Case Report

Drainage Vein Induced Hydrocephalus Caused by an Unruptured Arteriovenous Malformation in an Adult Presenting with Visual Loss

Yasar BAYRI[1, 2], Mustafa SAKAR[1, 2], Ali OZEN[1, 2], Mustafa Ibrahim ZIYAL[1, 2]

[1]Marmara University, School of Medicine, Department of Neurosurgery, İstanbul, Turkey
[2]Marmara University, Institute of Neurological Sciences, İstanbul, Turkey

ABSTRACT

Arteriovenous malformations (AVMs) commonly present with seizures and hemorrhage. Hydrocephalus associated with an unruptured AVM in an adult patient is exceedingly rare. A 37-year-old male patient presented with total visual loss in his right eye and severe impairment in his left eye. His evaluation showed an unruptured right frontal AVM and bilaterally dilated lateral ventricles. The major draining vein of the AVM was obstructing the third ventricle. An urgent external ventricular drainage was used as the first line intervention and followed with a ventriculoperitoneal shunt two days later. His definitive treatment for AVM was staged stereotactic gamma-knife radiosurgery. Unruptured AVMs in adult patients may rarely cause hydrocephalus. Visual loss caused by such a hydrocephalus has not been reported before. Both communicating and non-communicating type hydrocephalus can be seen with unruptured AVMs, and have different pathophysiological mechanisms. Our patient was treated with ventriculoperitoneal shunting, and visual examination remained unchanged after the operation. Visual loss caused by hydrocephalus associated with an unruptured AVM in an adult patient has not been reported before. It may indicate an association of different pathophysiological mechanisms. The treatment must depend on the neurological condition of the patient.

KEYWORDS: Adult, Arteriovenous malformation, Hydrocephalus, Unruptured, Visual loss

ABBREVIATIONS: AVM: Arteriovenous malformation, **AVMs:** Arteriovenous malformations, **CSF:** Cerebrospinal fluid, **CT:** Computed tomography, **MRI:** Magnetic resonance imaging.

■ INTRODUCTION

True arteriovenous malformations (AVMs) are classified as arteriovascular malformations of brain, along with capillary telangiectasias, cavernous malformations and developmental venous anomalies (11). Symptom presentation is mostly at the third and fourth decades and includes hemorrhage, seizures, headaches, focal neurological deficits etc. Hydrocephalus is a very rare entity in the adult population as the presenting symptom of an unruptured AVM. There are 14 reported adult patients having hydrocephalus associated with unruptured AVMs in the literature (3-7,9,10,12,13). Here we present an adult patient, complaining of visual loss resulting from hydrocephalus induced by the major draining vein of an unruptured AVM, which has not been reported previously.

■ CASE REPORT

A 37 year-old-male patient presented with total blindness on his right eye and some visual impairment on his left eye. His right pupilla was not reactive to light and he had bilateral papilledema. His Glasgow Coma Scale was 15/15 and rest of his neurological examination was normal. He reported a headache episode 5 months ago which subsided spontaneously and he was normal except a slight gait disturbance for the last 4 months. One month ago his vision

Corresponding author: Mustafa SAKAR

E-mail: docmsakar@gmail.com, mustafa.sakar@marmara.edu.tr

started to blur. He lost vision on his right eye progressively for the last week. His vision on his right eye continued to deteriorate. Admission cranial computed tomography (CT) of the patient showed a right frontal AVM, and bilaterally dilated lateral ventricles but not the third and fourth. There was no bleeding. Elevated intracranial pressure was evident on cranial CT by hypodensity of brain tissue surrounding the lateral ventricles reflecting the cerebrospinal fluid (CSF) reflux (Figure 1A-H). An urgent ventricular drainage catheter was introduced as the first line intervention. The biochemical analyses of the CSF obtained during external ventricular drainage (EVD) was normal with glucose 68mg/dl and protein 43mg/dl. Cytology and cultures were also found to be negative later. MRI showed the nidus was located at the right frontal lobe. A tortious, venous signal void was seen at the medial side of the nidus, travelling through the level of the third ventricle. This vein seemed to obstruct the third ventricle and both foramina of Monro and caused bilaterally dilated lateral ventricles (Figure 1A-H). Conventional angiography showed a right frontal high-flow AVM, which had the feeders mainly from the right middle cerebral artery. Venous drainage was mainly to the deep venous system through the highly dilated and tortuous thalamostriate and internal cerebral veins, finally reaching the vein of Galen and straight sinus (Figure 2A-C). The diagnosis was a Spetzler-Martin Grade 3 right frontal AVM. A shunt procedure was performed after two days of drainage, and the lateral ventricles showed a significant reduction in size (Figure 1A-H). The patient was scheduled for staged stereotactic gamma-knife radiosurgery for his AVM immediately after the shunt procedure. One week later, his right pupilla was still non-reactive to light. However his vision on his left eye was at least stable, if not better.

