



Case Report

Idiopathic fourth ventricular outlet obstruction misdiagnosed as normal pressure hydrocephalus: A cautionary case

Joshua A. Cuoco¹, Michael J. Benko¹, Brendan J. Klein¹, David C. Keyes², Biraj M. Patel³, Mark R. Witcher¹

¹Section of Neurosurgery, Carilion Clinic, ²Diagnostic Radiology, Department of Radiology, Carilion Clinic, ³Neurointerventional Surgery, Department of Radiology, Carilion Clinic, Roanoke, Virginia, United States.

E-mail: *Joshua A. Cuoco - jacuoco@carilionclinic.org; Michael J. Benko - mjbenko@carilionclinic.org; Brendan J. Klein - bklein1@carilionclinic.org; David C. Keyes - dckeyes@carilionclinic.org; Biraj M. Patel - bmpatel@carilionclinic.org; Mark R. Witcher - mrwitcher@carilionclinic.org



*Corresponding author:

Joshua A. Cuoco,
Section of Neurosurgery,
Carilion Clinic, Roanoke,
Virginia, United States.

jacuoco@carilionclinic.org

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ABSTRACT

Background: Fourth ventricular outlet obstruction is an infrequent but well-established cause of tetraventricular hydrocephalus characterized by marked dilatation of the ventricular system with ballooning of the foramina of Monro, Magendie, and Luschka. Multiple processes including inflammation, infection, hemorrhage, neoplasms, or congenital malformations are known to cause this pathological obstruction. However, true idiopathic fourth ventricular outlet obstruction is a rare phenomenon with only a limited number of cases reported in the literature.

Case Description: A 61-year-old female presented with several months of unsteady gait, intermittent headaches, confusion, and episodes of urinary incontinence. Conventional magnetic resonance imaging demonstrated tetraventricular hydrocephalus without transependymal flow, but with ventral displacement of the brainstem and dorsal displacement of the cerebellum without an obvious obstructive lesion on pre- or post-contrast imaging prompting a diagnosis of normal pressure hydrocephalus. However, constructive interference in steady state (CISS) and half-Fourier acquisition single-shot turbo spin echo (HASTE) sequences followed by fluoroscopic dynamic cisternography suggested encystment of the fourth ventricle with thin margins of arachnoid membrane extending through the foramina of Luschka bilaterally into the pontocerebellar cistern. Operative intervention was pursued with resection of an identified arachnoid web. Postoperative imaging demonstrated marked reduction in the size of ventricular system, especially of the fourth ventricle. The patient's symptomatology resolved a few days after the procedure.

Conclusion: Here, we describe an idiopathic case initially misdiagnosed as normal pressure hydrocephalus. The present case emphasizes the necessity of CISS sequences and fluoroscopic dynamic cisternography for suspected cases of fourth ventricular outlet obstruction as these diagnostic tests may guide surgical management and lead to superior patient outcomes.

Keywords: Arachnoid web, Endoscopic third ventriculostomy, Fourth ventricular outlet obstruction, Noncommunicating hydrocephalus, Obstructive hydrocephalus, Suboccipital craniectomy

INTRODUCTION

Fourth ventricular outlet obstruction is an infrequent but well-established cause of tetraventricular hydrocephalus characterized by marked dilatation of the ventricular system with ballooning of the foramina of Monro, Magendie, and Luschka. However, the majority of cases result from prior parenchymal insults, including inflammatory, infectious, hemorrhagic, neoplastic, or congenital

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etiologies.^[8,11] Indeed, true idiopathic fourth ventricular outlet obstruction is a rare phenomenon with only a limited number of cases reported in the literature.^[1-3,5-7,9,10] Here, we describe an idiopathic case initially misdiagnosed as normal pressure hydrocephalus. Moreover, we discuss our diagnostic workup as well as current surgical management and operative approaches based on the relevant literature.

