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Controversial neuroendoscopic Monro foraminoplasty in the management of isolated lateral ventricle in an adult



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Max O. Krucoff^a, Moshe Chinn^b, Parker Babington^b, Zachary N. Litvack^{b,*}

^a Division of Neurosurgery, Duke University Medical Center, Durham, NC, USA

^b Department of Neurosurgery, The George Washington University Medical Center, Washington, DC, USA

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ABSTRACT

This report describes the clinical presentation, differential diagnosis, imaging characteristics, and current literature regarding the minimally invasive neuroendoscopic treatment of a rare condition: idiopathic unilateral hydrocephalus presenting late in adulthood. This paper also addresses the current controversy regarding septum pellucidotomy and/or Monro foraminoplasty as mono- or dual therapies for UH. A 63 year old African American female presented with headaches, blurry vision, and gait instability for six months. She was referred for neurosurgical consultation after an MRI demonstrated enlargement of the right lateral ventricle with left septal deviation. The patient underwent endoscopic transventricular fenestration of an idiopathic membrane occluding the foramen of Monro, as well as foraminoplasty of the foramen of Monro and septum pellucidotomy. Postoperatively the patient had transient difficulty with short term recall that improved rapidly, and she was discharged home on postoperative day 2. Follow-up one month later demonstrated complete resolution of her headache, blurry vision, and imbalance, as well as continued improvement of her memory. At six months, she had durable resolution of hydrocephalus and no short term memory complaints.

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Introduction

Unilateral hydrocephalus (UH) is a rare finding in adults and is usually caused by an asymmetrical obstruction of the foramen of Monro [1]. Etiologies of such an obstruction have included neoplastic (subependymal giant cell astrocytoma, hypothalamic glioma), infectious (ventriculitis, abscess), inflammatory (scarring from intraventricular hemorrhage), vascular (basilar artery aneurysm/ectasia, AV malformation of choid plexus, enlarged thalamostriate vein) and congenital (choroid plexus cyst/hypertrophy, anomaly of foramen of Monro), but rarely idiopathic [1-4]. In adults, this condition may present with symptoms on increased intracranial pressure, such as headache, incontinence, seizures, altered mental status, focal weakness, gait disturbance, visual field deficits, and/or papilledema [1]. Treatment options for this type of obstruction include CSF diversion (e.g. ventricular shunting, cyst shunting), craniotomy for excision of obstructive lesion, or creation of an alternate pathway for CSF flow (e.g. septum pellucidotomy, foraminoplasty, or fenestration of the obstruction) [4–7]. Here we present a case wherein the patient was diagnosed and treated successfully with neuroendoscopic exploration and fenestration of an obstructive membrane, septum pellucidotomy, and foraminoplasty of the foramen of Monro. We present a review and discussion of the neuroendoscopic treatment options and their

* Corresponding author.

E-mail address: zlitvack@mfa.gwu.edu (Z.N. Litvack). http://dx.doi.org/10.1016/i.inat.2015.03.004 associated risks and benefits, including a discussion of the controversial foraminoplasty of the foramen of Monro.

Case presentation

Clinical presentation

A 63 year old African American female presented with headache, blurry vision, and gait instability for six months. Her headaches occurred daily, were severe (10/10 on pain scale), and located at the vertex. She had no urinary incontinence or focal neurologic symptoms. Aside from balance instability on tandem walk, her exam and laboratory work up were unremarkable. Her past medical, family, and social histories were also noncontributory. She was referred for neurosurgical consultation after an MRI demonstrated enlargement of the right lateral ventricle with left septal deviation (Fig. 1). This finding was thought to be most consistent with an obstruction of the right foramen of Monro with transependymal flow secondary to trapping of the right lateral ventricle.

Surgical intervention

The patient underwent right frontal transventricular endoscopy for exploration and treatment. A 14 mm burr hole was drilled at 3 cm anterior to the coronal suture and 4 cm lateral to midline, similar to the placement used for endoscopic third ventriculostomy or resection of a colloid cyst. Proper placement is confirmed with neuronavigation and allows a trajectory that crosses orthogonal to the foramen of Monro, as well as one

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Fig. 1. T2 (left) and FLAIR (right) MRI sequences demonstrating unilateral hydrocephalus of the right lateral ventrical with septal deviation toward the left and transependymal flow.

that allows for both ventriculo-ventriculostomy and septum pellucidotomy. Based on the confluence of the septal vein, thalamostriate vein, and choroid plexus, the presumed location of the foramen of Monro was identified, but no foramen was seen (Fig. 2). Instead, a thin membrane was found. A Gaab Ventriculostomy Forceps (Karl Storz Endoskope, El Segundo, CA USA) was used to pierce this membrane, and a 3 French Fogarty catheter was used to cross into the third ventricle and dilate this new stoma. A Myriad micro-endoscopic debrider (manufactured by NICO Corp [Indianapolis, IN USA]) with combination variable suction and side cutting instrument was used to resect gliotic tissue at the foramen which was subsequently cauterized with bipolar to prevent re-closure. The floor of the third ventricle was visualized through this new Foramen of Monro. In a similar fashion, an endoscopic septum pellucidotomy was completed. Pulsatile flow was noted across the both ventriculoventriculostomies. On removal of the trocar under direct endoscopic visualization, a pulsatile membrane separate from the ependyma was noted. A clamped external ventricular drain (EVD) was placed for 23 h transduction of ICP in the right lateral ventricle.

