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Clinical commentary

Idiopathic bilateral occlusion of the foramen of Monro: An unusual entity with varied clinical presentations

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ABSTRACT

We review our experience with four patients who presented to our Medical Center from 2005–2015 with adult idiopathic occlusion of the foramen of Monro (FM). All patients underwent CT scanning and MRI. Standard MRI was performed in each patient to rule out a secondary cause of obstruction (T1-weighted without- and with gadolinium, T2-weighted, fluid-attenuated inversion recovery [FLAIR] and diffusion-weighted imaging [DWI] protocols). When occlusion of the FM appeared to be idiopathic, further high-resolution MRI with multiplanar reconstructions for evaluation of stenosis or an occluding membrane at the level of the FM was performed (T1-weighted without- and with gadolinium, T2-weighted 3D turbo spin-echo). Occlusion of the FM was due to unilateral stenosis and septum pellucidum deviation in two patients, to an occluding membrane in one, and to bilateral stenosis in one patient. Urgent surgical intervention is mandatory when there are signs of increased intracranial pressure while asymptomatic patients may be managed conservatively. In this patient series, truly bilateral stenotic obstruction of the FM was best managed with ventriculoperitoneal shunt and patients with membranous obstruction or unilateral stenosis with septum deviation were treated endoscopically.

1. Introduction

Bilateral occlusion of the foramen of Monro (FM) can be caused by a wide range of disorders, including meningioma, subependymal giant cell astrocytoma, hypothalamic glioma, choroid plexus tumors, colloid cysts, inflammatory conditions (e.g. bacterial ventriculitis, brain abscess), congenital problems (e.g. choroid plexus cyst, FM atresia), vascular malformations, basilar artery aneurysm or ectasia, and iatrogenic complications.

In rare cases, no cause is identified and the occlusion is considered idiopathic. Idiopathic occlusion may present with a spectrum of clinical characteristics in adult patients. Determining the physiopathology of the occlusion may be challenging, resulting in a therapeutic dilemma [1].

We present a series of four consecutive patients with adult idiopathic occlusion of the FM, present our algorithm for diagnosis and treatment, and review the literature.

2. Material and methods

We reviewed clinical records from 2005–2015 to identify all patients 18 years of age and above with a primary diagnosis of obstructing hydrocephalus at the level of the FM who were treated in our Department. In the current study, we included patients who were found to have an idiopathic occlusion at the FM. We reached a diagnosis of idiopathic occlusion in patients with bilateral enlargement of the lateral ventricles and a comparatively small third ventricle, with no evidence of a space-occupying lesion (SOL) or vascular lesion, congenital malformation, or inflammatory process that could cause biventricular obstruction. Patients with an obstructing lesion at the level of the FM, signs of communicating hydrocephalus, unilateral lateral ventricle enlargement, or signs of obstruction at another cerebrospinal fluid (CSF) pathway but not at the FM were excluded from the study.

The clinical and imaging records of patients included in the study were retrospectively reviewed. Details regarding their clinical presentation, imaging findings, management, and neurological status were recorded. Our Institutional Review Board approved the study design with a waiver of the requirement for informed consent.

2.1. Diagnostic procedure and management of the idiopathic obstruction

All patients underwent initial head CT scan without and with contrast if there were no contraindications to contrast

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administration. As a second step, our standard head MRI imaging examination was performed, including a T1-weighted study without and with gadolinium, T2-weighted, fluid-attenuated inversion recovery (FLAIR), and diffusion-weighted imaging (DWI) protocols to rule out secondary causes of Monro occlusion.

In patients where the obstruction of the FM was determined to be idiopathic, high resolution T1-weighted MRI without- and with gadolinium, and high-resolution T2-weighted 3-dimensional turbo spin-echo (TSE) MRI studies were performed. Multiplanar reconstructions at the level of the FM were then examined to determine whether the obstruction was due to true bilateral stenosis, unilateral stenosis with septum pellucidum deviation, or an occluding membrane. Patients with bilateral stenosis were managed with ventriculoperitoneal (VP) shunt insertion and septum pellucidotomy; obstructions due to unilateral obstruction or a membrane at the FM were treated using neuroendoscopy techniques. In patient who presented mild symptoms and no neurological deficit, conservative management was considered.

