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Case Report

Intraventricular Cavernous Malformation Radiologically Mimicking Meningioma

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We report a case of trigonal cavernous malformation (CM) radiologically mimicking meningioma. The computed tomographic (CT) head angiography and magnetic resonance imaging (MRI) showed a partially calcified lesion with slight contrast enhancement located in the area of the left atrium of lateral ventricle. The lesion was completely removed using microsurgery with a parieto-occipital transcortical approach. The resected mass was histologically confirmed as CM. CM should be considered as differential diagnosis in case of the atrial mass lesion due to lack of hemosiderin ring characteristically seen other seated CM.

KEY WORDS : Cavernous malformation · Meningioma · Trigone · Atrium.

INTRODUCTION

Intraventricular cavernous malformations (CM) are very rare vascular malformations that account for 2.5% of all brain cavernous malformations[15]. Finkelnburg first described intraventricular CM in 1905. Since then, less than 50 cases have been reported[3,8,11,14]. On imaging, these lesions may be misdiagnosed as arteriovenous malformations (AVM), meningiomas, or high grade gliomas.

We report a clinical case of trigonal CM which was diagnosed initially as a trigonal meningioma; the results of neuroimaging were more compatible with trigonal meningioma than with trigonal cavernous malformation.

CASE REPORT

A 48-year-old woman presented with sudden-onset severe headache for the first time. Brain computed tomography (CT) and brain magnetic resonance images (MRI) at another hospital revealed a 2.5 cm of maximal diameter, well-defined, partly calcified round mass in the left trigonal region with slight contrast enhancement. There was no perileional edema or midline shift. There was also no hydrocephalus (Fig. 1A, B). Severe headache was gradually decreased and resolved. The physical examination demonstrated right homonymous temporal hemianopia. CT head angiography at the time of admission showed that the lesion was not definitely supplied by feeding arteries (Fig. 1C). The lesion, atypical features of meningioma in radiological images, was diagnosed initially as an intraventricular tigonal meningioma based on incidence and epidemiological factors. A differential diagnosis of choroid plexus papilloma was considered less likely. She underwent left parieto-occipital craniotomy, inferior parietal gyrus approach, and en block excision of the mass. Inferior parietal gyrus corticectomy and track were created to the shortest distance of the lesion under frameless navigation guidance. There was a multi-lobulated purple mass within the atrium extending anteriorly into the temporal horn. The gross appearance of the resected mass, purple colored multiple lobulated, was thought to be more compatible with CM rather than Meningioma (Fig. 2). Postoepratively, the visual field deficit, right upper homonymous hemianopsia, remained to be unchanged. The permanent pathology was reported not as meningioma but as cavernous malformation unlike our intial preoperative radiological diagnosis. Postoperative CT confirmed the total resection of the CM (Fig. 3). We reviewed the preoperative radiological images of the patient with trigonal CM postoperatively. Gradient echo images which were neglected preoperatively showed multiple low signal intensity suggesting thrombosis or hemorrhage (Fig. 1D).

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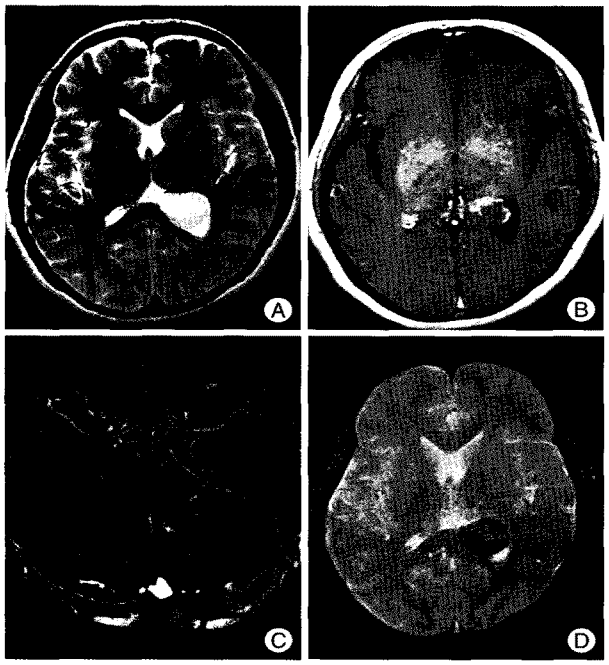

Fig. 1. Preoperative Brain magnetic resonance (MR) image and computed tomography (CT) head angiography. A : T2-weighted image showing a mass at the left trigonal region without hydrocephalus or edema. B : Preoperative gadolinum-enhanced T1-weighted MR image demonstrating a scanty enhanced lesion. C : Preoperative CT-head angiography revealing no definite feeding arteries. D : preoperative gradient echo MR image showing intratumoral hemorrhage or thrombosis.

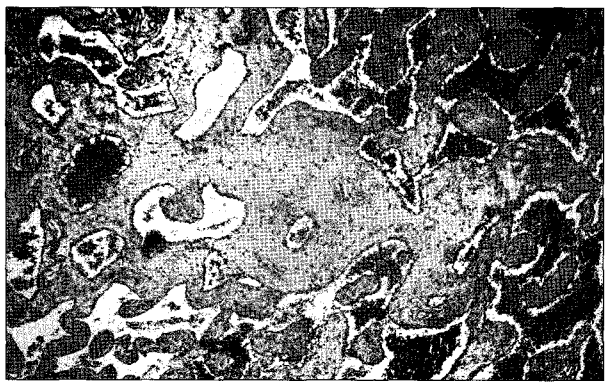

Fig. 2. Photomicrograph of the cavernous malformation resected. The mass consists of multiple large, dilatated and hyalinized vessels, lined by flattened endothelia.