■ DISCUSSION

Two of the 14 patients had communicating hydrocephalus where an unruptured AVM was documented as the cause. Geibprasert et al and Ebinu et al coincidently emphasized venous outflow obstruction and the potential role of a hemodynamic disequilibrium (4,6). Although explaining hydrocephalus in this circumstance by venous hypertension and malabsorption of the CSF from arachnoid villi seems reasonable, the high flow shunt may also cause a venous hypertension and hinder CSF absorption through trans ependymal flow. This is a previously described mechanism in pediatric population with vein of Galen AVMs and high flow pial shunts (1,14). However, the exact mechanism through which the elevated venous hypertension results in communicating hydrocephalus is not apparent enough.

On the other hand, it is obvious that non-communicating hydrocephalus is a result of compression on CSF pathways. The reason for blockage of the CSF pathways can be a draining vein, a venous varix, or the nidus itself. This late onset blockage of CSF pathways may have a role in the dynamic nature and growing tendency of some of these congenital malformations as stated before (15).
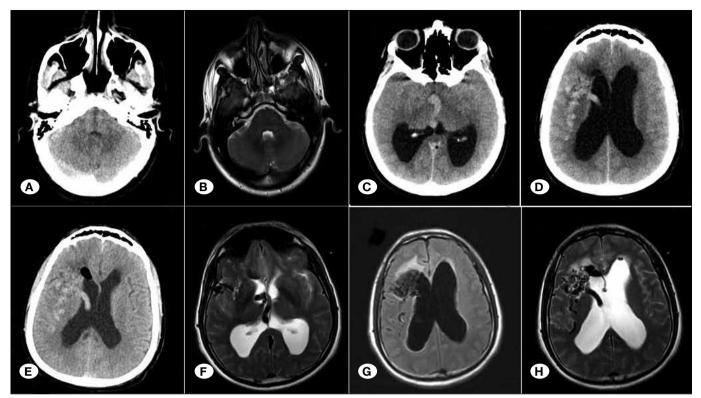
Other 12 patients had non-communicating hydrocephalus caused by an unruptured AVM (3,5-7,9,10,12,13). It seems that

there is tendency for the posterior fossa lesions to obstruct the Sylvian aqueduct and supratentorial lesions to obstruct third ventricle, which is reasonable. The level of obstruction may play an important role in decision making when managing hydrocephalus. In their well-documented patient series, Geibprasert et al. reported an unusually high percentage of shunt problems (6). Esparza et al. also reported a patient with AVM causing hydrocephalus in which a shunting procedure resulted in stupor and Parinaud's Syndrome suggesting an upward herniation (5). Treating a non-communicating hydrocephalus with third ventriculostomy is a well-established procedure and has many advantages over shunting. Kehler stated modern hydrocephalus treatment modalities that can be the choice of intervention for these cases, in his comment (8). These modalities include endoscopic third ventriculostomy for aqueductal stenosis and endoscopic septum pellucidotomy for unilateral blockage of the foramen of Monro. Although we agree with the author, in the setting of a neurosurgical emergency such as visual loss that we experience in our patient, a direct CSF diversion method such as ventricular drainage or shunting seems a more logical and safe option. Also, since the huge major draining vein was near totally obstructing the third ventricle, endoscopic third ventriculostomy seemed dangerous and troublesome, if not impossible. We chose a ventricular drainage as the first line treatment in this neurosurgical emergency setting, and replaced it with a ventriculoperitoneal shunt two days later. The patient was discharged without having a shunt problem.

Fourteen patients reported in the literature had both communicating and non-communicating hydrocephalus associated with an unruptured AVM (3-7,9,10,12,13). Most of these patients presented with headache or with signs of chronic obstructive hydrocephalus, but none of them had visual impairment of any degree. Visual impairment and papilledema are also reported with unruptured AVMs, although rare (2). Most of these patients present with a pseudotumor cerebri like clinical appearance but without hydrocephalus. In light of above mentioned reports, explanations and theories, our patient can be a unique example that both mechanisms took action together. While increased intracranial pressure leaded to headache episode, a slight change in the configuration of vascular anatomy of the malformation in a later stage caused obstruction of the third ventricle and resulted in non-communicating hydrocephalus. Increased intracranial pressure and non-communicating hydrocephalus together might result in visual disturbances.

