showed subdural plaques over the left parietal convexity and right tentorium (Figure 1). Blood examination revealed raised C-reactive protein of 228 mg/L (normal range, 0-80 mg/L).

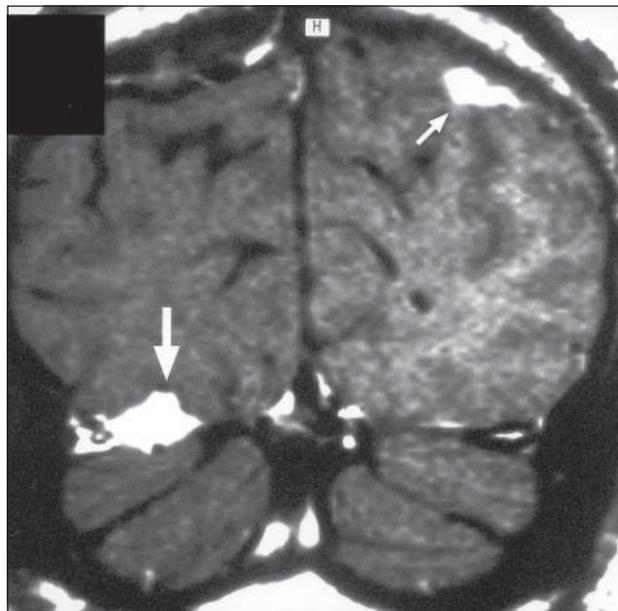


Figure 1. Contrast coronal magnetic resonance images of the brain show enhancing dural-based lesions at the right tentorium (large arrow) and left parietal region (small arrow).

The patient underwent craniotomy and partial excision of the plaques in 1992. Intraoperative findings revealed multiple subdural plaques diffusely involving the meninges. Histological examination showed fibrotic meninges that revealed a variegated appearance with basophilic areas alternating with light-staining areas (Figure 2). The basophilic areas were composed predominantly of lymphocytes and plasma cells with occasional Russell's bodies, while the light-staining

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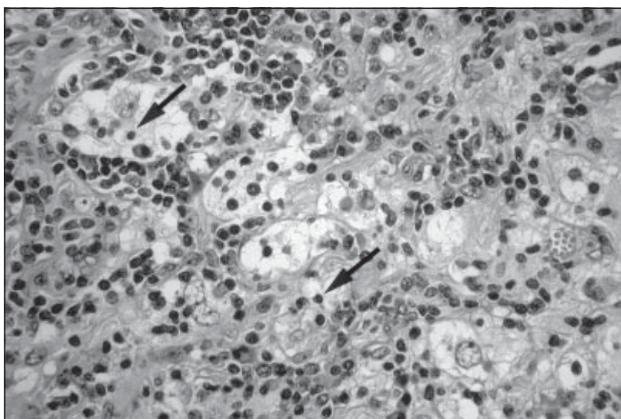


Figure 2. Histological section showing infiltration of lymphocytes, plasma cells and large pale histiocytes with large vesicular nuclei, prominent nucleoli, and abundant pale cytoplasm. Emperipolesis is seen in some of the histiocytes (arrows).

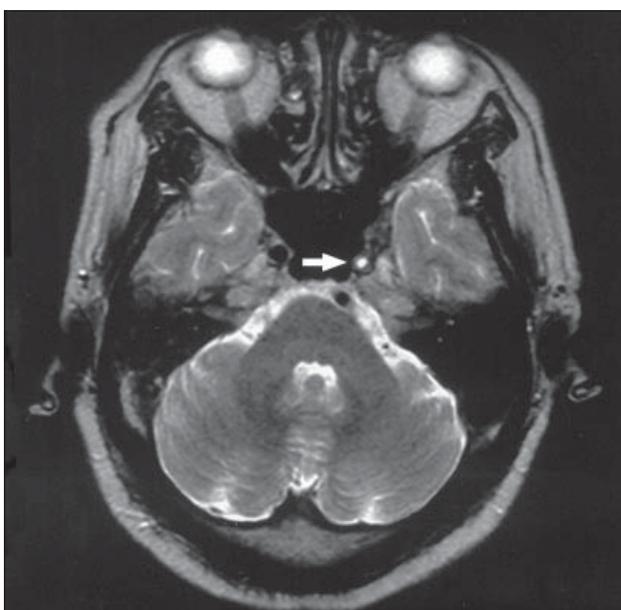


Figure 3. Axial magnetic resonance image of the brain (4000/90) shows hyperintense signal at the cavernous part of the left internal carotid artery (arrow).

areas were formed by numerous histiocytes with large vesicular nuclei and prominent nucleoli. Emperipolesis was noted in some of these histiocytes. Fibrosis was prominent. The pathology was compatible with Rosai-Dorfman disease. The patient underwent cervical lymph node biopsy, which showed no evidence of Rosai-Dorfman disease. He was discharged home with oral steroids and regular follow-up. MRI in 1996 showed that the size of right tentorial plaque remained stable.