DISCUSSION

CM is a type of vascular malformation that are composed of tightly packed thin-walled vessels of various sizes, lined by a single layer of endothelial cells and collagen fibers. There is no intervening nervous tissue within the malformations.

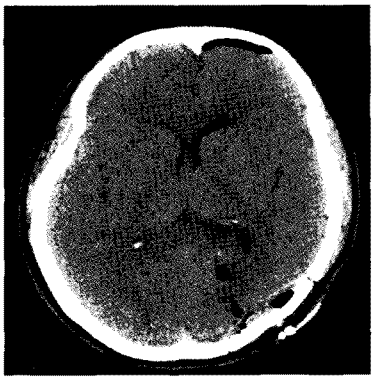

Fig. 3. Postoperative Brain computed tomography demonstrating the route approaching the left trigone and confirming complete removal of the tumor.

The incidence of CM has been reported in the range of 0.4-0.5% in MRI and autopsy series[2,12]. CM is typically located in the subcortical areas, deep white matter and basal ganglia. Supratentorial areas account for 80% of locations where cavernous malformation occur. In the infratentorial regions, cavernous hemangiomas mostly occur in the brainstem and cerebellum[5]. CM rarely occurs within the ventricular system. About 50 well-documented cases have been published[3,8,11,14].

Trigonal cavernous malformation may be misdiagnosed as malignant neoplasms, meningiomas, and AVMs because of their enhancement and heterogeneous appearance of CT and MRI. The intraventricular location, size, hyper intensity, and partial calcification suggest a neoplastic lesion. However, the lack of surrounding edema on the fluid-attenuated inversion recovery and T2 MRI sequences, the peripheral hemosiderin seen on the T2-weighted axial MRI scan, and the lack of enhancement make a tumor less likely and suggest vascular malformation[1]. Typical CT findings of CM usually demonstrate a well circumscribed moderately hyper-intense nodular lesion with mild contrast enhancement and calcifications[4,15]. The characteristic appearance on MRI is a mixed signal intensity resulting from blood products of varying ages. However, Kumer et al.[6] have described intraventricular cavernous angiomas lack of the hemosiderin ring. In retrospective investigation of our case, slight hyper intensity on T2-weighted images and hyper intense on T1-weighted images in radiologic features of our case may be more closely correlated with subacute stage hemorrhage rather than pure mass lesion. Meningiomas generally are hypo- to isointense on T1-weighted images and iso- to hyperintense on T2-weighted images relative to cortex[7,10]. Homogenous signal intensity with nodular mass lesion may suggest a first episode of hemorrhage within mass lesion without repetitive bleeding. Despite a first attack of hemorrhage, well-circumscribed intraventricular mass lesion should be differentially diagnosed as cavernous malformation; we simply overlooked this lesion as trigonal meningioma on the base of incidence

without more careful consideration. We reviewed the preoperative radiological images of the patient with trigonal CM postoperatively. Preoperative Brain MRI and Brain CT suggested that the lesion was hemorrhagic with scanty enhancement and well circumscribed tumor. If we had examined the preoperative radiological images carefully, trigonal CM would have been included in the differential diagnoses of the lesion.

The natural history of CM is not fully understood. The estimated annual risk of hemorrhage of supratentorial CM is about in the range of 0.25-0.7%[9]. The natural history of intraventricular CM is not known due to rare incidence. Reins et al.[11], in a review of 27 cases of intraventricular CM, found that 64% of patients presented with mass effect and 20% with acute hemorrhage. The tendency of rapid growth and extralesional hemorrhage of intraventricular cavernomas may suggest the need to treat these lesions more aggressively[11,13].

Only a few reports suggested the effectiveness of radiotherapy in the treatment of CM. Proliferation and recurrence have been described in several cases treated conservatively or with radiotherapy alone[4,9]. Complete surgical resection should be the gold standard of treatment in case of intraventricular CM[15]. Using any surgical approach, callostomy or corticectomy of normal brain territory is necessary to reach a trigonal tumor in the lateral ventricle. The surgical approach to trigonal lesions depends on the size of the lesion and whether the lesion is in the dominant hemisphere or not. A various surgical approaches to the trigone have been described in past reports. Trigonal CM of our case was resected by this paramedian parieto-occipital transcortical approach. No additional neurologic sequela with the preexisting visual field disturbance developed postoperatively.

The radiologic diagnosis of intraventricular cavernoma may be difficult as these lesions may mimic neoplasm such as germinoma, astrocytomas, or meningiomas. Incorrect preoperative diagnosis has sometimes resulted in inappropriate therapy, including radiotherapy[11].

CONCLUSION

Although trigonal CM with unusual radiologic features is extremely rare, they should be considered in the differential diagnosis of trigonal lesions. Because the treatment plan of intraventricular CM, complete removal of the lesion should be gold standard of treatment with no other treatment modalities, are somewhat different from other intraventricular pathologies.

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