CASE REPORT

GMJ.2017;6(1):61-65
www.gmj.ir

Received 2016-07-26
Revised 2016-10-17
Accepted 2016-11-02

An Occipital Headache as the First Presentation of Multiple Third, Fourth, and Lateral Ventricular Cavernous Malformations: A Case Report and Review of Literature

Ali Reza Vakilian [1], Amir Moghadam Ahmadi [2], Habib Farahmand [3] ✉

[1] Department of Neurology and Geriatric care research center, School of Medicine, Ali-Ebn Abitaleb Hospital, Rafsanjan University of Medical Sciences, Rafsanjan, Iran
[2] Department of Neurology, School of Medicine, Ali-Ebn Abitaleb Hospital, Rafsanjan University of Medical Sciences, Rafsanjan, Iran
[3] Department of Radiology, School of Medicine, Ali-Ebn Abitaleb Hospital, Rafsanjan University of Medical Sciences, Rafsanjan, Iran

Abstract

Background: Cavernous hemangiomas are common benign vascular malformations. Their existence in the intraventricular region is very rare. **Case Reports:** A 43-year old woman with an occipital headache was admitted to the emergency ward. Brain computed tomography scan showed mild hydrocephalus and multiple intraventricular isodense lesions. Imaging findings, especially of Gradient Resonance Echo imaging, were in favor of multiple intraventricular cavernous malformations. **Conclusion**: This is a rare presentation of multiple cavernous malformation as occipital headache without needing surgical intervention in this phase. Coexistence of periventricular plaques like Radiologically isolated syndrome of Multiple sclerosis is another unique aspect in this report. **[GMJ.2017;6(1):61-65]**

Keywords: Multiple; Cavernous Malformations; Ventricular Hemangiomas

Introduction

Cavernous malformation (CM) or cavernoma of the central nervous system are vascular malformations that usually present in the cerebral hemispheres. Cavernous hemangiomas are common benign vascular malformations and are ubiquitously distributed in the central nervous system. It constitutes 5-10% of vascular malformations occurring in the central nervous system and commonly seen in the 2nd-5th decades of life [1].
However, their existence in the intraventricular region is very rare, constituting 2.5-10% of all cerebral CM [1, 2].
In total, 17 trigonal CMs were reported, and their clinical presentation included mass lesion, hemorrhage, and seizure [1].
There are more than 200 cases of CMs in literature, but this study's case is unique because it considered the multiplicity of intraventricular CMs. Due to the rarity of these lesions in the intraventricular region and multiplicity of them in various ventricles and difficulty of diagnosis without special magnetic resonance imaging (MRI) techniques, we reported this case for educational objectives.

GMJ

Tel/Fax: +98 71 36474503
PO Box 7193616563
Email:info@gmj.ir

✉ **Correspondence to:**
Habib Farahmand, Department of Radiology, Ali-Ebn Abitaleb Hospital, Rafsanjan University of Medical Sciences, Rafsanjan, Iran
Telephone Number: +983434280185
Email Address : crcdc2research@gmail.com

Case Presentation

A 43-year old woman with an occipital headache was admitted to the emergency ward of Ali-ibn-Abitaleb Hospital in Rafsanjan city. The onset of symptom was acute, and the quality of pain was pulsatile with nausea. No fever or blurred vision was detected.

There was a history of mild periodic headache in recent years. Patient's medical history showed hypertension, diabetes and periodically used an oral contraceptive. At the time of admission, her blood pressure was 140/75 mmHg, respiratory rate was 15/min, pulse rate was 88/min, and no papilledema was found on the fundoscopy. The level of consciousness and its quality were normal. Examinations of cranial nerves and other neurologic examinations were normal.

In laboratory examination complete blood count (CBC) results were: red blood cells (RBC) 3.95 million/mc liter, white blood cells (WBC) 9700 cells/ml, hemoglobin (Hb) 13.1 gram/dl, hematocrit (Hct) 36.5%, mean corpuscular volume (MCV) 92.4 fL/cell, mean corpuscular hemoglobin (MCH)33.2 picograms (pg)/cell, Mean corpuscular hemoglobin concentration (MCHC) 35.9 gr/dl, platelet 251000 /mcL. Blood urea nitrogen (BUN) was 25mg/dL, creatinine was 0.8, fasting blood sugar (FBS) 148, Sodium 144 mEq/L, Potassium 4.5 mEq/L, serum glutamic-oxaloacetic transaminase (SGOT) 16, serum glutamic-pyruvic transaminase (SGPT) 32, total bilirubin was 0.1, direct bilirubin 0.5. Erythrocyte sedimentation rate was 13. The both c-reactive protein (CRP) and rheumatic factor were negative.