CASE REPORT

A 61-year-old female presented with several months of unsteady gait, intermittent headaches, confusion, and episodes of urinary incontinence. Her past medical history was unremarkable. Physical examination was notable for impaired tandem gait without focal deficits. Conventional magnetic resonance imaging (MRI) demonstrated tetraventricular hydrocephalus without transependymal flow, but with ventral displacement of the brainstem and dorsal displacement of the cerebellum without an obvious obstructive lesion on pre- or post-contrast imaging. Previous outpatient neurology workup concluded a diagnosis of normal pressure hydrocephalus based on her clinical symptomatology and radiographic findings, which prompted neurosurgical consultation. However, given the degree of fourth ventricular dilatation and subtle displacement of the brainstem and cerebellum, additional imaging sequences were ordered to rule out fourth ventricular outlet obstruction before considering surgical intervention (e.g., ventriculoperitoneal shunting). Constructive interference in steady state (CISS) and half-Fourier acquisition single-shot turbo spin echo (HASTE) MRI sequences suggested encystment of the fourth ventricle with thin margins of arachnoid membrane extending through the foramina of Luschka bilaterally into the pontocerebellar cistern [Figure 1]. These findings suggested the diagnosis of noncommunicating tetraventricular hydrocephalus secondary to mechanical obstruction at the outlet of the fourth ventricle. As such, a fluoroscopic dynamic cisternogram through an external ventricular drain was performed to confirm outlet obstruction,

defined the boundaries of encystment, and determine the operative approach for definitive surgical intervention.

Digital subtraction cisternography demonstrated contrast outflow through the foramina of Monro and cerebral aqueduct; however, there was a paucity of flow through the preponderance of the fourth ventricle with contrast tracking peripherally along the ependymal walls and stopping just past the apertures of the foramina of Magendie and Luschka without extension into the basal cisterns [Figure 2]. Delayed fluoroscopic images confirmed persistent obstruction at the level of the fourth ventricle outflow pathways. Given this localization of obstruction, a suboccipital craniectomy was performed for exploration and intervention. A thick, dense, opaque-white arachnoid membrane was identified at the foramen of Magendie masking the obex [Figure 3]. This arachnoid membrane was completely excised after mobilization of the tonsils, restoring cerebrospinal fluid (CSF) flow dynamics. Frozen sections were consistent with benign fibrovascular tissue, however, a meningotheelial lining was not observed to confirm the diagnosis of an arachnoid cyst. CSF studies were unremarkable. The external ventricular drain was clamped to monitor for 3 days and then removed on postoperative day 3 without complication. Postoperative MRI demonstrated a significant decrease in the size of the ventricular system with marked reduction in the size of the fourth ventricle and interval resolution of brainstem displacement [Figure 4]. Postoperatively, her gait returned to baseline with complete resolution of her headache, confusion, and urinary incontinence. At 3-month clinical follow-up, she was without symptomatology.

DISCUSSION

Idiopathic fourth ventricular outlet obstruction is an exceedingly rare cause of noncommunicating hydrocephalus.^[1-3,5-7,9,10] In pediatrics, it is typically the consequence of posterior fossa malformations, whereas

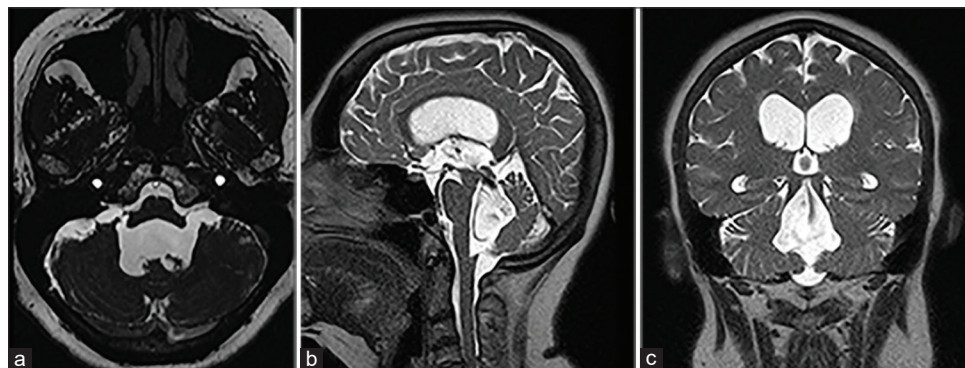


Figure 1: Preoperative MRI with tetraventricular hydrocephalus. Note the marked dilatation of the fourth ventricle with ventral displacement of the brainstem and dorsal displacement of the cerebellum with ballooning of the foramina of Luschka and Magendie. (a) CISS. (b and c) HASTE.

common causes in adults are secondary to processes such as inflammation, infection, hemorrhage, trauma, neoplasms, or Chiari malformations.^[8,11] Such pathologies impede CSF flow primarily in narrow pathways, such as the cerebral aqueduct or foramina, causing upstream ventricular dilatation.

