Case Report

Late-onset occlusion of the Monro foramina after endoscopic third ventriculostomy in adults: Case discussion and review of the literature

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ABSTRACT

Background: Few cases of adult idiopathic occlusion of the foramen of Monro (AIOFM) are described in the literature. The diagnosis of AIOFM after an endoscopic procedure is even more infrequent.

Case Description: We described the case of a 50-year-old woman who developed bilateral membranous occlusion of both Monro foramina 20 years after an endoscopic third ventriculostomy (ETV) for triventricular hydrocephalus due to an aqueductal stenosis. The patient underwent an endoscopic treatment (left foraminoplasty and septostomy) to check the patency of the stoma on the floor of the third ventricle. After the endoscopic procedure, the symptoms improved and the postoperative magnetic resonance imaging (MRI) demonstrated the resolution of the biventricular hydrocephalus.

Conclusion: Bilateral occlusion of both FM with consequent bilateral lateral ventricles enlargement is an extremely rare condition, especially if we consider the cases of biventricular hydrocephalus after endoscopic procedures. In our opinion, an endoscopic approach should be attempted as first choice procedure, avoiding any intraventricular stent or shunt placement.

Keywords: Endoscopic third ventriculostomy, Foraminoplasty, Hydrocephalus, Idiopathic, Neuroendoscopy, Septostomy

INTRODUCTION

Adult idiopathic occlusion of the foramen of Monro (AIOFM) is an exceeding rare condition, with few cases reported in the literature.[1,3,5,6,8-14,16,22-25] In particular, the diagnosis is established in patients with bilateral enlargement of the lateral ventricles with a small third ventricle on computed tomography (CT) scan or magnetic resonance imaging (MRI), ruling out any secondary cause of FM obstruction.[6]

A recent review has classified the AIOFM into four classes, on the bases of the symmetry of the lateral ventricles enlargement (and, consequently, on the deviation of the septum) and the presence/absence of membranes at the level of the FM.[6]

In our experience, only four cases of bilateral occlusion of FM after previous endoscopic procedures have been reported in the literature.[7,15,17,18] In this paper, we describe the case of a...
woman with bilateral symmetric dilatation of the lateral ventricles with a slit third ventricle due to membranous occlusion of both FM twenty years after an endoscopic third ventriculostomy (ETV).

**CASE DESCRIPTION**

Due to frequent episodes of headache, a 30-year-old female underwent neuroradiological investigations that documented the presence of an aqueductal stenosis with triventricular hydrocephalus and transependymal transudation [Figure 1]. She underwent an ETV through a right-sided burr hole.

During the procedure, no hemorrhage occurred. Neither symptoms nor signs of infection were identified during the hospitalization and the patient's symptoms improved. She underwent regular clinical and neuroradiological follow-up. In particular, the MRI demonstrated a decrease in size of the third and the lateral ventricles, with the resolution of the transependymal transudation. The fast spin-echo (FSE) sequences revealed the patency of the stoma and the regular flow void inside it.

Twenty years after the endoscopic procedure, the patient complained again of severe headaches, acute onset of gait imbalance, dizziness, and vomiting. The MRI showed a dilatation of the lateral ventricles with slight deviation of the septum and slit third ventricle, indicating an obstruction of both FM [Figure 2]. Because of a mild dilatation of the left lateral ventricle and an apparent enlargement of the Monro foramen aditus, we performed a left-sided approach for homolateral planned foraminoplasty and septostomy. Using a rigid endoscope (OI HandyPro Neuro-Endoscope Karl Storz Endoskope, Tuttingen, Germany), we observed the left FM to be completely occluded by a fibrous membrane. We performed a foraminoplasty using a laser coagulator (2 μm thulium laser, RevoLix Jr, LISA Laser USA, Pleasanton, CA, USA) to pierce the membrane and a 4 Fr NeuroBalloon catheter (Integra, Plainsboro, NJ, USA) to enlarge the slot. Through the left Monro foramen, we entered into a slit third ventricle and reported the patency of the ventriculostomy that was enlarged during the procedure. We observed, when retracting the endoscope, a clear pulsation of the cerebrospinal fluid (CSF) through the stoma. We also fenestrated the septum and, through the septostomy, evaluated the contralateral FM, which was also occluded by a fibrous membrane [Figure 3]. No complications occurred during the procedure nor the hospitalization. The analysis of the CSF did not reveal any abnormality.

After surgery, the symptoms improved and the early postoperative MRI showed the resolution of the biventricular hydrocephalus and the patency of septostomy, foraminoplasty, and ETV [Figure 4]. Trying to objectify the volumetric reduction of the ventricular system, we segmented the data of the pre operative and of the postoperative MRI (Elements, BrainLab, Munich, Germany). We obtained a preoperative ventricular system’s volume of 191.8 cm³, while in the postoperative MRI, the value was reduced to 135.9 cm³ [Figure 5].

A year later, the patient remained asymptomatic and no changes on the cerebral MRI were observed.

