Fourth Ventriculostomy in Occlusion of the Foramen of Magendie Associated with Chiari Malformation and Syringomyelia

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We present four cases of hydrocephalus caused by occlusion of foramen of Magendie associated with Chiari Type I malformation and syringomyelia. The aim of this study is to evaluate the results of surgical treatment via fourth ventriculostomy with catheter from the fourth ventricle to the upper cervical subarachnoid space. Obstructive tetraventricular hydrocephalus due to occlusion of the foramina of Luschka and Magendie can be treated with cerebro-spinal fluid shunting, opening the membranes with suboccipital craniotomy, placement of a catheter, endoscopic third ventriculostomy, and endoscopic fourth ventriculostomy. Our aim was to solve all the pathologies such as Chiari malformation, hydrocephalus, and syringomyelia in one approach. Thus, the treatment consisted of posterior fossa decompression and exploration. All the patients were treated with suboccipital craniectomy and C1 laminectomy with excision of the membrane obstructing the foramen of Magendie. Fourth ventriculostomy with cathetering from fourth ventricle to upper cervical subarachnoid space was performed. The postoperative period was uneventful in all the patients. Neurological status of all the patients improved. Tetraventricular hydrocephalus and syrinx were reduced in the control cranial magnetic resonance imaging. Complications such as infection and catheter migration were not observed during the follow-up period. Treatment with fourth ventriculostomy using a catheter from fourth ventricle to upper cervical subarachnoid space could be a treatment of choice in cases with hydrocephalus caused by occlusion of the foramina of Magendie, with associated Chiari Type I malformation and syringomyelia.

Keywords: Chiari malformation, foramen of Magendie, tetraventricular hydrocephalus, ventriculostomy

Introduction

Fourth ventricular outlet obstruction (FVOO) may be congenital or acquired. Congenital occlusive lesions occur primarily in children and are seen mainly in association with Chiari malformations, Dandy–Walker cysts, tuberous sclerosis, spina bifida, platybasia, acondroplasia, basilar impression, and atlantooccipital fusion.[1–6] Idiopathic stenosis or closure of the foramina of Magendie and Luschka

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is a rare cause of obstructive hydrocephalus.[7] However, a distinct subgroup of patients present with radiological evidence of FVOO, in their adulthood, without any significant underlying etiology.[3] It is most often seen associated with an underlying pathology that is responsible for the inflammation and scarring of the outlet foramina; however, there have been a few reports in which no predisposing causes have been identified. Histopathological evaluation of the membrane has revealed neural tissue and gliosis without any active inflammation.[4]

The treatment for these malformations is still controversial. Hydrocephalus can be treated with cerebro-spinal fluid (CSF) shunting, opening the membranes with suboccipital craniotomy, placement of a catheter, and endoscopic third ventriculostomy (ETV) or endoscopic fourth ventriculostomy.

Materials and Methods

Informed patient consent was obtained for this procedure. All patients' data are summarized in Table 1. A 40-year-old man approached us with complaints of headache and neck pain. The patient's neurological examination was normal. Magnetic resonance imaging (MRI) revealed tetraventricular hydrocephalus, dilatation level of aquaductus Sylvii, tonsillar ectopia, and compression to the brain stem and cerebellum, and a membrane causing an obstruction at the level of foramen Magendie. Cervical MRI showed syringomyelia between C1 and C7. Cine-MR revealed no CSF flow through the foramen Magendie and foramen magnum. At the level of aquaductus Sylvii, turbulence, and tertiary flow were observed.

Second patient; a 44-year-old woman presented with weakness in bilateral upper extremity. She complained of numbness in the arms, defect in discriminating between hot and cold, and difficulty in swallowing. MRI showed tetraventricular hydrocephalus, tonsillar ectopia, and syringomyelia between C1 and L1. Neurological examination showed left 11th and 12th cranial nerve deficiency, reduced GAG reflex, quadriparesis, hyperactive reflexes, and bilaterally ankle clonus.

Third patient; a 33-year-old woman complained of headache. MRI revealed tetraventricular hydrocephalus and Chiari Type I malformation with syringomyelia between C1 and C5. Neurological examination was normal.