Brain computed tomography (CT) scan showed mild hydrocephalus and multiple intraventricular isodense lesions (Figure-1). Brain MRI revealed multiple T1-T2 weighted isointense nodular lesions in the 3rd and 4th ventricles and frontal horns of lateral ventricles. The lesions were mildly hyperintense in the FLAIR sequences (Figure-2). Contrast-enhanced MRI showed no enhancement of the lesions (Figure-3).

For more evaluation, lumbar puncture was done which revealed lymphocytic pleocytosis with normal protein and glucose (aseptic meningitis).

In the cerebrospinal fluid (CSF) study, there was bloody appearance, 600 RBC, 100 WBC [85% lymphocyte), protein 23 mg/dL, glucose 160 mg/dL and lactate dehydrogenase was 17 IU/L. Every intraventricular lesion showed significant blooming at T2* gradient resonance echo (GRE) and susceptibility weighted imaging (SWI) sequences (Figure-4).

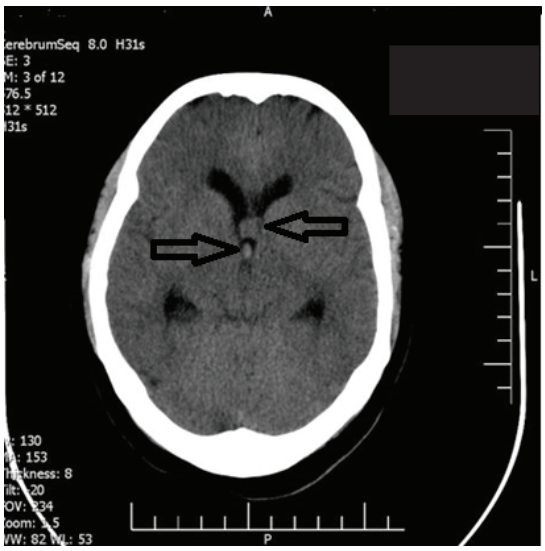

Figure 1. Non-contrast CT scan revealed nodular lesions (blank arrows) within the 3rd ventricle and frontal horns of lateral ventricles. Mild hydrocephalus is seen.

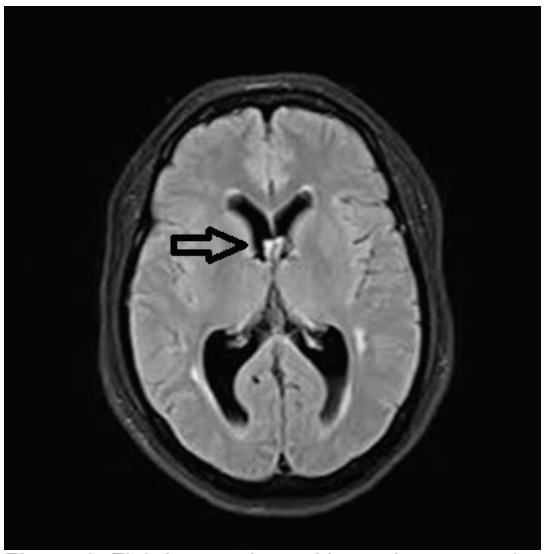

Figure 2. Flair Image showed hyper intense nodular lesions (blank arrow) within the frontal horns of lateral ventricles.

Brain magnetic resonance angiogram (MRA) (Figure-5) and magnetic resonance venography (MRV) were normal. Imaging findings, especially of GRE images, were in favor of multiple intraventricular CMs. Incidentally, multiple T2-FLAIR hyperintense lesions were seen in the white matter of cerebral hemispheres including deep periventricular and juxtacortical regions without enhancement in the post-contrast images. Although these findings suggest multiple sclerosis (MS), no clinical sign and symptom in favor of MS were present. Thus, it is believed that these later findings were due to a radiologically isolated syndrome (RIS).

Our patient had no need for surgery in this phase because the hydrocephalus was not severe and there was no hemorrhage during our followed up. No deterioration was seen in the patients, five months after discharging from the hospital.

Discussion

Prior to this study, there was no case of multiple intraventricular CMs in literature. So far, to the best of our knowledge, more than 200 cases of intraventricular CMs have been reported. Those located in the third ventricle account for approximately half of all intraventricular CMs. Only 14 cases of cavernous hemangiomas at the foramen of Monro have been reported previously [3].

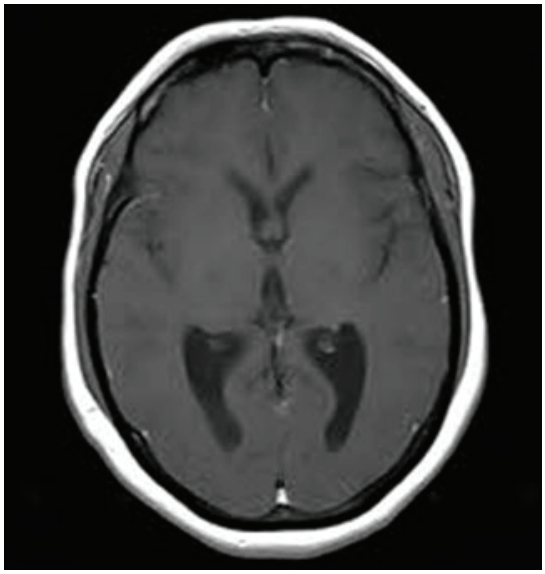

Figure 3. Contrast enhanced T1 weighted images revealed no enhancement in the intraventricular lesions.