**DISCUSSION**

AIOFM is a rare condition without a clear etiology or a standardized treatment. In particular, various surgical treatments have been proposed, including endoscopic approaches (septostomy and/or unilateral or bilateral foraminoplasty, with the eventual use of an intraventricular stent) and monolateral or bilateral shunt placement.

The first report is that of Marions, in 1986, who described two cases of true bilateral stenosis of the FM in the pre-MRI era. One patient was treated microsurgically, and in both cases, a ventriculoperitoneal shunt (VPS) was implanted.

In 1992, Maxwell reported the case of a female with striking rapid memory loss and documented a type 1 AIOFM (sec. Ebrahimzadeh) on a CT scan. The patient underwent craniotomy and microsurgical septostomy with subsequent symptoms release.

The first case of AIOFM treated endoscopically was reported in 2001 by Javier-Fernández who performed a foraminoplasty using a bipolar cauterization without septostomy and Schonauer also used a similar combination. Other patients with integral septum pellucidum underwent unilateral foraminoplasty with septostomy. Only in rare cases did the left lateral ventricle dilate to such an extent that the third ventricle was also slit.

![Figure 1: T1-weighted MRI sequence performed before the endoscopic third ventriculostomy. On this MRI, performed 20 years ago and for this reason of suboptimal quality, we identified the presence of a dilatation of the supratentorial compartment of the ventricular system and the presence of a membranous aqueductal stenosis.](image-url)
cases, such as the acute onset of hydrocephalus ascribable to a reaction to the contrast used in ventriculography[^8] and in patients with bilateral obstruction due to true stenosis of the FM,[^13,16] the authors preferred a VPS placement.

Some observations have been made on the imaging data and the modality to perform the stomas. In particular, while in some cases, the diagnosis was reached using ventriculography[^9,12,14,25] or CT scan[^1,8,12-14,16,22,24,25] in others, MRI was preferred.[^1,3,5,8-11,13,14,16,22-25] Only De Bonis formulated the diagnosis of bilateral membranous occlusion of FM using an FSE and steady-state free processing (SSFP) high-resolution three-dimensional (3D) sequences MRI.[^5]

Relatively to the instrumentation, Schonauer concluded that the use of the thulium laser, instead of the Fogarty balloon, could reduce the risk of unintended violation of the fornix or of the other neural structures.[^24] Analogous considerations were made by Prontera who pointed out that the fenestration of an obstructing membrane is safer than the dilatation of a true stenotic foramen because of the high risk of damage to the fornix and the thalamostriate vein.[^22] Azab described a case of bilateral FM obstruction treated with bilateral foraminoplasty with the use of scissors and septum pellucidotomy, underlining that the initial use of blunt forceps and a Fogarty balloon catheter offered help with...
creating a larger opening through which sharp scissors’ blade could be safely utilized to complete the membrane opening.[13]

While there are some cases of AIOFM treated endoscopically,[1,3,5,6,8-11,13,16,22-25] patients with idiopathic occlusion of both FM after endoscopic procedures are rare.[7,15,17,18] In particular, Mori described a bilateral non-tumoral occlusion of the FM in a 22-year-old woman who had previously undergone a VPS placement through a right frontal approach because of an aqueductal stenosis. Six years after the shunt insertion, it was demonstrated a right-sided slit ventricle syndrome with a stenosis of the homolateral FM, which was treated with ETV and septostomy, with contextual VPS removal. Two years later, the patient presented with bilateral lateral ventricles dilatation due to bilateral membranous FM obstruction and occlusion of the septostomy. The authors performed bilateral foraminoplasty and septostomy with stent placement. It was speculated that multiple surgical stimuli with minor intraoperative bleeding associated with shunt surgery and/or ETV may have resulted in non-specific reactions with consequent FM obstruction.[17]

A similar case was reported by Migliorati: he described the case of a 52-year-old woman with bilateral occlusion of both foramina, who was initially treated with VPS insertion. Because of the onset of an abdominal infection, the shunt was removed and the bilateral dilatation of both lateral ventricles was treated with endoscopic foraminoplasty and septostomy. Rapidly, the hydrocephalus recurred and the patient underwent a ventriculoatrial (VA) shunt insertion resulting in resolution of her symptoms.[15]

El Rafee presented the case of a 30-year-old female who underwent ETV for occlusive hydrocephalus due to aqueductal stenosis. After 30 months, she reported recurrent headaches and the MRI demonstrated bilateral enlargement of the lateral ventricles with a collapsed third ventricle caused by bilateral stenosis of the FM. A left-sided endoscopic foraminoplasty and stenting of the left FM were performed with immediate neurological improvement. The authors ascribed the bilateral occlusion of the FM to the small diameters of these foramina before the first surgery: after the ETV, the diminished size of the lateral ventricles induced a decreasing in the FM diameter and the voluminous choroid plexus blocked the CSF flow within the foramina.[7]

Figure 4: Postoperative MRI. The postoperative MRI demonstrated the resolution of the dilatation of both lateral ventricles and of the transependymal transudation (a). We could identify the signs of our left-sided foraminoplasty (b), septostomy (c), and endoscopic third ventriculostomy (d).

