Trapped fourth ventricle
Case report and narrative review

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Abstract

A trapped fourth ventricle is a rare complication following successful shunting of cerebrospinal fluid from the lateral ventricles in patients with acquired post-haemorrhagic or post-infectious hydrocephalus. Inadequate drainage combined with ongoing cerebrospinal fluid production leads to progressive enlargement of the fourth ventricle with compression of surrounding anatomical structures. We present the case of a four-month-old girl born extremely prematurely at 25 weeks with bilateral intraventricular haemorrhage whose management was complicated by the rare combination of trapped fourth ventricle and subsequent multiloculated posterior fossa hydrocephalus, presenting a neurosurgical dilemma.

Introduction

In 2020, approximately 7% of newborns in Flanders were born premature (gestational age less than 37 weeks), of which 1% was born very preterm (gestational age between 28 and 32 weeks) or extremely preterm (gestational age less than 28 weeks) (1). Despite considerable advances in perinatal and neonatal care, complications still occur, including the development of intraventricular haemorrhage (IVH) in 20 to 25% of very low birth weight (<1500 grams) infants, of which 15% is complicated by a parenchymal haemorrhagic infarction (PHI). In 25%, IVH is followed by progressive post-haemorrhagic ventricular dilatation (PHVD) of which 35% will require surgery (2). The management of these conditions is a major challenge for families, neonatologists and the health care system as a whole.

We present the case of a four-month-old girl, born extremely prematurely at 25 weeks with progressive PHVD and lateral ventricular shunting, complicated by the development of a trapped fourth ventricle (TFV) and subsequent multiloculated posterior fossa hydrocephalus after multiple neurosurgical interventions, posing a management dilemma. A comprehensive review of the literature on the current understanding of the pathophysiology, diagnosis and treatment of a TFV is provided.

Case report

The patient was born extremely prematurely at 25 weeks and 2 days with a birth weight of 740 grams and quickly presented with respiratory distress syndrome requiring intubation for surfactant replacement therapy. She had multiple other complications, including left-sided pneumothorax, prolonged intubation evolving into mild bronchopulmonary dysplasia, late-onset sepsis, spontaneous bowel perforation requiring a temporary double-barrelled ileostomy and retinopathy of prematurity treated with intravitreal injection and laser therapy. In addition, there were significant neurological complications, which prompted the writing of this case report. On the second day postpartum, a bilateral grade 3 IVH of germinal matrix origin with venous infarction in the left terminal vein area occurred. This was complicated by the development of bilateral PHVD, and the formation of left-sided porencephalic cysts (Figure 1). A Rickham® ventriculostomy reservoir was initially placed in the right lateral ventricle to evacuate CSF. Three weeks later, ventriculostomy-associated ventriculitis was suspected and treated with antibiotics. One month after the initial operation, a second Rickham® reservoir had to be placed in the left ventricle due to progressive dilatation of the left ventricle. Serial transcranial ultrasound scans revealed the development of a TFV (35 mm in height and 20 mm in anteroposterior diameter) at the age of four months.

Figure 1: brain MRI showing bilateral post-haemorrhagic ventricular dilatation and formation of left sided porencephalic cysts.

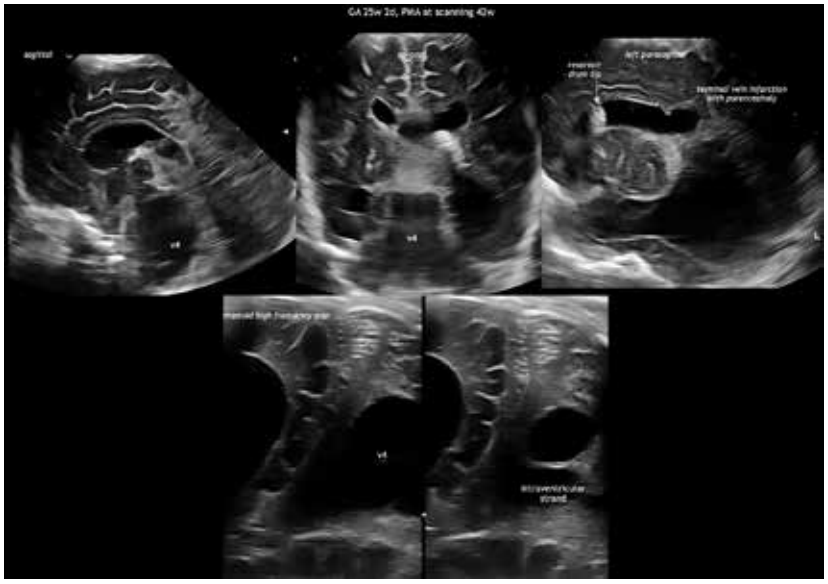

Figure 2: brain MRI showing dislocation of the Ommaya® catheter and septation dividing the fourth ventricle into two isolated cysts with consequent brainstem compression.

