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Case Report Open Access

Isolated Intracranial Rosai-Dorfman Disease Mimicking Convexity Meningioma: A Case Report

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Abstract

Intracranial Rosai-Dorfman disease without systemic involvement is extremely rare. A 59-year-old woman presented with headaches. Magnetic resonance imaging revealed an enhancing posterior fossa convexity lesion with the dural tail sign. The preoperative diagnosis was meningioma. The histopathological examination revealed reactivity for S-100 and CD68 and non-reactivity for CD1a with emperipolesis (lymphophagocytosis) characteristic of Rosai-Dorfman disease.

Keywords: Rosai-Dorfman disease; Meningioma; Emperipolesis

Introduction

Rosai-Dorfman Disease (RDD) is a benign histioproliferative disease of unknown etiology, first described by Rosaiand Dorfman [1], which causes bilateral painless enlarged cervical lymph nodes, fever, leukocytosis, Elevated erythrocyte Sedimentation Rate (ESR), and hypergammaglobulinemia. Extranodal involvement is believed to occur in approximately 25% to 43% of cases [2-4], but isolated intracranial RDD with no systemic involvement is extremely rare [4-16]. It is usually tightly attached to the dura mater and resembles meningioma in both clinical and surgical findings [2,3,6-10,13,15,17-19]. Meningioma usually occurs in middle-aged and older women, whereas RDD is more often seen in children and young adults and has a definite male predominance [4,7,8,17,12]. Histological and immunohistochemical confirmation is essential for definitive diagnosis. In addition to emperipolesis (lymphophagocytosis), reactivity for S-100 protein and CD68, but non reactivity for CD1aimmunostaining are characteristic features of this histioproliferative disease [9,11,15].

Case Report

A 59-year-old female presented with a 7-month history of headaches. Magnetic Resonance Imaging (MRI) at another hospital indicated a neoplastic lesion in the posterior fossa, so the woman was referred to this hospital. There were no neurological deficits, and a physical examination disclosed no extra cranial lesions. Laboratory examination of the blood revealed no abnormalities.

MRI showed a round-shaped mass in the left posterior fossa convexity that showed isointensity on T1-weighted imaging and low intensity on T2-weighted imaging. This was homogeneously enhanced with Gadolinium Diethylenetriaminepenta-Acetic Acid (Gd-DTPA) with the dural tail sign (Figure 1a). T2-weighted imaging revealed perilesional edema and a high signal abnormality in the brain parenchyma (Figure 1b).

The preoperative diagnosis was convexity meningioma. The patient underwent a suboccipital craniotomy, and a total excision of the lesion with the dural attachment was performed. The mass was whitish-gray, vascular, and elastic. It was arising from the cerebellar convexity dura, and loosely adherent to the pia mater.

On histological examination, a round-shaped mass was tightly attached to the dura mater (Figure 2a), and there were sheets of histiocytes interspersed with foci of small lymphocytes and mature plasma cells (Figure 2b). Immunohistochemical staining showed that the histiocytes were reactive for S-100 protein (Figure 2c) and CD68 (Figure 2d), and were nonreactive for CD1a (Figure 2e). The histiocytic cells occasionally showed emperipolesis (lymphophagocytosis) (arrows) (Figure 2d). Four years after surgery, the patient is in good health without recurrence.

Discussion

RDD, also known as Sinus Histiocytosis with Massive Lymphadenopathy (SHML) [11,20], is a benign histioproliferative disease with systemic symptoms and lymphadenopathy that was first described by Rosai and Dorfman in 1969 [1]. It usually causes bilateral painless enlarged cervical lymph nodes, fever, leukocytosis, elevated ESR, and hypergammaglobulinemia. Lesions occur mainly in the systemic lymph nodes, but extranodal involvement has been reported in the skin, nasal cavity, soft tissue, orbit, bone, salivary glands, and central nerves [11]. Isolated intracranial RDD is extremely rare, with approximately 40 previously reported cases in the literature [2-6,9,11-13,15-19,21-24].

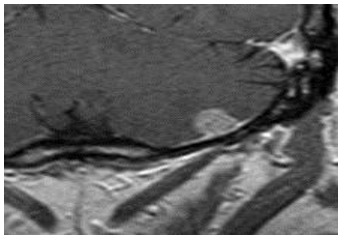

Figure 1: Preoperative MRI
a: Coronal contrast-enhanced T1-weighted MRI showing an enhanced mass with the dural tail sign in the left posterior fossa.