Fourth patient; a 21-year-old man presented with weakness in the hands and urinary incontinence. MRI demonstrated tetraventricular hydrocephalus, tonsillar ectopia, and syringomyelia between C1 and L1. Neurological examination

Table 1   Clinical characteristics of the patients

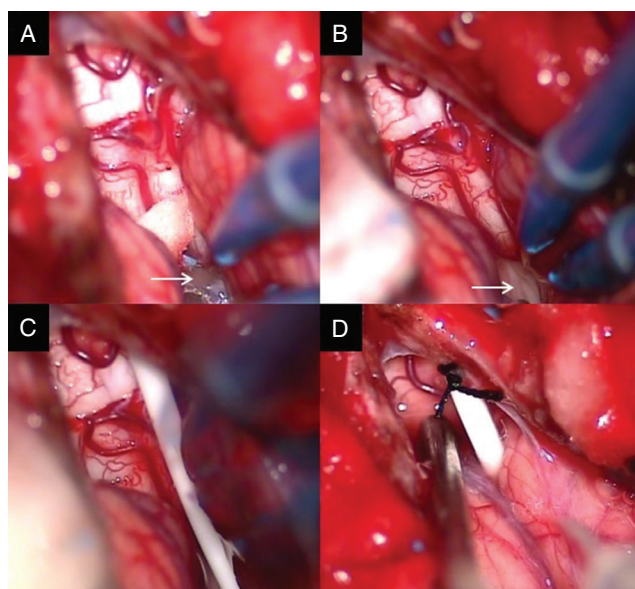| Age	Gender	Complaints	Neurological examination	Tetraventricular hydrocephalus	Syringomyelia	Chiari malformation
41	M	Head and neck pain	Normal	+	+	+
45	F	Numbness in arms, defect in discrimination between hot and cold, difficulty in swallowing	Left 11th and 12th cranial nerve deficiency, reduced GAG reflex, quadriparesis, hyperactive reflexes bilaterally ankle clonus	+	+	+
33	F	Headache	Normal	+	+	+
21	M	Head and neck pain, weakness in the hands, urinary incontinence	Upper extremity paresis: proximal muscle group is 4/5, distal group is 2/5, hyperactive reflexes, bilaterally Hoffmann's sign	+	+	+

F: female, M: male.

Fig. 1   Cranial computed tomography shows fourth ventriculostomy with a catheter (white arrows).


Fig. 2   Peroperative images. A: Occluded foramen of Magendie with transparent membrane (white arrow), B: Excised membrane and inside the fourth ventricle (white arrow), C: The catheter connecting the fourth ventricle to the cervical subarachnoid space, D: The catheter sutured to the arachnoid.

indicated upper extremity paresis and hyperactive reflexes, bilaterally Hoffmann's sign. The proximal muscle group was 4/5, and distal group was 2/5.

All the patients underwent MRI with morphological sequences. None of the patients had history of any known causes of hydrocephalus such as infection, hemorrhage, or head injury. Chiari Type I malformation, tetraventricular hydrocephalus, and syringomyelia were presented in all cases. Posterior fossa decompression and exploration constituted the treatment. All of these patients were operated while in prone position. Midline suboccipital craniectomy and C1 laminectomy were performed. After opening the dura, the cerebellar tonsils were separated. A transparent membrane had occluded the foramen of Magendie without evidence of inflammation. No CSF outflow was seen. The membranes were excised. After this excision, inside fourth ventricle and

passage of the foramen were seen clearly, and CSF drainage was achieved. One side of the catheter was inserted toward the fourth ventricle and connected with the cervical subarachnoid space (Fig. 1). The catheter was sutured to the arachnoid to prevent its migration (Fig. 2). With the same approach, Chiari malformation, syringomyelia, and hydrocephalus treatment were performed in the same session.