■ CONCLUSION

Diseases related to CSF dynamics such as hydrocephalus and pseudotumor cerebri can present with visual symptoms. Most patients show a recovery of visual functions after a succesful intervention. Recovery of functions is a reliable measure that shows the effective treatment of the situation, also this is not the rule. In most situations, stabilizing the progressive neurological deterioration is also regarded as a sign of effective treatment. Late recovery of neurological functions can be seen in some patients both in visual symptoms and other neurological deficits. Our patient's progressive visual loss stabi-

Figure 1: A, B) Admission CT and MRI showing the size of the fourth ventricle. **C)** Admission CT showing the drainage vein obstructing third ventricle. **D)** Admission CT showing marked ventricular enlargement of lateral ventricles. **E)** Significant reduction in size of both lateral ventricles after external ventricular drainage and shunting. **F)** Axial T2 MRI image showing drainage vein obstructing third ventricle. **G, H)** T2 axial Flair and T2 axial MRI images showing the nidus and slight ependymal reflux.
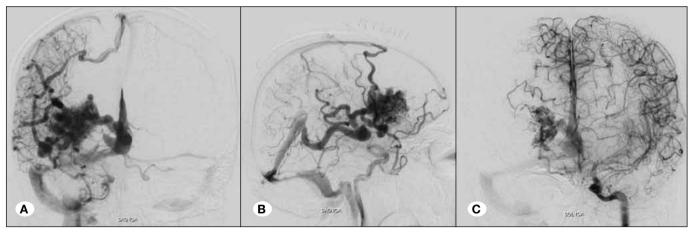

Figure 2: Conventional angiography showing highly dilated and tortious thalamostriate and internal cerebral veins finally draining to vein of Galen and straight sinus. Other smaller cortical draining veins to the superior sagittal sinus and right transverse-sigmoid sinuses are seen.

lized after EVD and shunt replacement. There was a failure of recovery after the first week but there was also a stabilization of ongoing neurological deterioration. We also believe that there is still a chance for late recovery.

■ **REFERENCES**

1. Alvarez H, Garcia Monaco R, Rodesch G, Sachet M, Krings T, Lasjaunias P: Vein of galen aneurysmal malformations. Neuroimaging Clin N Am 17:189-206, 2007

2. David CA, Peerlerss SJ: Pseudotumor syndrome resulting from a cerebral arteriovenous malformation: Case report. Neurosurgery 36:588-590, 1995

3. DeFoe DR, Kusske JA, Rush JL, Pribram H: Aquaductal occlusion by midline arteriovenous malformation. Surg Neurol 5: 59-62, 1976

4. Ebinu JO, Matouk CC, Wallace MC, Terbrugge KG, Krings T: Hydrocephalus secondary to hydrodynamic disequilibrium in an adult patient with a choroidal-type arteriovenous malformation. Interv Neuroradiol 17:212-216, 2011

5. Esparza J, Lobato RD, Munoz MJ, Shillon D, Portillo JM, Lamas E: Giant cerebral arteriovenous malformation producing a noncommunicating hydrocephalus. Surg Neurol 15:76-80, 1981

6. Geibprasert S, Pereira V, Krings T, Jiarajongmun P, Lasjaunias P, Pongpech S: Hydrocephalus in unruptured brain arteriovenous malformations: Pathomechanical considerations, therapeutic implications, and clinical course. J Neurosurg 110:500-507, 2009

7. Hoi S, Kerber C: Ventricular obstruction secondary to vascular malformations. Neurosurgery 12:572-575, 1983

8. Kehler U: Hydrocephalus in unruptured brain arteriovenous malformations. J Neurosurg 112: 696, 2010

9. Liu HM, Wang YH, Chen YF, Tu YK, Huang KM: Endovascular treatment of brainstem arteriovenous malformations: Safety and efficacy. Neuroradiology 45: 644-649, 2003

10. Lobato RD, Lamas E, Cordobes F, Munoz MJ, Roger R: Chronic adult hydrocephalus due to uncommon causes. Acta Neurochir (Wien) 55:85–97, 1980

11. McCormick WF: The pathology of vascular ("arteriovenous") malformations. J Neurosurg 24:807-816, 1966

12. Mindea SA, Yang BP, Batjer HH: Unruptured arteriovenous malformation in a patient presenting with obstructive hydro-cephalus. Case report and review of the literature. Neurosurg Focus 22: E11, 2007

13. Pribil S, Boone SC, Waley R: Obstructive hydrocephalus at the anterior third ventricle caused by dilated veins from an arteriovenous malformation. Surg Neurol 20:487-492, 1983

14. Rodesch G, Malherbe V, Alvarez H, Zerah M, Devictor D, Lasjaunias P: Nongalenic cerebral arteriovenous malformations in neonates and infants. Review of 26 consecutive cases (1982-1992). Childs Nerv Syst 11: 231-241, 1995

15. Spetzler RF, Wilson CB: Enlargement of an arteriovenous malformation documented by angiography. Case Report. J Neurosurg 43:767-769, 1975