In 2002, the patient developed diabetes mellitus that was controlled by oral hypoglycaemic agents. MRI performed in July 2002 demonstrated no interval change of the right tentorial plaque. However, there was an



Figure 4. Magnetic resonance angiogram demonstrates occlusion of the left internal carotid artery (arrow).

abnormal signal with absent flow void in the left internal carotid artery (ICA) [Figure 3]. MR angiography showed left ICA occlusion (Figure 4). The right ICA and the Circle of Willis were patent, and supplied the left anterior and middle cerebral arteries. The intracranial arteries were unremarkable. The brain was normal with no sign of ischaemia. Carotid Doppler ultrasound and computed tomography revealed similar radiological findings. As the patient showed no symptoms of left ICA occlusion, conservative treatment was continued.

DISCUSSION

Rosai-Dorfman disease is an uncommon entity characterised by massive cervical lymphadenopathy associated with fever, leukocytosis, hypergammaglobulinaemia, and increased erythrocyte sedimentation rate. Histologically, the disease shows a polymorphous infiltrate of histiocytes, lymphocytes, and plasma cells in fibrous stroma. The large histiocytes typically demonstrate emperipolesis. They contain intact lymphocytes and are usually S-100 protein immunoperoxidase-staining positive. The cause of Rosai-Dorfman disease remains uncertain. Although some infectious agents² and the disturbance of cell-mediated immunity³ have been postulated as possible causes, no definite causative factor can be identified. The biochemical and radiological studies of Rosai-Dorfman disease are non-specific. The purpose of treatment is to reduce the local aggressiveness of the disease. Surgery seems to be the most effective treatment method. Other treatment modalities

include steroids, radiotherapy, and chemotherapy. The prognosis is correlated with the number of nodal disease sites and the involvement of the extranodal system.⁴ Further information about Rosai-Dorfman disease is available in the literature.⁵

Intracranial involvement in Rosai-Dorfman disease is rare and has been reported in less than 5% of patients.⁶ To date, 54 patients have been reported in 41 articles, including 2 non-English journals.^{7,8} The patients include 35 men and 19 women (male to female ratio, 1.8:1). The mean age of onset was 39.8 years (2 to 78 years), which is significantly higher than that of nodal-based Rosai-Dorfman disease (mean age of onset, 20.6 years)⁹ [$p < 0.0005$ by one sample t-test]. Headache is the commonest symptom, occurring in 29 of the 54 patients. Other symptoms are related to the location of the disease — 90.7% of intracranial Rosai-Dorfman diseases are dural-based, 70.3% of which are solitary, although intraparenchymal and subarachnoid involvement has also been reported.¹⁰ Lymph node involvement is uncommon in patients with intracranial Rosai-Dorfman disease, and is only seen in 16.7% of patients. Nevertheless, 20.4% of patients with intracranial disease have extranodal involvement, such as bone, liver, trachea, breast, abdominal wall, and parotid glands. Most patients underwent operation and 41 patients have follow-up information — 80.5% were clinically stable and 17.1% showed progression of disease. One patient died 5 days after surgery.¹¹ The radiological differential diagnoses of dural-based Rosai-Dorfman disease include meningioma, lymphoma, plasma cell granuloma, Castleman's disease, and Langerhans' cell histiocytosis. The definitive diagnosis can be difficult and biopsy is usually required.

This case is unique in that it is the first reported patient with intracranial Rosai-Dorfman disease having ICA occlusion. As carotid Doppler study and MR angiogram showed normal intracranial and other neck arteries, the arterial occlusion appears to be an isolated involvement. In addition, the absence of symptoms relating to the left ICA occlusion suggests that the occlusion is chronic in nature. Although the patient has diabetes mellitus and is taking steroids long term, these 2 factors are not strongly associated with ICA occlusion. Moreover, other

contributory aetiologies of carotid artery occlusion such as fibromuscular dysplasia or Takayasu's arteritis were not seen in this patient. Therefore, the possibility of the arterial occlusion being a vascular manifestation of Rosai-Dorfman disease cannot be excluded. Only one report discussed the occurrence of vasculitis in a patient with cutaneous Rosai-Dorfman disease.¹² The vasculitis involved only small vessels, which is different from this patient. Patients with intracranial Rosai-Dorfman disease usually have cranial MRI during their follow-up. It is therefore important for radiologists to assess the carotid arteries to detect early occlusion and facilitate proper management.

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