Outcome and follow up

Post-operatively our patient had transient difficulty with short term recall but improved to baseline and was able to be discharged home on postop day 2. On one month follow up her headache and gait instability on tandem walk had completely resolved, and her memory continued to improve. At six months, a durable normalization of the ventricular



Fig. 2. Neuroendoscopic view of the septum pellucidum from the right lateral ventrical at the expected location of the foramen of Monro. Landmarks are noted (septal vein, thalamostriate vein, and choroid plexus), but no patent foramen is visible.

system was noted on CT (Fig. 3) and the patient continued to be without complaint of headache, gait imbalance, or short-term memory problems.

Discussion and literature review

Idiopathic unilateral hydrocephalus is a rare condition in adults [1]. In 1986 Shapiro et al. demonstrated that asymmetric ventricles on imaging alone can be a normal variant found on about 10% of head CTs [8]. They found that the normal variant is differentiated from a true abnormality by, among other findings, a significant shift of the septum pellicidum away from the dilated ventricle. In 2011, Vaz-Guimarães Filho et al. retrospectively reviewed a series of nearly 800 neuroendoscopic procedures from 1995 to 2010 and found only seven adult patients who had presented with unilateral hydrocephalus (six from intraventricular neurocysticercosis and one from congenital stenosis of the foramen of Monro) [6].

In 1985 Oi and Matsumoto defined the term progressive unilateral obstructive hydrocephalus [9]. They identified four causal categories based on the condition of the foramen of Monro: 1) congenital atresia, 2) acquired obstruction, 3) functional obstruction (e.g. from shunting), and 4) patent foramina with asymmetrical parenchymal abnormalities (i.e. alterations in compliance). As is the case with most literature on unilateral hydrocephalus, these authors dealt with an almost exclusively pediatric population. A literature review by Freudenstein et al. in 2002 found the most common obstructive lesion to the foramen of Monro in adults to be a colloid cyst, and that only seven cases of unilateral obstruction had ever been reported in this population [3].

In 2008, Schroeder et al. described several techniques for endoscopic treatment of different types of CSF obstructions, including septum pellucidum fenestration and foraminoplasty of the foramen of Monro which were used in this case [7]. Safe and efficacious results have been published most readily in the pediatric literature with data in adults limited to several small case series, as idiopathic unilateral hydrocephalus is exceedingly rare in this population. In 2010, Sharifi et al. published a report of 3 adult cases of unilateral hydrocephalus treated exclusively with neuroendoscopic septum pellucidotomy [2]. In this series and in Vaz-Guimarães Filho et al.'s series in 2011 mentioned above, septum pellucidotomy was found to be a sufficient as a monotherapy. However, both acknowledged that diverting CSF from two lateral ventricles through a single foramen of Monro may result in a partial obstruction.

Although septum pellucidotomy has become an accepted monotherapy for the treatment of unilateral hydrocephalus, there have been cases that warn against generating only one path for CSF flow. In 1989 Venkataramana treated a 31 year old patient for unilateral hydrocephalus with neuroendoscopic septum pellucidotomy [10]. After a two week asymptomatic period, the patient became unconscious with decerebrate posturing and died from respiratory



Fig. 3. Pre-operative (left) and post-operative (right) CT scan demonstrating unilateral hydrocephalus of the right lateral ventricle that resolved after treatment.

arrest. Additionally, in 2002, Freudenstein et al. reported a case of bilateral ventriculomegaly that was treated with endoscopic perforation of the obstructing membrane [3]. On postop day 3, however, the patient deteriorated clinically and required further intervention. The authors of that report believe the deterioration to have been caused by an inflammatory re-occlusion of the surgically created stoma vs. increased CSF production triggered by intraventricular contrast. They therefore they advocate creating an additional pathway (e.g. third ventriculostomy or septum pellucidotomy) in addition to fenestration of the occlusion.

In light of the above, a foraminoplasty was performed in our patient as a second pathway for CSF flow. This technique is more controversial given its proximity to the fornix and risk for post-operative memory deficits, yet it is well described as a therapy for bilateral foraminal stenosis [7]. Given the appropriate visualization, landmarks, and precautions, this technique can be performed without permanent memory deficits, as was seen in our case and the cited articles.

In regard to the surgical approach, Gangemi et al. advocate cannulating the occipital pole of the normal ventricle, as they feel it gives the best trajectory, the best opportunity to identify nondistorted landmarks, and the best chance to prevent healing of the stoma by not decompressing the enlarged ventricle intraoperatively [6]. They were also able to fenestrate the obstructing choroid cysts from the contralateral ventricle. In Sharifi et al.'s case report of 3 adults with unilateral hydrocephalus, they approached two via the enlarged ventricle and one via the non-enlarged ventricle because of severe septal deviation [2]. In the latter case, they felt that the difficulty and risk of perforating the septum from the enlarged side were too high. In our case, membrane fenestration, foraminotomy of the foramen of Monro, and septum pellucidotomy were achieved after cannulating the frontal pole of the larger ventricle. This approach enabled us to visualize the membrane obstructing the foramen of Monro as well as create a redundant pathway in case one were to heal over time. However, had the septum been pushed farther toward the contralateral ventricle, it is conceivable that we may have altered our approach.

Conclusion

Idiopathic unilateral hydrocephalus is a rare condition in the adult population, and in our case was caused by an idiopathic membrane obstructing the foramen of Monro. Currently the treatment of choice for UH is neuroendoscopic septum pellucidotomy, although we advocate creating a second, redundant pathway for CSF flow in light of case reports of delayed occlusions and their potentially devastating consequences. Although foraminoplasty is a controversial procedure, it can be performed safely and without permanent memory deficit if the proper landmarks are visualized and care taken during the procedure. The approach to the septum is left to the discretion of surgeon, mostly depending on the nature of the occlusion and degree of septal deviation.

Appendix A. Supplementary data

Supplementary data to this article can be found online at http://dx. doi.org/10.1016/j.inat.2015.03.004.

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