CASE REPORT

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Rosai-Dorfman Disease in Posterior Fossa

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Rosai-Dorfman Disease(RDD) is an idiopathic histiocytic proliferation affecting lymph nodes. Although extranodal involvement has been reported in the skin, orbit, upper respiratory tract, or testes, the isolated intracranial involvement without associated lymphadenopathy is extremely rare. We report our experience with 1 case of an isolated intracranial RDD without associated lymphadenopathy and any other organ involvement. A 61-year-old male presented with an isolated well-circumscribed brain mass in the posterior fossa, preoperatively thought to be a meningioma. But histology and immunohistochemistry confirmed that the lesion was RDD.

KEY WORDS : Idiopathic histiocytosis without lymphadenopathy · Rosai-Dorfman disease.

Introduction

Sinus histiocytosis with massive lymphadenopathy or Rosai-Dorfman disease(RDD) is a rare but well recognised benign disorder characterized by an unusual proliferation of histiocyte[14]. This histioproliferative disorder is associated with bilateral, painless, cervical lymphadenopathy in 81% of patients[8]. Fever, leukocytosis, elevated sedimentation rate, and polyclonal hypergammaglobulinemia may also be found. In 30% of cases, extranodal involvement is present and may include the skin, orbit, upper respiratory tract, or testes[9]. But isolated intracranial RDD without associated lymphadenopathy is extremely rare. The intracranial involvement is usually a dural-based lesion that mimics a solitary or multiple dural-based meningioma. The case in our study is a solitary intracranial lesion that mimicked a meningioma.

Case Report

A 61-year-old male without prior illness, had a 2-month history of progressive headache and dizziness. Upon presentation, neurological examination was unremarkable. Physical examination did not demonstrate any cutaneous lesions, lymphadenopathy, or hepatosplenomegaly.

A computerized tomography(CT) of the head revealed a 3×2cm sized well enhanced round mass in the right-side posterior fossa(Fig. 1A, B). Meningioma was considered the most likely diagnosis. Routine hematological studies were

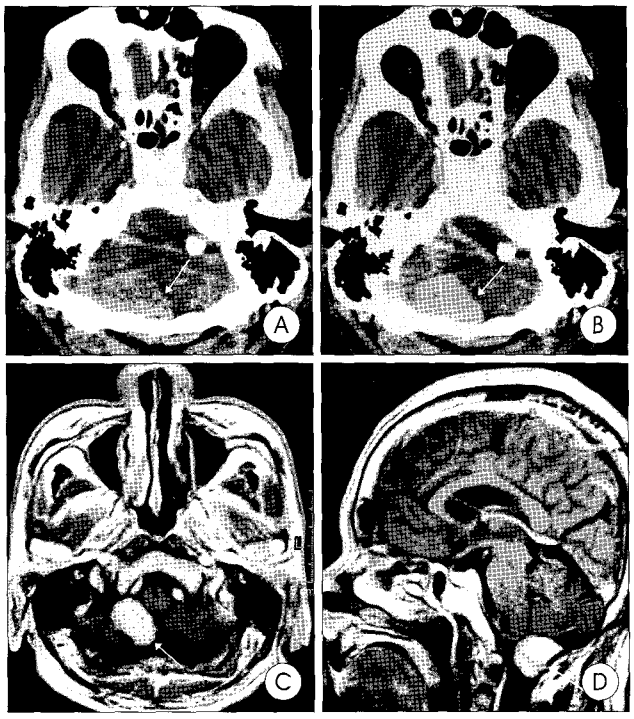

Fig. 1. Preoperative brain computed tomography (A, B) and magnetic resonance image (C, D) reveal a strongly enhancing homogeneous mass (arrow) near the right side of foramen magnum. The most likely preoperative diagnosis, based upon this image, is meningioma.

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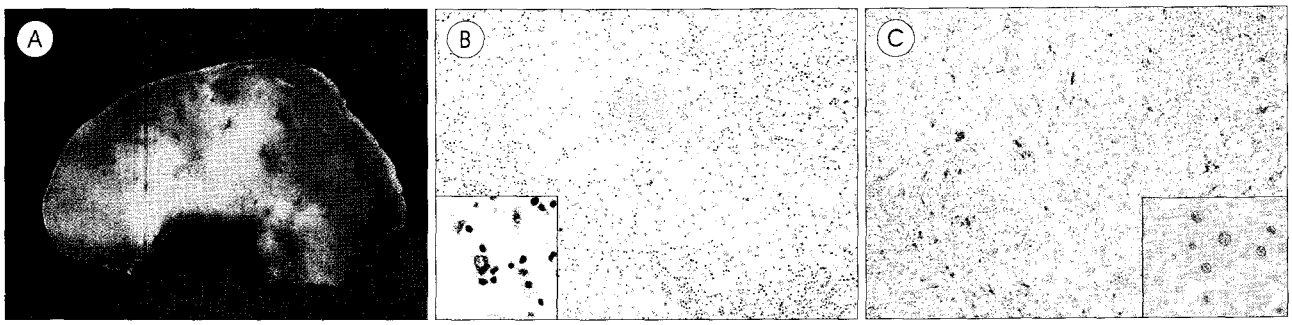