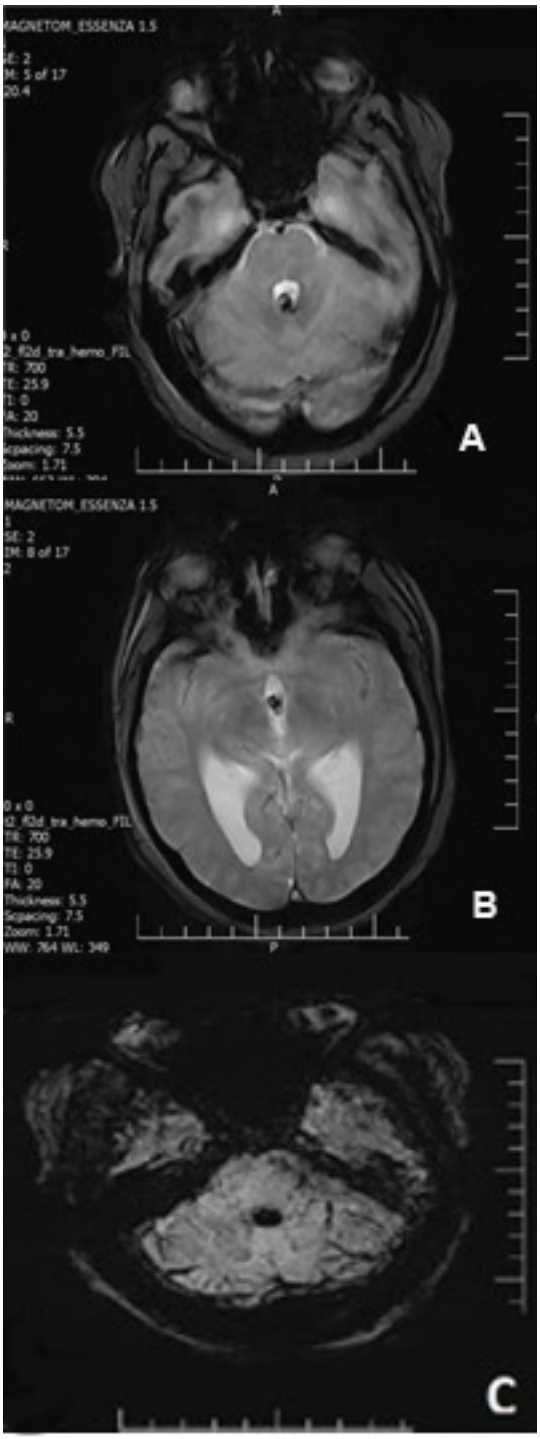

Figure-4: T2*GR images (A and B) and SWI (C) revealed significant blooming and susceptibility effect in the intraventricular lesions.

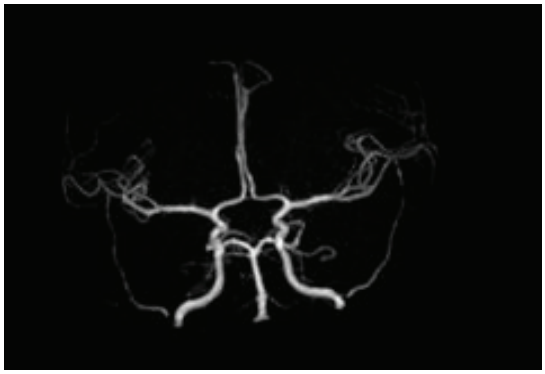

Figure 5. The MRA findings demonstrated no abnormality.

In particular, at the foramen of Monro, cavernoma was reported in about 14 cases as very rare and may be difficult to differentiate from other brain tumors [3].

However, CMs occur very rarely in the ventricles. A rarer case was in a patient with CM involving both the third and lateral ventricles [4]. The present case report describes a case of multiple intraventricular masses with mild hydrocephalus which turned out to be multiple CMs in the third, fourth and frontal horns of the lateral ventricles.

In the first few days we suspected cerebral venous thrombosis (CVT) and chronic meningitis, but normal cytology and CSF smear with normal glucose and the detection of protein in the CSF changed the condition. Another finding of this study is the occurrence of lymphocytosis in CSF. There was no xanthochromia or hemorrhage in the CSF study or siderosis in the MRI images, as we expected to find according to many reports of intraventricular hemorrhages and subarachnoid hemorrhage in multiple intracranial CMs [2].