This appeared to progress rapidly over two days with significant mass effect on the cerebellum and brainstem, leading to clinical deterioration with poor feeding, somnolence and bradycardia. Urgent decompression was achieved by performing an endoscopic fourth ventriculostomy with insertion of another Rickham® reservoir. The operation was successful and in a second stage, the three ventricular reservoirs were replaced by two ventriculoperitoneal shunts (VPS): a Y-shaped branched VPS to divert CSF from the lateral ventricles and another VPS to drain the fourth ventricle. Clinical improvement was observed and ventricular size remained stable on repeated ultrasound scans. On postoperative day 8, recurrent entrapment of the fourth ventricle occurred as consequence of Escherichia coli ventriculitis. Both VPS were immediately removed and replaced with three intraventricular Ommaya® reservoirs which provided adequate decompression.

However, since the last surgery, neurological function had not fully recovered and our patient had persistent axial hypotonia and poor feeding, corresponding to persistent dilatation of the fourth ventricle on ultrasound imaging. One month post-operatively, a brain MRI showed dislocation of the Ommaya® catheter and septation dividing the fourth ventricle into two isolated cysts with consequent brainstem compression (Figure 2). Further neurosurgical intervention was imminent and consisted of removal of the Ommaya® reservoirs and placement of new VPS: a Y-shaped branched VPS for the lateral ventricles and a second multi-perforated VPS for the fourth ventricle cysts. The MRI was repeated one week post-operatively and showed that the fourth ventricular cystic lesions had increased in number and size, with significant pressure on the brainstem and supratentorial extension towards the midbrain (Figure 2). There was an imminent risk of fatal brain herniation if left untreated, and it was decided to perform endoscopic fenestration of the posterior fossa cysts by bilateral ventriculostomy. The neurological status improved postoperatively, but MRI showed further expansion of the posterior fossa cysts with increased transtentorial herniation.

Following multidisciplinary consultation, it was agreed that our patient was unlikely to benefit from further surgical intervention given the increasing surgical complexity, extensive medical history and poor prognosis in terms of severe developmental delay. In consultation with the parents, palliative care was provided to minimise suffering. Twenty days later, aged 4 months, she died in hospital.

Informed consent was obtained from both parents to publish this case report.

Literature review and discussion

Pathophysiology

TFV, also known as 'isolated fourth ventricle', 'encysted fourth ventricle' and 'double compartment hydrocephalus', is a rare complication following lateral ventricle shunting in patients with acquired post-haemorrhagic or post-infectious hydrocephalus. TFV develops when the shunting system fails to drain the fourth ventricle (3-7).

The primary site of CSF production is the choroid plexus of the ventricles of the brain. CSF flows through the ventricular system from the lateral ventricles to the third ventricle through the foramen of Monro. From here it continues through the aqueduct of Sylvius to the fourth ventricle and then enters the subarachnoid space through the lateral apertures of Luschka and the median aperture of Magendie. Obstruction of the inlet (aqueduct of Sylvius) and three outlets (the paired lateral apertures of Luschka and the median aperture of Magendie) of the fourth ventricle isolates the fourth ventricle from the ventricular system and the subarachnoid space, resulting in TFV. The continued production of CSF causes progressive enlargement of the fourth ventricle with compression of surrounding anatomical structures (brainstem, cranial nerves, cerebellum, central canal of the spinal cord) and increased intracranial pressure (4-6).

TFV is most commonly secondary to the placement of a VPS for the treatment of congenital or acquired hydrocephalus. Dandy-Walker and Arnold-Chiari malformations are the main causes of congenital hydrocephalus associated with the development of TFV (8). Subarachnoid or intraventricular haemorrhage (IVH) and infections (bacterial/fungal meningitis/ventriculitis, VPS infection and cysticercosis) are the main causes of acquired hydrocephalus associated with the development of TFV (9-11). The incidence of TFV in shunted patients has been reported to be 2-3% (7).