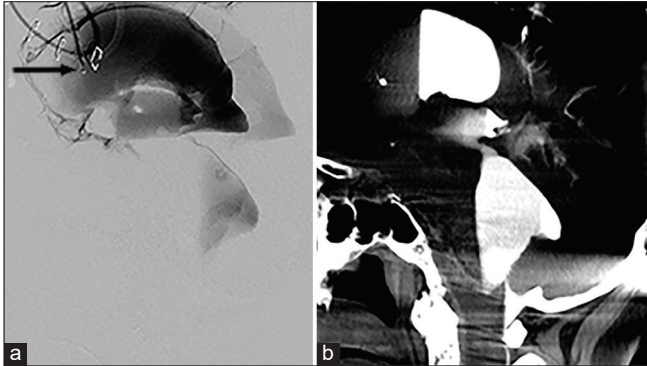


Figure 2: (a and b) Fluoroscopic dynamic cisternography demonstrating dilatation of the lateral and third ventricles with adequate flow through the foramina of Monro and cerebral aqueduct; however, a paucity of outflow is appreciated from the fourth ventricle through either foramina of Luschka or Magendie. The black arrow denotes the tip of the external ventricular drain catheter.

On the contrary, balanced ventricular enlargement is typically suggestive of communicating hydrocephalus, as is seen in the setting of adults with normal pressure hydrocephalus. As such, differentiating noncommunicating from communicating hydrocephalus based on imbalanced or balanced ventricular dilatation, respectively, is relatively straightforward. However, fourth ventricular outlet obstruction can also present with balanced tetraventricular hydrocephalus with the impediment of CSF flow at the outlet of the fourth ventricle to the subarachnoid space causing diagnostic difficulties. The present case demonstrated the classic findings seen in the fourth ventricular outlet obstruction including tetraventricular hydrocephalus with ballooning of all foramina, including Magendie and Luschka. Ventral displacement of the brainstem and dorsal displacement of the cerebellum are also classically associated with the diagnosis. Certainly, it is difficult to rule out fourth ventricular outlet obstruction based on conventional MRI sequences. As such, additional MRI sequences, including CISS and HASTE, may be able to confirm suspicion for membranous obstruction. For example, Dincer *et al.*^[2] investigated the utility of CISS compared with conventional MRI sequences in the diagnosis of obstructive membranes in 134 patients with a known diagnosis of communicating

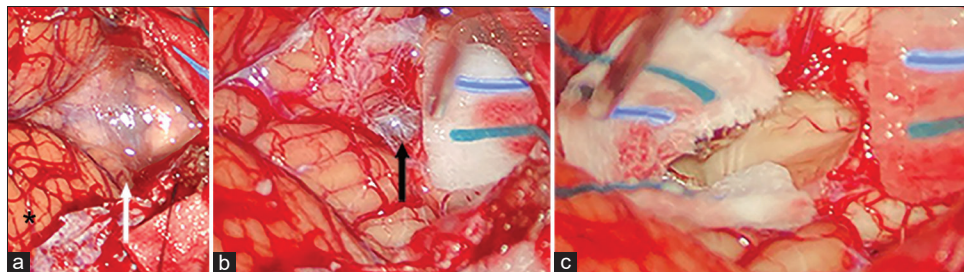


Figure 3: (a) Initial suboccipital craniectomy exposure of the cerebellar tonsils (black asterisk) and dense, adherent arachnoid within the cistern magna (white arrow). (b) Visualization of a thick, dense, and opaque arachnoid web (black arrow) found to be obstructing the foramen of Magendie. (c) Restoration of normal cerebrospinal fluid flow through a patent foramen of Magendie after circumferential excision of the arachnoid web.

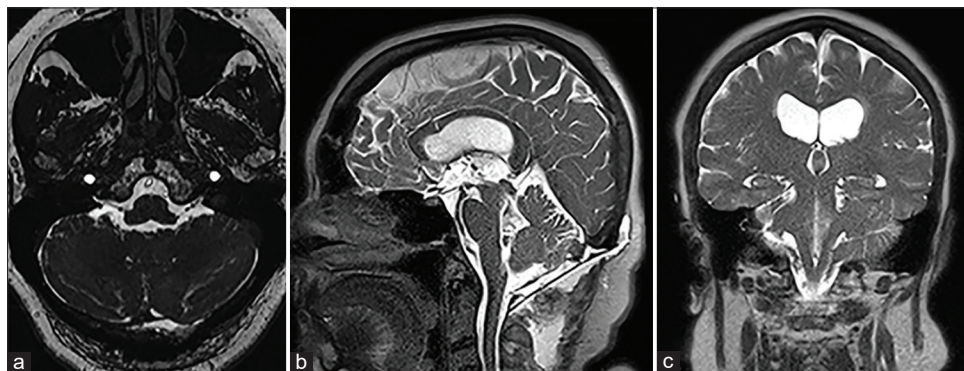


Figure 4: Postoperative magnetic resonance imaging with a significant decrease in the size of the ventricular system, including the fourth ventricle, and resolution of brainstem and cerebellar displacement. (a) CISS. (b and c) HASTE.