Results

The postoperative period was uneventful in all the patients. Neurological status of all the patients improved. Tetraventricular hydrocephalus and syrinx were reduced in the control cranial MRI. The postoperative imaging was performed at early CT scan and MRI scan, followed by controls in 3 months to 6 months. Follow-up period ranged between 1 year and 3 years. No complications such as
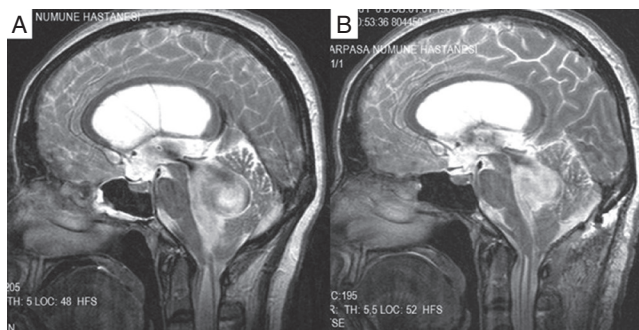
Fig. 3 T₂-weighted sagittal magnetic resonance imaging shows dilated fourth and lateral ventricle and syringomyelia preoperatively (A), reduced sizes of the ventricles and syringomyelia postoperatively (B).
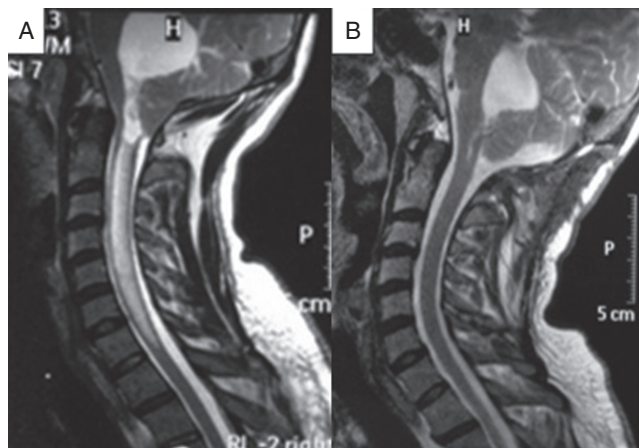
Fig. 4 T₂-weighted sagittal magnetic resonance imaging shows dilated fourth ventricle and syringomyelia preoperatively (A), reduced sizes of the fourth ventricles and syringomyelia postoperatively (B).

infection and catheter migration were observed during the follow-up period. Compared to the preoperative images, the sizes of the ventricles and syringomyelia were reduced in the postoperative images, and postoperative MRI findings demonstrated the dramatic effectiveness of the surgical procedure both in terms of ventricular size, CSF flow characterization, and reduction of the syringomyelia (Figs. 3, 4). A total disappearance of the tonsillar displacement and reduction in the syrinx size were observed in all the patients with syringomyelia.

Discussion

Idiopathic stenosis or closure of the foramen of Magendie is a rare cause of obstructive hydrocephalus.[7] Hydrocephalus resulting from FVOO is commonly observed to have a postinfectious or posthemorrhagic etiology in infants and young children although the secondary causes of obstruction are more common.[4] In adults, the occlusion is rather acquired than congenital, linked to infection, head trauma, intraventricular hemorrhage, tumors, or Chiari malformation.[6] The timing of clinical manifestations may vary and the ages of patients range from infants to adults. The patients presented with clinical sign of increased intracranial

hypertension and rhomboid fossa compression, cerebellar syndrome, vertebrobasilar insufficiency, nonspecific intermittent symptoms, and signs of normal pressure hydrocephalus.[2,3,7] Two of our patients complained of simple headache and neck pain with normal neurologic examination. In spite of these findings, the other two patients had severe pathological neurologic findings; tetraparesis, and carnial nerve involvement.

The diagnosis of hydrocephalus due to FVOO can be suspected in the face of an imaging study that shows tetraventricular hydrocephalus predominating in the fourth ventricle dilatation with dilated lateral recess. The study of the CSF flow shows a patent and enlarged cerebral aquaduct in which CSF flow is increased and the fourth ventricle outflow is obstructed. The foramen of Magendie shows no measurable flows. In Chiari and Dandy–Walker malformations, the cerebellar changes may help with the diagnosis. Although due to the dilatation of the fourth ventricle, it may be a downward displacement of the tonsils without true herniation.[2,3,5,6,8] In our group, all patients had Chiari Type I malformation, tetraventricular dilatation, and downward displacement of the posterior fossa structures and wide-spread syringomyelia.