2.2. Literature search

A comprehensive literature search was performed using Pubmed, Medline, and Google Scholar to identify relevant English-language studies published from 1980–2015. Search terms included foramen of Monro, hydrocephalus, biventricular hydrocephalus, stenosis, ventriculoperitoneal shunt, VP shunt, septum pellucidotomy, and neuroendoscopy. Additional reports were identified from reference lists. Details regarding the diagnosis and management of patients with idiopathic obstruction of the FM were extracted from the literature and summarized, in the hope that a thorough review would help to elucidate a possible treatment algorithm.

3. Results

A total of 312 adult patients with a primary diagnosis of hydrocephalus were reviewed, and only four (1.3%) met the criteria for idiopathic bilateral occlusion of the FM. Their demographic information, clinical presentation, and management data are presented in Table 1.

3.1. Patient descriptions

3.1.1. Patient No. 1

A 39-year-old man was admitted through the emergency room after being found on the floor of his home in a comatose state. The patient arrived intubated. He was in a superficial coma with mid-sized reactive pupils and alternating bilateral localizing motor response to pain (Glasgow Coma Score [GCS] 6). His history was notable for a diagnosis of arrested biventricular hydrocephalus 6 years earlier. His wife reported that he had ingested water compulsively the night before arrival. Admission head CT scan without contrast showed a massive enlargement of both lateral ventricles with small third and fourth ventricles, with no evidence of an SOL or other cause of obstruction (Fig. 1A). His laboratory reports revealed hemodilution (hemoglobin 12.2 g/dl compared to a normal range of 14–18 g/dL, hematocrit 35.2%), and hyponatremia (blood sodium 128 mmol/L vs. a normal range of 135–145 mmol/L). The patient underwent uneventful emergent bilateral ventriculostomy. He made a rapid and complete neurological recovery after 12 hours.

MRI performed 24 hours after ventriculostomy ruled out secondary causes of Monro occlusion. Additional high-resolution MRI studies were performed as described above. Multiplanar reconstructions at the level of the FM confirmed the absence of an SOL (Fig. 1B), and revealed true bilateral stenosis of the FM (Fig. 1C). On day 5, bilateral ventriculostomy was replaced by permanent navigation-guided biventricular peritoneal (VP) shunt with trans-septum pellucidotomy (Fig. 1D). At 6-month follow-up, the patient had completely recovered with no residual neurologic deficit.

3.1.2. Patient No. 2

A 27-year-old woman, with a history of headaches over the past 6 months was referred to our outpatient neurosurgical clinic. Neurological examination did not reveal any neurologic deficit; however, bilateral papilledema was noted. Head CT scan showed biventricular enlargement with transependymal edema and signs of obstruction at the level of the FM. MRI ruled out secondary causes of Monro occlusion. Further high-resolution MRI examination with multiplanar reconstructions at the level of the FM revealed unilateral FM stenosis with septum deviation. The patient underwent endoscopic septum pellucidotomy. Her headaches were fully relieved at 1-month follow-up and her papilledema was resolved after 3 months.

3.1.3. Patient No. 3

A 25-year-old woman was referred to the Department of Neurosurgery after 2 weeks of periodic headache and dizziness. Careful examination did not reveal any neurological deficit or papilledema. Head CT scan showed biventricular enlargement with transependymal edema and obstruction at the level of the FM. MRI ruled out secondary causes of Monro occlusion and further high-resolution MRI with multiplanar reconstructions revealed unilateral FM stenosis with septum deviation. The patient was managed conservatively due to the absence of neurologic deficits and signs of high intracranial pressure (ICP) (no papilledema). At 3-year follow-up the patient remains asymptomatic without papilledema.

Table 1
Presentation and management of patients with idiopathic bilateral occlusion of the foramen of Monro

Patient No.	Gender/ Age	Presentation	Imaging findings	Papilledema	Treatment
1	M/39	Comatose state, chronic headache	MRI – "Truly" bilateral stenosis	N/A	Bilateral vestriculostomies and VP shunt
2	F/27	Chronic headache	MRI – Unilateral stenosis and septum deviation	+	Neuroendoscopy
3	F/25	Subacute headache	MRI – Unilateral stenosis and septum deviation	–	Conservative
4	F/40	Chronic headache, focal seizure, vomiting	MRI – Bilateral membrane	–	Conservative – with anticonvulsant treatment

F = female, M = male, N/A = not available, VP = ventriculoperitoneal, + = present, – = absent.