Case Report

Delayed Fourth Ventricle Outlet Obstruction after Fourth Ventricle Tumor Removal Successfully Treated with Endoscopic Third Ventriculostomy in a Pediatric Patient

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ARTICLE INFO

ABSTRACT

Introduction: Fourth ventricle outlet obstruction (FVOO) at the level of Magendie's and Luschka's foramina is a rare cause of non-communicating hydrocephalus.

Case Report: We present a case of a 15-year-old woman successfully operated on a fourth ventricle WHO grade 1 pilocytic astrocytoma developing a delayed FVOO five months after surgery, when the patient experienced progressive headache, nausea and gate disturbances. Magnetic resonance imaging (MRI) study showed a tetra-ventricular hydrocephalus, with enlargement and bulging of both lateral recesses and Luschka's foramina. An endoscopic third ventriculostomy (ETV) was successfully performed. Postoperative computed tomography (CT) and MRI studies showed a significant improvement of the hydrocephalus.

Conclusion: FVOO is a rare cause of hydrocephalus. Posterior fossa and fourth ventricle microsurgical procedures can produce a delayed FVOO leading to an unexpected deterioration of the clinical status of the patient. The ETV is an effective and safe procedure to treat this unusual condition.

Introduction

Fourth ventricle outlet obstruction (FVOO) due to the obstruction of the Magendie's and Luschka's foramina, is a rare cause of non-communicating hydrocephalus. Very few cases have been reported on the literature, most of them in adult population [1-9]. This condition produces a disproportionate dilatation of the fourth ventricle and subsequently a forward displacement of the forebrain that can displace the basilar artery cranially. Progressively the perimesencephalic cisterns disappear, as the ventricular system gradually dilates. Increased cerebrospinal fluid (CSF) pressure at the fourth ventricle can eventually develop spinal cord's central canal dilatation, causing secondary hydromyelia [1]. Endoscopic third ventriculostomy (ETV) may effectively treat this rare entity. We report a pediatric case of hydrocephalus due to post-surgical obstruction of the Magendie's and Luschka's foramina, successfully treated by ETV.

Case Report

A 15-year-old woman underwent microsurgery at our institution on a fourth ventricle tumor through a telovelar approach, with uneventful gross total removal. Histopathological diagnosis was WHO grade 1

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pilocytic astrocytoma. Postoperative CT scan showed no postoperative complications, nor ventricular dilatation (Figure 1). The patient was discharged without neurological deficit and routine surveillance MRI was scheduled six months later. However, five months after surgery, the patient developed headache and mild intracranial hypertension symptoms. Two weeks later experienced gait instability and increased headache causing morning vomiting, so was urgently admitted in our department. MRI showed hydrocephalus, more eminent at the fourth ventricle and its lateral recesses (Figure 2).
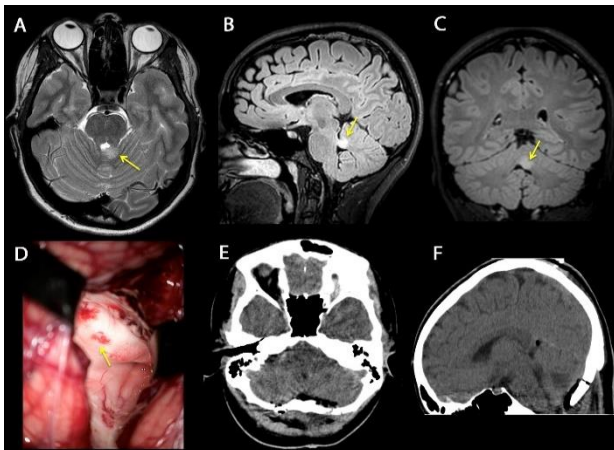

Figure 1: A-C) Preoperative T2 weighted MRI images in axial, sagittal and coronal planes showing a tumor in the left superior cerebellar peduncle; **D)** Intraoperative image from the microsurgical removal of the lesion; **E & F)** Postoperative CT scan in axial and sagittal planes showing no complications.
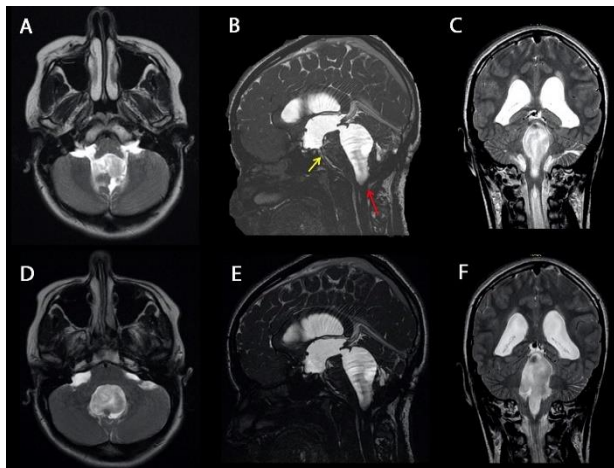