Therefore, in cases of pronounced dilatation of the fourth ventricle with an increase in size of the lateral recesses and no history of inflammation or hemorrhage, the diagnosis of atresia of the foramina of Luschka and Magendie is most probable.[2,3,5,6,8] High resolution MRI may directly visualize the membrane of the foramen of Magendie. The membrane may be thick and strong or transparent or semidiaphanous.[6,9] A late symptomatic presentation is supposedly explained by a late decompensation of the semipermeable property of these congenital or acquired membranes.[4,5,8] In accordance with this theory, the age range of our patients was between 21 years and 44 years.

Various approaches have been described for the treatment of hydrocephalus due to FVOO such as ventriculoperitoneal or ventriculoatrial shunts, ETV, endoscopic fourth ventriculostomy, a suboccipital craniectomy with simply opening of the obstructing membrane or placing a catheter to maintain its patency toward the cisterna magna or subarachnoid space of the anterolateral to the upper cervical spinal cord.[1,2,7] Prior to technological advances, it was managed by shunts. Shunt operation presents unwanted complications such as obstructions, malfunctions, infections, visceral injuries, subdural hematomas or hygromas, slit ventricle syndrome, and isolated fourth ventricle.[1,7]

The development of endoscopic techniques has led to ETV and fourth ventriculostomy as the treatment of choice for these patients.[2] An endoscopic technique is a less physiological approach but is effective and preferable to CSF shunting for the treatment of obstructive hydrocephalus caused by stenosis of the cerebral aquaduct.[3] However, some distortions may prevent a safe ETV procedure and unique ventricular anatomy may contraindicate ETV.[2] Gianetti et al. described a new technique which they called endoscopic fourth ventriculostomy.[2] The technique involves opening the lateral and middle foramina of the fourth ventricle. They indicate that their technique has a number of advantages: (1) it allows

the opening of all three ventricular exits, (2) it is not dependent on the anatomy and dilatation of the supratentorial ventricles, and (3) it provides better control of possible bleedings and decreases the risk of injury to brain structures.

Longatti et al.[7] reported endoscopic opening of Magendie by transaquaductal. They pointed that the endoscopic opening has specific risks such as potential damage to the PICA hidden by thick membrane and the compression of the calamus scriptorius structures by the Fogarty balloon. Another complication could be restenosis caused by postoperative arachnoiditis.[6] However, some authors reported that the cases who performed fourth ventriculostomy did not require the placement of a catheter to maintain its patency.[4,5,10]

Posterior fossa craniotomy and membrane excision are still advocated by some.[10,11] Suboccipital craniectomy is an invasive procedure; suboccipital craniectomy with microsurgical exploration of the foramen of Magendie and excision of the obstructing membranes with or without a catheter to maintain the communication between the ventricular cavity and the subarachnoid space is a more physiological approach.[7]

Early in 1962, a new modality of fourth ventriculocisternostomy using a polyethylene catheter was described. Reopened foramen of Magendie maintained patency with a catheter installed, and thus, the original pathway and CSF circulation were restored.[11] As a result, the physiological functions of the intracranial pressure are maintained by the normal CSF circulatory mechanisms.[1] The direct catheter used this way has obvious advantages over other shunt systems. In fourth ventriculostomy, the catheter's structure is simple; its length is short, caliber is wide, obstruction and complications are rare.[1] However, ventriculosubarachnoid shunt is a foreign material that can be responsible for subsequent complications such as infection or especially catheter migration.[3]

In our study group, different from the earlier cases, Chiari Type I malformation and syringomyelia were present. Our aim was to solve all the pathologies such as Chiari malformation, hydrocephalus, and syringomyelia in one approach. Thus, the treatment consisted of posterior fossa decompression and exploration. All of the patients were treated with suboccipital craniectomy and C1 laminectomy with excision of the membrane obstructing the foramen of Magendie. Fourth ventriculostomy with cathetering from fourth ventricle to upper cervical subarachnoid space was performed.

The sizes of the ventricles and syringomyelia were reduced in the postoperative period. No complications such as infection and catheter migration were observed during the follow-up period.

The outcome of our treatment was satisfactory, but the number of patients and follow-up period were limited. Treatment with fourth ventriculostomy using a catheter from fourth ventricle to upper cervical subarachnoid space could be a treatment of choice in cases with hydrocephalus caused by occlusion of the foramina of Magendie associated with Chiari Type I malformation and syringomyelia.

Conflicts of Interest Disclosure

The authors declare that they have no financial or other conflicts of interest in relation to this research and its publication.

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