Normal MRV and MRA reduced the risk of CVT and another vascular basis for this event. Typical RIS for MS was another finding in this patient without clinical presentation of MS. According to our MRI findings, these vascular abnormalities were considered as CMs. Differential diagnosis of these malformations are choroid plexus papilloma, ependymoma, teratoma, neurocytoma, metastasis, meningioma, astrocytoma, and arteriovenous malformation [5]. Germinoma, colloid cyst, and subependymal giant cell astrocytoma were also considered as the other differential diagnoses [3]. Our patient was not chosen for surgery because his condition was stable and her hydrocephalus was mild. Hence there was no access to any pathological sample. Based on the MRI findings, other diagnoses were ruled out. Imaging findings especially GRE and SWI images were in favor of multiple intraventricular CMs compatible with type III Zabramski classification [6]. Though the differential diagnosis of multiple intraventricular lesions are diverse, lack of enhancement in the post-contrast images and prominent susceptibility effect in the SWI and T2*GRE sequences were against a lot of diagnostic possibilities such as primary intraventricular neoplasms or the earlier described metastatic diseases. Indeed, our patient had no other neoplasm in her body. This study included SWI for better diagnosis that could represent type 5 of these malformations that were not shown on the T2-weighted GRE [7].

In lateral ventricular CM (LVCM), surgery has a role in treatment. The LVCMs can usually be easily dissected from the ventricular walls. In lateral ventricles, the best approach for surgery is from the frontal horn. Hydrocephalus is a rare complication and shunt is advised before CM surgery. Surgery is needed if the mass enlarges significantly [8]. Our case lesions in lateral ventricle was found incidentally, and their size were not large. Complete surgical resection should be the gold standard of treatment in case of intraventricular CM [8].

The surgical approach to trigonal lesions depends on the size of the lesion and whether the lesion is in the dominant hemisphere or not [9]. However, CMs located in the third ventricle, surrounded by vital structures, are especially dangerous. These lesions show rapid growth, resulting in significant morbidity. The most frequent postoperative complication was a hydrocephalus, large-sized lesions frequently involve the hypothalamus [10].

Conclusion

Our case was a rare one as an occipital headache with multiple intraventricular CM. Cur-

rently, management of asymptomatic CM is not by surgery as our case, and conservative management is recommended. Surgery is recommended if the lesion is safely accessible, is symptomatic either by mass effect and/or hemorrhage or seizure or shows evidence of having bled in the past. Microsurgical removal of the intraventricular lesions is safe, but in the fourth ventricle, as our case evolved it posed increased risk for the cranial nerve deficits.

Conflict of Interest

No conflict of interest was provided for this paper.

References

1. Patibandla MR, Thotakura AK, Panigrahi MK. Third ventricular cavernous malformation: an unusual lesion. Br J Neurosurg.. 2014;28(1):110-2.
2. Ohbuchi H, Osaka Y, Ogawa T, Nanto M, Nakahara Y, Katsura K, et al. Trigonal cavernous malformation with intraventricular hemorrhage: A case report and literature review. J Med Invest. 2012;59(3.4):275-9.
3. Lee B-J, Choi C-Y, Lee C-H. Intraventricular cavernous hemangiomas located at the foramen of monro. J Korean Neurosurg Soc. 2012;52(2):144-7.
4. Zakaria M, Abdullah J, George J, Mutum S, Lee N. Third ventricular cavernous angioma. Med J Malaysia. 2006;61(2):229.
5. Stavrinou L, Stranjalis G, Flaskas T, Sakas D. Trigonal cavernous angioma: a short illustrated review. Acta Neurochir (Wien). 2009;151(11):1517-20.
6. Zabramski JM, Wascher TM, Spetzler RF, Johnson B, Golfinos J, Drayer BP, et al. The natural history of familial cavernous malformations: results of an ongoing study. J Neurosurg. 1994;80(3):422-32.
7. Bulut HT, Sarica MA, Baykan AH. The value of susceptibility weighted magnetic resonance imaging in evaluation of patients with familial cerebral cavernous angioma. Int J Clin Exp Med. 2014;7(12):5296.
8. Jin S-C, Ahn J-S, Kwun B-D, Kwon DH. Intraventricular cavernous malformation radiologically mimicking meningioma. J Korean Neurosurg Soc. 2008;44(5):345-7.
9. Wang J, Gong X. Superficial siderosis of the CNS associated with multiple cerebral cavernous malformation. Neurology. 2009;72(13):1187.
10. Han M-S, Moon K-S, Lee K-H, Kim S-K, Jung S. Cavernous hemangioma of the third ventricle: a case report and review of the literature. World J Surg Oncol. 2014;12(1):237.