The pathophysiological mechanisms of TFV secondary to acquired hydrocephalus are complex and multifactorial. The main mechanism is the onset of inflammation of the ependyma (ependymitis) and subarachnoid space (arachnoiditis) due to haemorrhage and/or infection, leading to fibrosis of the surrounding structures. When the ventricular system is decompressed following shunt placement, adhesions may form at the level of the ventricles and the cerebral aqueduct as a result of chronic ependymitis, leading to occlusion of the aqueduct. Impaired CSF absorption at the level of the arachnoid villi and obliteration of the foramina of Luschka and Magendie are consequences of chronic arachnoiditis. Blood products and cellular debris from haemorrhage and infection may contribute to obstruction of the cerebral aqueduct and foramina. Repeated revisions and manipulations contribute to the overall inflammation (3-6, 10-13). It has also been suggested that 'overdrainage' after shunt placement causes a reduction in supratentorial pressure relative to infratentorial pressure, resulting in upward displacement of midline cerebellar structures into the tentorial incisura. This results in a distortion of the cerebral aqueduct with impaired cerebrospinal fluid flow (3, 5, 11, 13).

In addition, the presence of cellular debris and chronic ependymitis may induce the formation of fibrous adhesions leading to intraventricular septations, creating one (uninoculated) or more (multiloculated) non-communicating fluid-filled compartments, known as multiloculated hydrocephalus, which may occur at different levels of the ventricular system. It is a dynamic disorder with the development of new septae due to hydrodynamic changes and new membrane formation (14, 15).

Diagnosis

TFV usually presents as a sudden clinical deterioration after an initial period of improvement following successful lateral ventricle shunting,

most commonly in patients with post-haemorrhagic or post-infectious hydrocephalus. The time between shunting of the lateral ventricles and the presentation of TFV ranges from 1 month to 12 years (7, 16, 17). In our case, the interval between placement of the first Rickham® ventriculostomy reservoir and diagnosis of TFV was 102 days. A wide variety of symptoms and signs are reported, all related to increased intracranial pressure and compression of surrounding anatomical structures, resembling posterior fossa syndrome. Clinical signs depend on the age of presentation and range from irritability, lethargy, apnoea, spontaneous bradycardic episodes, vomiting, poor feeding, full to bulging anterior fontanel and increasing head circumference in neonates and infants to headache, ataxia and growth failure in older children. Tonic seizures, progressive spasticity, dysconjugate eye movements and cranial nerve palsies (third and sixth cranial nerve palsies) have also been reported (3-7). Clinical diagnosis may be complicated by pre-existing neurological deficits such as developmental delay, cerebral palsy and epilepsy (6, 10). Delayed diagnosis can lead to severe neurological impairment and death (4, 18). Approximately 15% of patients are asymptomatic with an incidental finding of TFV on neuroimaging during follow-up, as was the case in our patient (13). Repeat cerebral ultrasound revealed TFV, which was asymptomatic at the time. However, it appeared to be rapidly progressing with significant mass effect on the cerebellum and brainstem visible on MRI imaging, which subsequently led to clinical deterioration with poor feeding, somnolence and bradycardia.

The problem of discrepant dilatation is often detected by cranial ultrasound and confirmed by magnetic resonance imaging (MRI). The diagnosis is made when an enlarged 'ballooned' fourth ventricle is seen, often accompanied by membranous occlusion of the cerebral aqueduct and compression of the surrounding structures with effacement of the cerebellar tissue, flattening of the posterior aspect of the brainstem and reduced CSF in the prepontine cistern caused by ventral displacement of the brainstem (7, 19). In addition, MRI can be used to differentiate between a TFV, shunt malfunction and cystic lesions. Treatment decisions are based on the size of the lateral ventricles (slit or wide), the length of the aquaeductal stenosis (short or long), compression of the brainstem and cerebellum, and herniation through the tentorial notch on preoperative imaging (16, 20).

Treatment

Asymptomatic TFV with or without brainstem compression is usually treated conservatively without surgery, but, in some cases, with progressive brainstem compression surgery is to be considered (13). Symptomatic TFV requires surgery (20). The optimal surgical treatment has not yet been determined because, for obvious reasons, randomised studies on this topic are not available. The available surgical options are fourth ventricular shunt procedures, endoscopic procedures and a suboccipital craniectomy with outlet fenestration (18, 20).

