Fourth ventricle hydrocephalus

Synonyms: Entrapped fourth ventricle; Isolated fourth ventricle.

Udayakumaran and Panikar reiterate that a Fourth ventricle hydrocephalus or trapped fourth ventricle (TFV) is a functional concept with imaging being at most only corroboratory [1].

In adults, Ferrer and de Notaris call this condition the functional trapped fourth ventricle because in none of there cases they have found physical obstruction of CSF flow [2].

Etiology

Fourth ventricle hydrocephalus, or a “trapped” fourth ventricle is a rare and uncommon entity which has been observed as a complication after intraventricular hemorrhage, infection/ meningitis or as a result of chronic shunt overdrainage after hydrocephalic shunting [3] [4] [5] [6].

A postinfectious occlusion of fourth ventricle outflow (foramen of Luschka and Magendie) and aqueduct of sylvius is the second most common cause for the development of trapped fourth ventricle [7].

This condition is caused by blockage of both the outlets (Foramen of Luschka, Foramen of Magendie) and the inlet of the fourth ventricle at the level of the aqueduct of Sylvius [8].

Progressive dilation of the fourth ventricle is due to continuing CSF production by the choroid plexus of the fourth ventricle within a closed space.

An increased cerebrospinal fluid (CSF) pressure within the fourth ventricle can lead secondary to the enlargement of the central canal in terms of communicating secondary syringomyelia. The exact pathophysiological mechanism of developing syringomyelia generally is not well established and remains yet controversial although several theories have been postulated [9].

In a follow-up study of 164 hydrocephalic children without tumors treated with ventriculoperitoneal shunts, 46 (28.0%) developed slit ventricle syndrome, 5 (3.0%) developed isolated fourth ventricles, and 4 (2.4%) developed isolated unilateral hydrocephalus. All of the patients with isolated unilateral hydrocephalus and 3 with isolated fourth ventricles had associated slit ventricles, 2 of whom had enlarged ventricles as double-compartment hydrocephalus. Reopening of the foramen of Monro or the aqueduct was achieved in one of the former and two of the latter cases with re-expansion of the slit ventricles. It is suggested that in some cases, the slit ventricle could be a causative factor in post-shunt isolated ventricle [10].

Clinical features

Such an entrapment may lead to clinical symptoms secondary to distortion of the brainstem and lower CNs. The clinical findings are mostly non localizing, even when there are obvious bulbar signs.

Diagnosis

Imaging may corroborate clinical findings but may not be diagnostic by itself.

The diagnostic and treatment dilemma is to differentiate between a “true” symptomatic TFV and other conditions associated with a large fourth ventricle. This dilemma is especially significant when one is attempting to identify those patients who may benefit from surgery, as opposed to those patients with a well-compensated process that simply have a similar clinical and a radiological picture of a large fourth ventricle.

Differential diagnosis

Posterior fossa cysts are usually divided into Dandy Walker malformations, posterior fossa arachnoid cysts, and isolated and/or trapped fourth ventricles.

Treatment

Treatment of the TFV remains a formidable challenge. However, prompt recognition and intervention may aid in the preservation of life and neurological function [11].

Treatment extends from placement of fourth ventriculoperitoneal shunt, endoscopic aqueductoplasty and interventriculostomy to open fenestration via suboccipital craniotomy [12) 13)].

During the last years the development of neuroendoscopy has dramatically changed the outcome of these patients.

see Fourth ventricular shunt.

Aqueductoplasty

see Aqueductoplasty

Complications

Frassanito et al., report an exceptional case of Descending transtentorial herniation (DTH) complicating the implant of a CSF shunting device in the trapped fourth ventricle of a 17-year-old boy in whom a second CSF shunting device had been implanted for neonatal posthemorrhagic and postinfectious hydrocephalus. The insidious clinical and radiological presentation of DTH, mimicking a malfunction of the supratentorial shunt, is documented. Ultimately, the treatment consisted of removal of the infratentorial shunt and endoscopic acqueductoplasty with stenting. The absence of supratentorial mass lesion and other described etiologies of DTH prompted the authors to speculate on the hydrodynamic pathogenesis of DTH in the present case [14].