Fig. 2. A : The cut surface of mass shows white to dark yellow and homogeneous appearance with scattered hemorrhage. B : The lesion shows inflammatory infiltrates composed of histiocytes, mature lymphocytes, and plasma cells in the background of fibrosis. (H&E,X 100) Inset : The cytoplasm of the histiocyte contains intracytoplasmic lymphocytes (emperipolesis). (H&E,X 400). C : A positive reaction in the cytoplasm of the histiocytes is shown.(CD68 immunostain, X100). Inset : A strong positive reaction in the cytoplasm of the histiocytes is shown. (S-100 protein immunostain, X 400).

normal. After admission, Magnetic resonance Image(MRI) obtained on the patient revealed a well circumscribed and homogenous enhancing oval shaped mass below the right cerebellum in posterior fossa (Fig. 1C, D).

The patient underwent a suboccipital craniectomy with complete excision of the lesion, which showed 3×2.2×1.3cm sized encapsulated ovoid rubbery mass. The cut surface of the mass showed homogenously pinkish yellow color (Fig. 2A). Microscopically the lesion showed a inflammatory infiltrates composed of mainly histiocytes consisted of foamy cytoplasm and mature lymphocytes and plasma cells (Fig. 2B). The immunohistochemical stain for CD 68, S-100 showed a positive reaction in the cytoplasm of the histiocytes (Fig. 2C).

The postoperative course was eventful. Wound revision and hematoma evacuation was performed due to acute subdural hematoma in previous operation site at 3 days after surgery. And the patient underwent ventriculoperitoneal shunt because of hydrocephalus at 5 months after first surgery.

Discussion

Sinus histiocytosis with massive lymphadenopathy was first described by Rosai and Dorfman in 1969[14]. RDD has become a well-described entity. Since their original description, at least 400 cases have been enrolled in the registry[4]. But extranodal RDD with CNS involvement is a characteristic observed in fewer than 5% of all patients with RDD. The mean age of onset with nodal disease is 20.6 years with a male to female ratio of 1.4:1[4,12]. Patient who present or develop intracranial involvement, however, become symptomatic at a mean age of 34.9 years with a strong male predominance.

About 70% of intracranial cases had no lymphadenopathy and 52% had no associated systemic disease[9]. RDD is known as a sinus histiocytosis with massive lymphadenopathy. However, the usage of this term in CNS is not quite appropriate because of the lack of lymphadenopathy in the great majority of cases.

Imaging studies typically show an enhancing meningeal based mass with a variable amount of edema surrounding the lesion. Although it usually presents as a solitary dural-based lesion, multiple intracranial lesions have also been reported[2,10,14]. Intracranial involvement is more common than spinal involvement[13]. Intracranial involvement will usually present with headache, seizure, or cranial nerve deficit according to the location of the lesion. Hemiparesis, dysphasia, and neglect have also been reported[9].

The etiology of RDD is still obscure. The immunophenotypic profile and studies of monokine expression suggest derivation from activated macrophages that produce interleukin-1β and tumor necrosis factor-α[3].

Microscopic examination typically reveals a polymorphous infiltrate of histiocytes, lymphocytes, and plasma cells in a fibrous stroma. In some cases, eosinophils may also be seen. Two subtypes of histiocytes, differentiated by size, are present in RDD[2,11]. The larger histiocytes typically exhibit emperipolesis. Emperiplesis is an active penetration of one cell by another[6]. In RDD, the larger histiocytes contain well-preserved lymphocytes and are usually S-100 positive[11].

Although the conservative care for intracranial RDD can be a treatment of choice, surgical resection seems to be an appropriate treatment modality. Previously reported patients with intracranial manifestation of RDD have shown a benign course[1,6]. In case with incomplete resection, the resolution of remnant by low dose radiation was reported[1]. With the case of intracranial involvement of RDD, more experiences are required to define the clinical course and prognosis.

In our patients, the differential diagnosis included meningioma, other types of histiocytosis, granulomatous disease, and metastasis. Meningiomas are isolated, dural based neoplasms which are seen as homogeneous lesions, if it does not accompany calcifications. In our case, CT and MRI findings revealed a homogeneous dural based mass isolated in the posterior fossa,

with no associated calcification. We strongly suspected meningioma until histological diagnosis was confirmed.

Conclusion

Isolated intracranial RDD without associated lymphadenopathy, is extremely rare. We report our experience with this disease, and emphasize the difficulty in differentiation prior to surgery.

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