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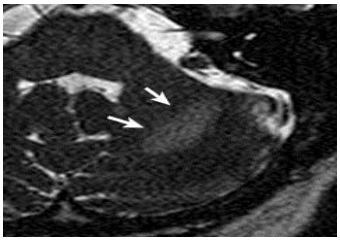

Figure 1: Preoperative MRI
b: Axial T2-weighted MRI showing perilesional brain edema and signal abnormality in the brain parenchyma (arrows).

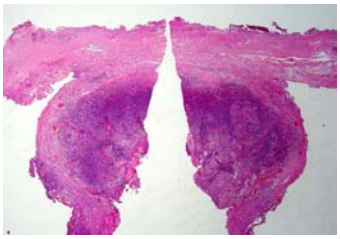

Figure 2: Pathological findings
a: Section of the mass demonstrating a round-shaped mass attached tightly to the dura mater.

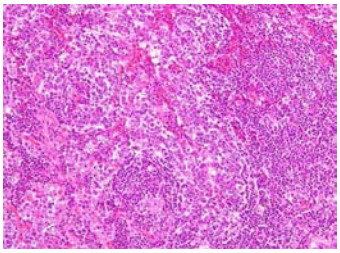

Figure 2: Pathological findings
b: Hematoxylin and eosin-stained specimen showing the sheets of histiocytes interspersed with foci of small lymphocytes and mature plasma cells.

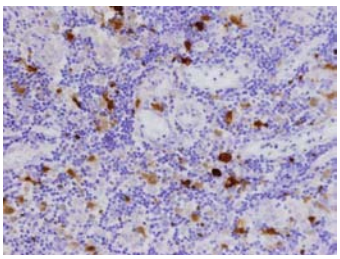

Figure 2: Pathological findings
c: The histiocytes are stained for S-100 protein.

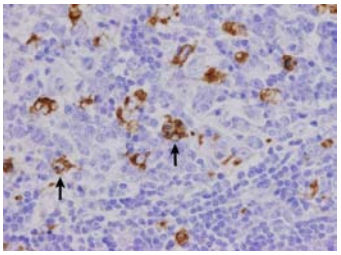

Figure 2: Pathological findings
d: The histiocytes are positive for CD68, demonstrating emperipolesis (lymphophagocytosis) (arrows).

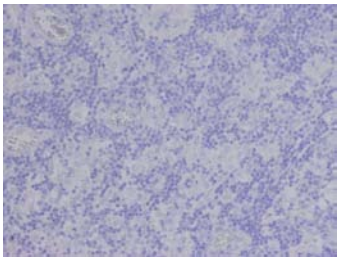

Figure 2: Pathological findings
e: The histiocytes are negative for CD1a.

The most common findings of intracranial RDD on imaging usually suggest a dural based, extra-axial, enhancing mass, and most are resected as meningioma [2,3,6-10,13,15,17-19]. In most cases, however, rather than forming the well-demarcated, lobulated, rounded mass typical of benign meningioma, RDD infiltrates the brain, as shown by the continuous enhancement of the meningeal mass extending to the brain parenchyma, and on imaging it more closely resembles en plaque or malignant meningioma [21,22]. A further characteristic is that, although the lesion is small, brain edema and signal abnormalities on T2-weighted MR images are pronounced in RDD. This may explain why patients commonly experience headaches and seizures despite its small size. However, in cases such as this, when the tumor forms a rounded mass, differentiation from meningioma on preoperative MRI imaging is more difficult [9,13].

Microscopic examination of RDD typically reveals a polymorphous infiltrate of histiocytes, lymphocytes, and plasma cells in a fibrous stroma. The large histiocytes typically exhibit emperipolesis (lymphophagocytosis), which means that the lymphocytes are within the histiocytes. Emperipolesis is not unique to RDD and has been seen in other conditions, but it appears to be a prerequisite for the diagnosis. On immunehistochemical examination, these large histiocytes are reactive for S-100 protein and CD68, and are nonreactive for CD1a, a reliable maker 5 RDD, also known as Sinus Histiocytosis with Massive Lymphadenopathy (SHML) [17,11], is a benign histioproliferative disease with systemic symptoms and lymphadenopathy that was first described by Rosai and Dorfman in 1969 [1]. It usually causes bilateral painless enlarged cervical lymph nodes, fever, leukocytosis, elevated ESR, and hypergammaglobulinemia. Lesions occur mainly in the systemic lymph nodes, but extranodal involvement has been reported in the skin, nasal cavity, soft tissue, orbit, bone, salivary glands, and central nerves [11]. Isolated intracranial RDD is extremely rare, with approximately 40 previously reported cases in the literature [2-9,11,13,15,16-19,22].

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