Figure 2: Pre ETV MRI study. **A & D)** Axial T2 MRI: Hydrocephalus affecting mainly the fourth ventricle and its lateral recesses; **B & E)** Sagittal T2 and FIESTA MRI: Perimesencephalic cisterns were barely patent and the forebrain was displaced anteriorly, with cranial displacement of the top of the basilar artery (yellow arrow) and secondary tonsillar descent (red arrow); **C & F)** CoronalT2 and FIESTA MRI: Dilatation of supratentorial ventricular system, fourth ventricle and lateral recesses. Hydromyelia and edema in the cervical spinal cord.

Perimesencephalic cisterns were barely patent and there was a displacement of the midbrain anteriorly and cranial displacement of the basilar artery's top. MRI also showed a secondary 16 mm tonsillar

herniation through the foramen magnum and funnel like fourth ventricle enlargement into the central canal of the cervical spinal cord, suggesting hydromyelia. Cine MRI showed patency of the sylvian aqueduct. Finally, gadolinium T1MRI showed no enhancement, ruling out any possibility of tumor remnant, recurrence, seeding, scar or infection. An ETV with a rigid endoscope (Aesculap Inc, Pennsylvania, USA) was performed through a right burr hole 0.5 cm medial to the Kocher's point. The third ventricle was accessed, the imprint of the basilar artery anterior to the mammillary bodies was visualized and ETV was performed. The fourth ventricle was not explored. Surgical procedure and postoperative recovery were uneventful. The patient improved after 48 hours and was discharged after demonstrating a significant reduction of the ventricular size in the CT scan (0.26 Evans ratio vs 0.40 preoperatively). Six-month postoperative MRI showed improvement of hydrocephalus and complete remission of tonsillar herniation and hydromyelia. The anatomy of the brainstem, ventricular system and cervical spinal cord was completely restored. After 3 years the patient remains asymptomatic with no imaging changes (Figure 3).
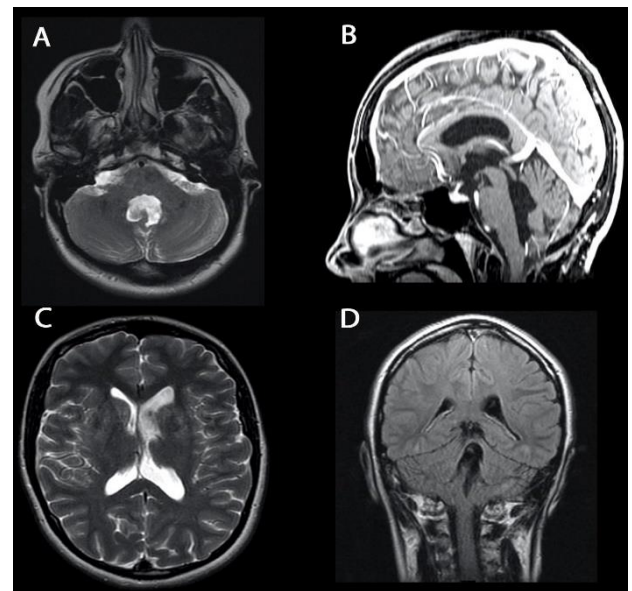

Figure 3: Post ETV MRI study showing the improvement of the hydrocephalus; **B)** Sagittal T1GD MRI: complete remission of the cervical spinal cords hydromyelia and tonsillar herniation. The anatomy of the brainstem, ventricular system and cervical spinal cord has been completely restored. **A & C)** T2 weighted MRI; **D)** Flair MRI.

