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Posterior fossa intracranial inflammatory pseudotumor: a case report and literature review

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Abstract

Background: Intracranial inflammatory pseudotumors are rare. This study describes an intracranial inflammatory pseudotumor at the left cerebellopontine angle. It is the second documented posterior fossa intracranial inflammatory pseudotumor, and it was treated by surgery and radiotherapy.

Case Description: A 49-year-old man presented with dizziness for 3 months and mild hoarseness for 1 month. Brain CT detected an intracranial tumor at the left cerebellopontine angle. Magnetic resonance imaging revealed a 3.6-cm heterogeneously enhancing mass. Suboccipital craniectomy with ventriculostomy was performed. The mass was well defined with a smooth surface, enclosed the low cranial nerves, and adhered to the dura matter. Pathologic examination revealed fibrous collagenous stroma with dense infiltrates of small lymphocytes and uninucleated histiocytes. Immunopositivity for T-200 and CD-68 was noted. Special staining for mycobacteria and fungus was negative. Serologic tests were positive for EBEA-Ab, EBNA-Ab, and EB-VCA-IgG. An inflammatory pseudotumor was diagnosed. Local recurrence was found 6 months later with a left oculomotor nerve palsy. Whole-brain irradiation with a total dose of 1200 cGy in 6 fractionations was done. Remission was found in follow-up neuroimages, and no recurrence was noted in 2 years' follow-up.

Conclusion: Based on serologic findings and a literature review, the pathogenetic mechanism of this rare intracranial tumor is believed to be chronic reactive EBV infection. We propose that radiotherapy may be the best treatment option in the case of local recurrent intracranial inflammatory pseudotumors.

Keywords: Cerebellopontine angle tumor; Inflammatory pseudotumor

1. Introduction

Histopathologically, inflammatory pseudotumor consists of a collagenous stroma and an inflammatory infiltrate of mononuclear elements [7,14]. Although the pathologic mechanism remains obscure, such a mechanism is believed to result from a nonneoplastic, reactive inflammatory process [1]. Synonymous expressions include fibrous xanthoma, xanthomatous pseudotumor, plasma cell granuloma, plasma cell/histiocytoma complex, pseudosarcomatous fibromixoid tumor, and inflammatory myofibrohistiocytic proliferation

Abbreviations: ANA, antinuclear antibody; CMV, cytomegalovirus; CMV-IgG, cytomegalovirus immunoglobulin G; CMV-IgM, cytomegalovirus immunoglobulin M; CT, computed tomography; EBV, Epstein-Barr virus; EBEA-Ab, Epstein-Barr virus early antigen antibody; EBNA-Ab, Epstein-Barr virus nuclear antigen antibody; EB-VCA-IgG, Epstein-Barr virus capsid antigen immunoglobulin G; EB-VCA-IgM, Epstein-Barr virus capsid antigen immunoglobulin M; GFAP, glial fibrillary acidic protein; HSV, herpes simplex virus; RF, rheumatic factor.

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[4]. Pseudotumors occur most often in the intraorbital cavity, lung, mesentery, omentum, retroperitoneum, genitourinary tract, or upper respiratory tract [3,7,14]. Most intracranial inflammatory pseudotumors arise from the dural and meningeal structures [3,7,14]. A literature review revealed only one previous report of a posterior fossa intracranial inflammatory pseudotumor [12].

We present here an additional case of posterior fossa intracranial inflammatory pseudotumor and discuss the treatment methods.

2. Case description

A 49-year-old male patient with a history of hypertension presented at the neurosurgery ward with dizziness of insidious onset for the past 3 months. Intermittent vertigo, especially when changing body position, was noted. The vertigo was sometimes accompanied by a dull, bitemporal headache. The patient had no evidence of head trauma, fever, or episodes of upper respiration tract infection. Mild hoarseness, mild slurred speech, frequent hiccup, nausea, and unsteady gait were noted during the previous month. Neurologic examination indicated clear and alert consciousness. Neither muscle weakness nor paresthesias was noted. Uvula deviation to the right and a slow gag reflex were noted in the cranial nerve function reviews. Cerebellar examination, including both the finger-nose-finger test and the heel-to-shin test, revealed left side dysmetria. A broad-based, unsteady gait and truncal ataxia were also observed.