Figure 5: Volumetric analysis. To objectify the reduction of the ventricular system dilatation, we segmented the preoperative (a-c) and postoperative MRI (d-f). Comparing axial (a,d), coronal (b,e), and sagittal (c,f) images, we identified a reduction of the biplanar dimensions of the ventricular system. This reduction was confirmed by the analysis of the preoperative (191.8 cm$^3$) and postoperative (135.9 cm$^3$) volumes.
Nagata reported the case of a 10-year-old girl with triventricular hydrocephalus due to aqueductal stenosis. She underwent ETV and reported no symptoms for 32 months. After that period, the patient presented with an exacerbation of headaches and the MRI findings demonstrated the bilateral occlusion of FM with a slim third ventricle. She underwent a septostomy (with monopolar microendoscopic electrode) and VPS insertion. The authors could not exclude the possibility of slight mechanical injury during the first endoscopic procedure nor of a subclinical postoperative intraventricular hemorrhage or of a ventriculitis. Because of the small dimension of the choroid plexus and consequent little production of CSF, Nagata hypothesized that the stagnation of the CSF could contribute to the occlusion of the FM.[18]

Similar to our experience, as in the majority of these cases, no sign of infection nor hemorrhage was reported.[7,17,18] Despite the other cases, our patient became symptomatic after 20 years from the first endoscopic procedure. We suppose that the *primum movens* in the etiology of this bilateral occlusion of the FM has to be ascribed to an alteration of the normal CSF dynamics that developed chronically. In this way, the symptoms manifested when the compensating systems failed. In particular, the reducing size of the lateral ventricles induced the walls of the FM to collapse around the choroidal plexus and to block the CSF flow. A non-specific fibrous reaction could have a role in the creation of a membrane at the level of the foramen, causing the CSF to accumulate in the lateral ventricles and the third ventricle to collapse.

Not relying only on macroscopic findings, we could not exclude the role of cellular abnormalities in the occlusion of both FM. While some authors focused their attention on ependymal cells[2,20] and choroid plexus epithelium[4,19] during brain ventricular system development, in a recent study, Olstad showed that motile cilia are crucial for generating a directional flow, which prevents the obstruction of ventricular ducts and, thereby, maintain the communication of CSF across ventricles in animal models.[21] Although zebrafish larvae with obstructed ducts do not develop hydrocephalus,[21] we could not exclude that a lack of flow causes the aggregation of molecules and could clog the ducts in adult human ventricles. Indeed, in our patient, there was not only an obstruction of both foramina but also the presence of several septa in the aqueduct, therefore, considering all these features as the result of a ciliopathy. Maybe, the absence or reduction of motile cilia function (without a clear etiology) could result in bilateral hydrocephalus due to the collapse of both FM, while the presence of an ETV could avoid the dilatation of the third ventricle. Even if the presence of these septa on the first MRI (and thus, preexistent respect any surgical intervention) could support the hypothesis of an individual tendency to develop intraventricular membranes, we cannot effectively rule out the possibility of a mechanical injury nor of a subclinical inflammation/infection nor of a minimal hemorrhage (not detectable on the postoperative CT) following the first endoscopic procedure.

Regarding the technique, we used a rigid endoscope obtaining a good visualization of the third ventricle and both lateral ventricles (through the septostomy). Mori reported a more maneuverability of the fiberscope than rigid endoscope inside the ventricular system.[17] Freudenstein noted that extensive inspection of the contralateral lateral ventricle was not possible due to restricted freedom of motion using a rigid endoscope.[16] Wong performed bilateral foraminoplasty for bilateral FM occlusion (after VPS implant and infection) through bilateral frontal burr holes using a rigid endoscope.[26]

We performed the left foraminoplasty, the septostomy using a laser coagulator, similarly to Schonauer,[24] and enlarging the slot with a 4 Fr NeuroBalloon catheter (Integra, Plainsboro, NJ, USA). We did not observe any symptom or sign of forniceal or thalamostriate vein damage, confirming the observations of other authors.[22,24]

Despite Mori[17] and El Rafee,[7] we did not use any stent and, unlike Nagata,[18] no VPS was inserted. Indeed, we obtained good CSF flow through the stomas not requiring any device and avoiding CSF shunts.

**CONCLUSION**

Bilateral occlusion of FM after endoscopic procedures is an extremely rare condition with no clear etiology nor encoded treatment.

In our opinion, the endoscopic approach should be attempted as the first choice procedure without any intraventricular stent or shunt placement. The use of the intraoperative laser could be of aid to avoid any damage of the internal veins during the foraminoplasty and the septostomy.

**Declaration of patient consent**

Institutional Review Board (IRB) permission obtained for the study.

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**Conflicts of interest**

There are no conflicts of interest.

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