Discussion

FVOO unrelated to inflammatory processes, Chiari malformation, Dandy-Walker and other malformations with abnormal posterior fossa anatomy, is a rare cause of hydrocephalus. The occlusion of Magendie's and Luschka's foramina by membranes in the absence of arachnoiditis is related to idiopathic, congenital or acquired causes [2-10]. Carpentier *et al.* describes three types of Luschka's obstruction. The first two, so-called dysgenesis and agenesis, should be idiopathic or congenital [2]. To explain the usually late onset of FVOO symptoms in adults, the authors hypothesize that in Luschka's dysgenesis there is a valve-like obstruction leading to an intermittent CSF flow. In Luschka's agenesis, obstruction is not complete and a punctiform aperture slows CSF flow,

but doesn't completely stop it. This balanced situation can change eventually if the communication suddenly closes and thus obstructs CSF flow. This would explain why some patients have a chronic insidious course and others an acute or subacute deterioration. Gianetti *et al.* describes fine translucent membranes with minute perforations occluding the foramina in a FVOO case operated through fourth ventriculostomy [3].

Longatti *et al.* describes different types of velar blockage of Magendie's foramen; closed Luskchka's foramen by membranes; engorged lateral recesses and inflammatory fourth ventricle's plexi arachnoiditis [4]. The third type of FVOO described by Carpentier, named Luschka's arachnoiditis, would be acquired and secondary to local inflammatory changes. A net-like membrane would lead to poor and turbulent CSF flow. The etiology should be secondary to arachnoiditis and scar tissue in the posterior fossa's cisterns due to haemorrhage or infection [1, 2, 7]. In fact, some patients with true FVOO often recall old subtle traumatic or inflammatory events [4]. However, most patients develop hydrocephalus in the acute or subacute stages in these situations, with rapid clinical deterioration. Patients treated in these early stages do not develop typical features of FVOO in MRI studies. Our patient has the antecedent of fourth ventricle microsurgery.

Although the procedure and clinical course were uneventful, we assume that surgery was the cause of FVOO. Moreover, the MRI revealed all anatomical features observed in idiopathic FVOO. Finally, the clinical course and the complete response to ETV are congruent with FVOO syndrome. These facts suggest that our patient had an idiopathic FVOO and surgery triggered a subacute obstruction [2]. CT scan can diagnose hydrocephalus, secondary tonsillar herniation and hydromyelia [1, 11]. However, MRI is the gold standard for FVOO diagnosis, with FIESTA, T1, T2 and FLAIR sequences. Cine MRI can help locating the exact level of FVOO. However, it seems to have less sensibility when compared to the diagnosis of aqueduct stenosis [1-3, 7]. Visualization of the membranes causing FVOO can be achieved with T1-T2 MRI and ventriculography [8].

Classical treatment of FVOO was based on ventriculoperitoneal shunt. Some cases have been treated by direct surgical exploration and microsurgical opening of the membranes [5, 8, 10]. Others by endoscopic third or fourth ventriculostomy and Magendie's foramina opening [4-6, 10]. However, ETV has demonstrated to be safe an effective for treating FVOO hydrocephalus. It is a low risk procedure with low rate of recurrences and complications [1, 2, 4-9]. Technically it is relevant to account the upward displacement of the basilar artery's top, imprinting into the third ventricle's floor, that reduces the available space for ETV [10]. Recurrence has been described in post hemorrhagic and post infectious patients, probably due to extensive adhesions in the basal cisterns, and also in infants younger than six months [5]. Exploration of the basal cisterns during ETV might have relevance in assessing the risk of recurrence [5]. The most relevant complication is the development of isolated fourth ventricle, which can force revisions, need for a second connection catheter or complex endoscopic procedures [12-14]. ETV could allow exploration and opening of the fourth ventricle outlets if navigation through the aqueduct was feasible using a rigid or a flexible endoscope) [4, 5, 13]. If the endoscopic procedure allowed opening any of the fourth ventricle foramina along with the ETV, the chances of an isolated fourth ventricle would reduce [15].

Conclusion

We describe a pediatric case of symptomatic idiopathic FVOO successfully treated with ETV developed five months after removing a fourth ventricle glioma. ETV is a safe technique for treating this unusual condition but it is necessary to account the technical difficulty due to the distortion of the ventricular system and the limited space available in the premammillary membrane. When feasible, it is recommended to associate the opening of the fourth ventricle outlets. It is also mandatory to carry out a long-term clinical and imaging surveillance, to rule out the appearance of secondary complications, particularly an isolated fourth ventricle.

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