Fourth ventricular shunts (FVS) are considered the mainstay of treatment to drain the TFV. The catheter can be connected to the existing lateral ventricle shunt with a Y-connector or to a separate system. FVS has a high risk of complications, including shunt malfunction (obstruction, disconnection), infection, intra-cystic haemorrhage, reversible and rarely irreversible cranial nerve dysfunction, fourth ventricle flour injury and brainstem injury (18, 20). Approximately 40% require shunt revision within one year of initial placement (21). In our patient, it was decided to first perform an urgent life-saving decompression through endoscopic fourth ventriculostomy with placement of a Rickham® reservoir, which was later replaced by a ventriculoperitoneal shunt draining the fourth ventricle. To date, there are insufficient case reports to compare the risk of TFV for shunts and reservoirs. Both techniques by definition reduce the flow through the aqueduct and, in reducing the size of the third ventricle, may predispose to adhesions of denuded and infected ventricular walls. Reservoir puncture causes intermittent changes in ventricle size and cerebrospinal fluid flow, in contrast to contrary to the continuous effect of a shunt. This intermittent effect would not theoretically increase the risk of TFV. Shunt complications that were encountered in our case included ventriculoperitoneal shunt infection and ventriculitis, followed by improper CSF drainage due to shunt malfunction requiring shunt revision surgery.

Due to the high complication rate of FVS and advances in endoscopy, other treatment options are now available (22). Endoscopic procedures include aqueductoplasty with or without stent placement (trans frontal trans-third ventricle or suboccipital approach), third or lateral interventriculostomy with or without stent placement (translateral ventricle approach) and fenestration of the obstructed aqueduct with stent placement (trans foramen of Magendie approach). Aquaeductoplasty penetrates the membranous occlusion of the cerebral aqueduct, restoring communication and equalizing pressure between the infra- and supratentorial ventricular system (23). Due to the high risk of restenosis ranging from 39 to 73%, stent placement is recommended even if there is a history of infection (7, 16, 23, 24). The transfrontal trans-third ventricle approach is usually performed in the presence of ventriculomegaly, which is used to facilitate safe endoscopic navigation (20). If the lateral ventricles are well decompressed, the pre-existing VPS is externalised and the ventricles are gradually dilated, or a suboccipital approach is performed. It has been suggested in the to differentiate between short segment aquaeductal stenosis (< 5 mm on MRI) and long segment aqueductal stenosis (> 5 mm) to avoid stent complications, such as stent migration, infection, cranial nerve palsies (especially third and sixth cranial nerve palsies), Parinaud syndrome and brainstem injury. Short segment aqueductal stenosis is eligible for aqueductoplasty with stent placement and long segment aqueductal stenosis for FVS (20). A third interventriculostomy with or without stent placement may be considered in cases of cystic dilatation of the fourth ventricle with upward herniation through the tentorium and distorted ventricular anatomy with unidentifiable anatomical landmarks. The thinnest membranous barrier between the third and fourth ventricles is then perforated, with the risk of damaging the basilar artery and hypothalamus when perforating the third ventricular floor (22-25). Suboccipital craniectomy and microsurgical canalisation of the obstructed aqueduct and fenestration of the outlet membranes is considered in complicated patients or when other approaches have failed (19, 20).

The neurosurgical management of our case was particularly challenging due to the formation of multiple posterior fossa cysts secondary to intraventricular infection, defined as multiloculated hydrocephalus. The presence of compartmentalisation within the fourth ventricle system is extremely rare and may be the most difficult form of hydrocephalus to treat. There is a paucity of data on posterior fossa shunting in the neurosurgical literature, so there is no widely accepted surgical treatment. Cyst fenestration to restore communication between these isolated cystic compartments is the main strategy of treatment, yet the approach remains controversial. The strategy in our case was an initial shunt revision with insertion of a multi-perforated ventricular catheter. Unfortunately, this did not adequately drain the isolated posterior fossa cysts. Endoscopic surgery consisting was then performed with intraventricular septum fenestration, which was also unsuccessful. After enduring numerous shunt revisions with high morbidity, a multidisciplinary decision was made to offer palliative care.

Conclusion

Advances in perinatal medicine have contributed to significant improvements in the survival of preterm infants. However, a significant number of these infants still suffer from disabling and life-threatening health conditions. The development of a trapped fourth ventricle was a major setback for our patient, who had previously been successfully for PHVD with lateral ventricle shunting. TFV develops when the fourth ventricle becomes isolated due to the occlusion of the inlets and outlets. Continued CSF production with progressive enlargement of the fourth ventricle leads to compression of the surrounding anatomical structures and increased intracranial pressure. Delayed diagnosis can lead to severe neurological impairment and death. Treatment options depend on a number of factors and include fourth ventricle shunting or endoscopic procedures. Neurosurgical management in this case was particularly challenging due to the formation of multiple posterior fossa cysts secondary to intraventricular infection. Early diagnosis of ventriculitis including close monitoring with high-resolution magnetic resonance imaging to detect multiloculated transformation and early treatment are key to a better prognosis.

Conflict of interest

The authors report no conflict of interest.

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