

Idiopathic Fourth Ventricle Outlet Obstruction Successfully Treated by Endoscopic Third Ventriculostomy: a Case Report

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Abstract

Introduction: Fourth ventricle outlet obstruction (FVOO) is a rare cause of obstructive hydrocephalus. We describe a case of idiopathic FVOO that was successfully treated with endoscopic third ventriculostomy (ETV).

Case report: A 22-year old female without any remarkable medical history presented with a headache and vomiting and blurring of vision. Magnetic resonance imaging (MRI) showed tetra-ventricular hydrocephalus associated with the dilatation of the fourth ventricle outlets, without any obstructive lesions. However, MRI suggested mechanical obstruction of the cerebrospinal fluid (CSF) at the fourth ventricle outlets. Thus, the patient was diagnosed as FVOO and ETV was performed; the hydrocephalus was subsequently resolved.

Conclusion: ETV should be considered for FVOO treatment, particularly in idiopathic cases without CSF malabsorption.

Keywords: Hydrocephalus, Fourth ventricle outlet obstruction (FVOO), Endoscopic third ventriculostomy (ETV).

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Background:

Fourth ventricle outlet obstruction (FVOO) is an uncommon clinical condition that causes obstructive hydrocephalus. In FVOO, cerebrospinal fluid (CSF) is blocked at the fourth ventricle outlets by a

membranous structure in the absence of any additional obstructive organic pathologies¹. Various terms for referring to FVOO have been used in previous reports, such as fourth ventricle/ventricular outlet obstruction^{2,3,4,5}, fourth ventricular outflow obstruction⁶, membranous obstruction of the fourth ventricle outlet⁷, obstruction of Magendie's and Luschka's foramina⁸, obstruction of fourth ventricular exit⁹ and primary obstruction of the fourth ventricle outlets¹⁰. Far distal obstructive hydrocephalus is a term that includes Dandy Walker or Arnold Chiari malformation, membranous obstruction or fourth ventricle and intercisternal external obstruction of the CSF¹¹. The etiology and pathogenesis of FVOO are unclear, although some cases present with a history of meningitis or intraventricular hemorrhage. In the present report, we describe the case of idiopathic FVOO without any remarkable medical history that was successfully treated by endoscopic third ventriculostomy (ETV).

Case report :

A 22-year-old female presented with a headache, vomiting and blurring of vision. She had no previous significant medical history. The patient had no neurological deficit except papilloedema on admission.

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Magnetic resonance imaging (MRI) showed enlargement of all ventricular systems associated with the dilatation of the foramina of Magendie and Luschka, with no obstructive organic lesions (such as brain tumors) on contrast enhanced MRI (Fig. 1 and 2). We had initially considered this case might be a communicating hydrocephalus because the fourth ventricle was dilated on MRI. However, the highly expanded fourth ventricle and its outlets were inconsistent with communicating hydrocephalus.

These findings suggested a mechanical obstruction at the outlets of the fourth ventricle. We therefore diagnosed the patient with FVOO and chose to

perform an ETV using a neuro-endoscope (Storz, Germany). A standard third ventriculostomy in the floor of third ventricle was performed on 27.03.2016. On endoscopic observation of the lateral ventricle and pre-pontine cistern, no abnormalities suggestive of previous meningitis or intraventricular hemorrhage were found.

The patient's postoperative course was uneventful with no signs of neurological deficit. Her vision was dramatically improved immediately after operation. MR images obtained 4 and a half month post-surgery revealed significant resolution of the hydrocephalus (Fig. 4a,b). The patient has remained in good