or noncommunicating hydrocephalus. The authors found 26 new cases (19.4%) of noncommunicating hydrocephalus with CISS images that were initially misdiagnosed as communicating hydrocephalus based on conventional imaging.^[2] Importantly, fourth ventricular outlet obstruction at the foramina of Luschka was found in 22 of the 26 newly diagnosed noncommunicating cases.^[2] Indeed, these data emphasize the additive value of CISS in identifying obstructive membranes and, ultimately, drastically altering patient treatment strategies and prognosis. Alternative methods to detect outlet obstruction include cisternography, the most sensitive diagnostic method, and direct endoscopic visualization of the fourth ventricle.^[1] Longatti *et al.*^[7] utilized transaqueductal endoscopic navigation to visualize outlet obstruction in 10 patients classifying the various degrees of membranous stenosis including (i) thick and opaque membrane, (ii) transparent spider web-like membrane, and (iii) a dense membrane with fissures acting as valves. In the present case, we identified a thick, dense, and opaque membrane at the foramen of Magendie without fissures or valves.

The surgical management of idiopathic fourth ventricular outlet obstruction remains controversial. Numerous approaches have been documented in the literature including suboccipital craniotomy for posterior fossa exploration with excision of the membranous obstruction, endoscopic third ventriculostomy, endoscopic transventricular transaqueductal foraminoplasty of Magendie, and/or Luschka, fourth ventriculocisternostomy and ventriculoperitoneal shunting for functional treatment.^[1,3-11] Bai *et al.*^[1] advocated for microsurgical excision of the membranous occlusion through suboccipital craniotomy as an effective and reliable treatment. At 1-year follow-up, the authors reported no radiographic evidence of recurrent hydrocephalus in their case.^[1] Mohanty *et al.*^[8] assessed the efficacy of endoscopic third ventriculostomy in 22 patients with fourth ventricular outlet obstruction. The authors reported a 65% success rate with the majority of radiographic failures (6 of the 7 patients) occurring within 6 weeks from the initial procedure.^[8] Torres-Corzo *et al.*^[11] reported success rates of endoscopic transventricular transaqueductal foraminoplasty of Magendie and/or Luschka in 30 patients with obstructive hydrocephalus secondary to the fourth ventricular outlet obstruction. Seventeen of the 26 patients (65.3%) with long-term follow-up improved clinically and did not require further intervention; however, eight patients failed to improve and required an additional procedure.^[11] Ventriculoperitoneal shunts have been utilized with varying degrees of success, but caution should be used in these cases as CSF shunting from the lateral or third ventricles to the peritoneal cavity may only serve as a temporizing measure and fail to relieve a “trapped” fourth ventricle.^[4,9] For example, Shimoda *et al.*^[9] reported a case initially misdiagnosed as normal

pressure hydrocephalus that underwent ventriculoperitoneal shunting. Nine months after the initial procedure, repeat imaging demonstrated slit-like lateral ventricles with marked dilatation of the fourth ventricle and return of the patient’s presenting symptomatology.^[9] The authors removed the ventriculoperitoneal shunt and temporarily replaced it with an external ventricular drain followed by endoscopic third ventriculostomy.^[9] Postoperative imaging demonstrated marked reduction in the size of the fourth ventricle. This highlights the importance in obtaining additional MRI sequences and, if necessary, fluoroscopic dynamic cisternography for suspected cases of fourth ventricular outlet obstruction before considering ventriculoperitoneal shunt placement.

CONCLUSION

Here, we described a rare case of idiopathic fourth ventricular outlet obstruction diagnosed with fluoroscopic dynamic cisternography and treated successfully with suboccipital craniectomy for microsurgical excision of an encountered arachnoid web. Noncommunicating hydrocephalus attributable to outlet obstruction can be misdiagnosed as communicating hydrocephalus due to conventional MRI, which lacks the sensitivity to identify membranous occlusions. The present case emphasizes the necessity in obtaining CISS sequences and fluoroscopic dynamic cisternography for suspected cases of the fourth ventricular outlet obstruction as the results of such diagnostic tests may guide surgical management strategies and lead to superior patient outcomes.

Declaration of patient consent

Patient’s consent not required as patients identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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