Brain magnetic resonance imaging (MRI) revealed an irregular 3.6 × 4.2-cm mass with heterogeneous enhancement over the left anterior cerebellopontine angle and diffuse ventricular dilatation (Fig. 1). The preoperative diagnosis was meningioma. A left suboccipital craniectomy and a right Kocher point ventriculostomy were performed. During surgery, the mass was found to be well defined with a smooth surface, elastic consistency, and was adhered to the convexity of the dura. The 7th and 8th cranial nerves were clearly visible and spared from the tumor. This tumor enclosed the 9th or 10th cranial nerve. The tumor size was markedly reduced by bipolar thermocoagulation, and en bloc resection was easily achieved with gross preservation of the cranial nerves.

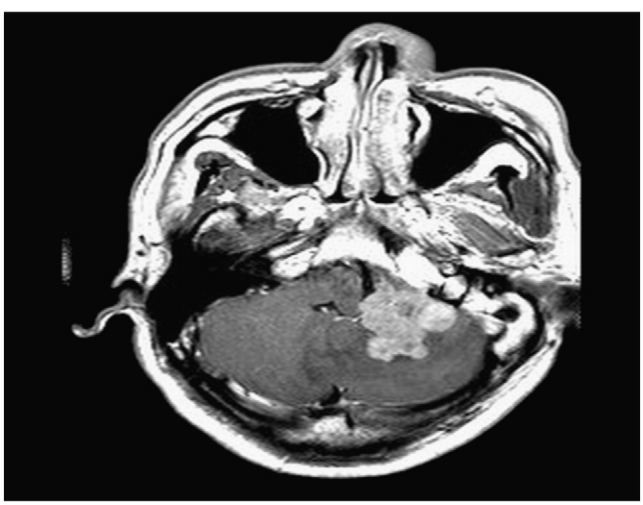

Fig. 1. Radiographic findings. Axial T1-weighted image with contrast enhancement reveals a heterogeneously enhanced lesion at left anterior cerebellopontine angle with lobulation formation.

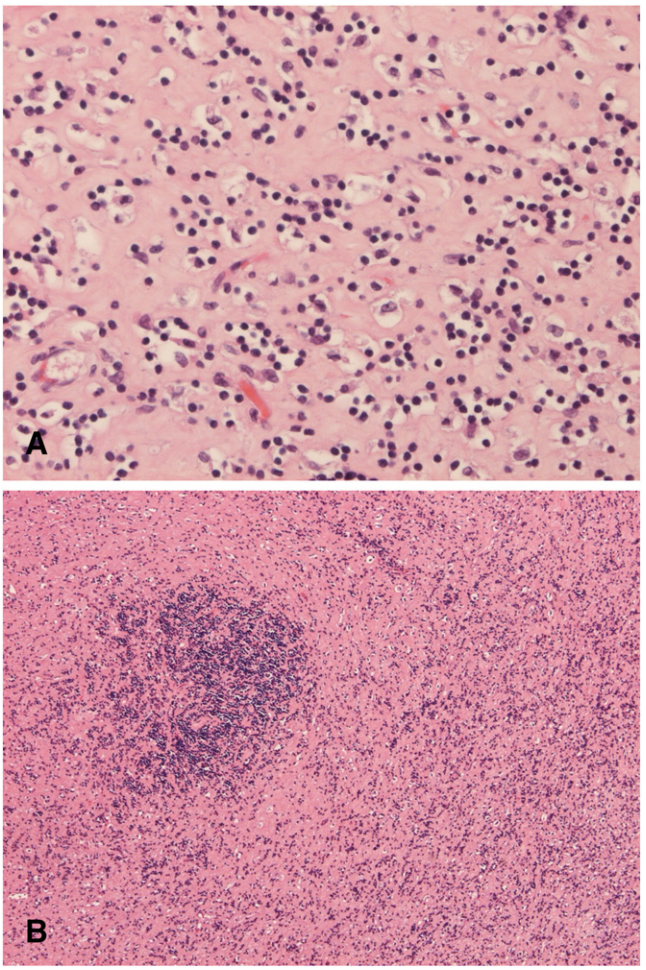

Fig. 2. Microscopic findings. A: Photomicrograph shows infiltrating small lymphocytes and uninucleated histiocytes in the fibrous stroma (hematoxylin and eosin stain). B: Lymphoid follicle with surrounding lymphocytes (hematoxylin and eosin stain).

The pathologic examination showed fibrous, collagenous stroma with dense infiltrates of small lymphocytes and some uninucleated histiocytes (Fig. 2). Immunopositivity for T-200 and CD-68 and immunonegativity for S-100 and GFAP were noted. Special staining for mycobacteria and fungi was negative. Immunochemical staining was negative for both HSV and EBV. However, serologic tests were positive for EBEA-Ab, EBNA-Ab, EB-VCA-IgG, and CMV-IgG but were negative for EB-