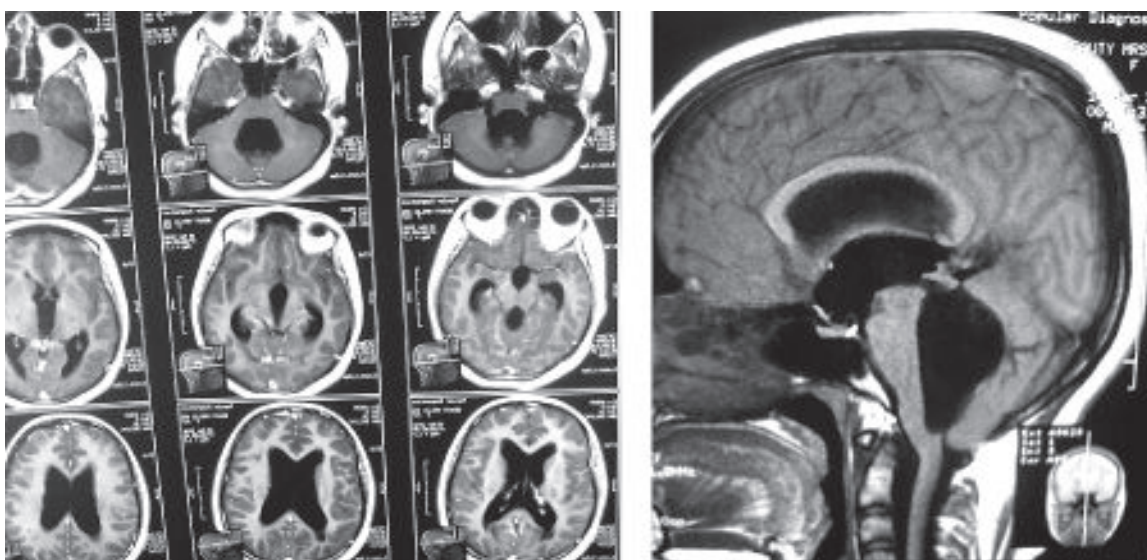


Fig.-1: MRI of brain contrast-enhanced T1-weighted axial and sagittal image on admission showed dilatation of both lateral, third and fourth ventricles, no obstructive lesion.

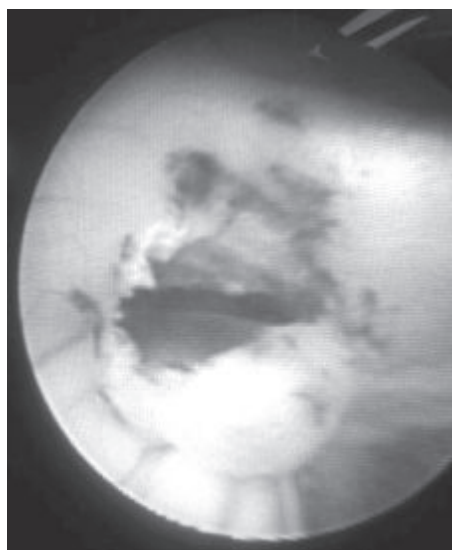


Fig.-2: Intraoperative view of ETV after fenestration of 3rd ventricular floor.

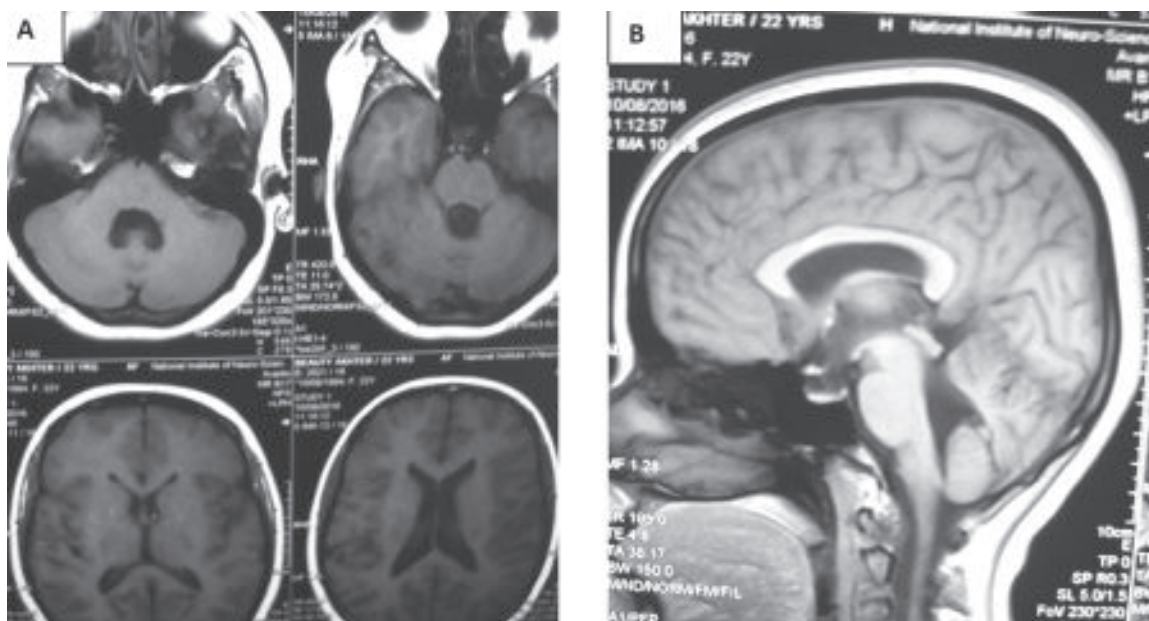


Fig.-3 A,B: Magnetic resonance imaging (MRI) of brain, performed 4 and a half months after ETV showed significant shrinkage of each ventricle, improvement of the hydrocephalus.

condition without recurrence of hydrocephalus followed up to 11 months in the postoperative period.

Discussion:

FVOO is a rare cause of obstructive hydrocephalus. Although many studies on FVOO have been published, the pathogenic mechanism of this condition remains unclear. FVOO tends to occur in children and may be congenital^{12,13,14}, but adult cases are also in fact common. There have been no reports that gender affects the prevalence of this condition. The present case had no obvious medical histories. These facts suggest that the present case represents a case of 'idiopathic' FVOO.

Diagnostic modalities for FVOO:

It is difficult to confirm the presence of a membranous obstruction via conventional MRI. High-resolution three-dimensional constructive interference with steady state sequence on 3T MRI may be able to detect obstructive membranes¹⁵, although this may not be possible in all cases¹⁰. The most sensitive diagnostic method is CT ventriculography, with the injection of contrast medium through a ventricular catheter^{13,3}. Serial CT images after injection will show collected contrast medium in the outlets of the fourth ventricle and subsequent blockage of its diffusion to the pre-pontine cistern. One concern about this method is radiation exposure of the brain, particularly in younger children. It is recommended the use of MRI instead of CT as the diagnostic modality for FVOO in order to avoid exposure to radiation¹⁶. As

alternative examination to access the dynamics of CSF, efficacy of phase-contrast MRI^{8,6}, cine-MRI^{7,8,17,12,5,9,4} or radioisotope cisternogram^{8,4} is also reported. Another diagnostic option is direct endoscopic inspection of the fourth ventricle in case where the aqueduct is sufficiently expanded to safely insert a neuro-endoscope through it². Although this technique needs to be done under general anesthesia and carries a risk of damaging the midbrain around the aqueduct, it has recently been reported to be relatively safe^{18,19,2,20}. When FVOO is highly suspected solely with MRI, this technique could allow simultaneous diagnosis and treatment, thereby reducing the chance of radiation exposure, duration of hospitalization, and risk of drainage infection.

Treatment options of FVOO:

Although a ventriculo-peritoneal (V-P) shunt is the most conventional treatment for FVOO⁹, it is not in fact preferable in children, who represent the majority of patients. In the past, direct fenestration of the membranous occlusion through craniotomy was attempted for treating FVOO^{9,2}; however, recent studies have suggested that ETV is a less invasive and effective treatment strategy^{1,17,9,21,2,10,4}. Therefore, correct preoperative diagnosis is very important because ETV can eliminate the need for surgical implantation of a V-P shunt. In the imaging study with the 3D-CISS sequence on 3T MRI, they found 26 endoscopically treatable noncommunicating cases among 134 cases who had been previously diagnosed as communicating hydrocephalus by

conventional MR images¹⁵. It is reported the entire success rate of ETV for FVOO is 65 % (13 successes in 20 cases)². Although they did not evaluate the success rates of primary and secondary FVOOs separately, they speculated that failure was attributable to CSF malabsorption as a result of prior meningitis or intraventricular hemorrhage. Other case reports or case series^{7,17,5,9,2,4} of ETV performed exclusively for primary FVOO demonstrated obviously better outcome (75–100 %). According to these previous reports, ETV would be more effective in patients of primary FVOO. It is reported that most failures of ETV for treating FVOO occur within 6 weeks of surgery and that subsequent endoscopic re-exploration revealed patency at the fenestration site². Endoscopic exploration not only confirmed a highly stenosed fenestration site, but re-expansion of the fenestration also relieved the hydrocephalus. Endoscopic foraminoplasty by direct fenestration of membranous obstruction at the fourth ventricle outlets is another previously reported treatment option^{18, 9, 19}.

Conclusion:

In the present report, we describe a case of idiopathic FVOO with no remarkable medical history that resulted in the development of hydrocephalus. ETV should be considered as a treatment option for FVOO, particularly in idiopathic cases without CSF absorption disorders. Moreover, in such cases, endoscopic re-exploration of the fenestration would be effective, even when hydrocephalus recurs.

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