Cranial nerve palsy is rarely seen after shunt placement in an isolated fourth ventricle. In the few reports of this complication, neuropathies are thought to be caused by catheter injury to the brainstem nuclei either during the initial cannulations or after shrinkage of the fourth ventricle. The authors treated a child who suffered from delayed, progressive palsies of the sixth, seventh, 10th, and 12th cranial nerves several weeks after undergoing ventriculoperitoneal shunt placement in the fourth ventricle. Magnetic resonance imaging revealed the catheter tip to be placed well away from the ventricular floor but the brainstem had severely shifted backward, suggesting that the pathogenesis of the neuropathies was traction on the affected cranial nerves. The authors postulated that the siphoning effect of the shunt caused rapid collapse of the fourth ventricle and while the cerebellar hemispheres were tented back by adhesions to the dura, the brainstem became the only mobile component in response to the suction forces. Neurological recovery occurred after surgical opening of the closed fourth ventricle and lysis of the basal cistern adhesions, which restored moderate ventricular volume and released the brainstem to its normal position [15].

Case series

2016

Pomeraniec et al retrospectively reviewed 8 consecutive cases involving pediatric patients with TFV following VP shunting for IVH due to prematurity between 2003 and 2012. The patients ranged in gestational age from 23.0 to 32.0 weeks, with an average age at first shunting procedure of 6.1 weeks (range 3.1-12.7 weeks). Three patients were managed with surgery. Patients received long-term radiographic (mean 7.1 years; range 3.4-12.2 years) and clinical (mean 7.8 years; range 4.6-12.2 years) follow-up.

The frequency of TFV following VP shunting for neonatal posthemorrhagic hydrocephalus was found to be 15.4%. Three (37.5%) patients presented with symptoms of posterior fossa compression and were treated surgically. All of these patients showed signs of radiographic improvement with stable or improved clinical examinations during postoperative follow-up. Of the 5 patients treated conservatively, 80% experienced stable ventricular size and 1 patient experienced a slight increase (3 mm) on imaging. All of the nonsurgical patients showed stable to improved clinical examinations over the follow-up period.

The frequency of TFV among premature IVH patients is relatively high. Most patients with TFV are asymptomatic at presentation and can be managed without surgery. Symptomatic patients may be treated surgically for decompression of the fourth ventricle [16].

2012

Of 1044 aneurysms treated, 3 patients were identified who required fourth ventricular shunting, for the treatment of the isolated ventricle. All 3 patients underwent microsurgical clip obliteration of their aneurysms and had subsequent frontal approach ventriculoperitoneal cerebrospinal fluid diversion. These patients had no evidence of infection of the cerebrospinal fluid as measured by serial cultures. Subsequently, all 3 patients presented in a delayed fashion with symptoms attributable to a dilated fourth ventricle and syringomyelia or syringobulbia. Either exploration or percutaneous tapping confirmed the function of the supratentorial shunt. These patients then underwent fourth

ventriculoperitoneal cerebrospinal fluid diversion by the use of a low-pressure shunt system. The symptoms attributable to the isolated fourth ventricle resolved rapidly in all 3 patients after shunting. This clinical improvement correlated with the fourth ventricular size.

Isolated fourth ventricle, in an adult, is a rare phenomenon associated with intracranial posterior circulation aneurysm rupture treated with microsurgical clip obliteration. Fourth ventriculoperitoneal cerebrospinal fluid diversion is effective at resolving the symptoms attributed to the trapped ventricle and associated syrinx [17].

2011

Between February 1998 and February 2007, 12 children were treated for TFV in Dana Children's Hospital by posterior fossa craniotomy/craniectomy and opening of the TFV into the spinal subarachnoid space. The authors performed a retrospective analysis of relevant data, including pre- and postoperative clinical characteristics, surgical management, and outcome.

Thirteen fenestrations of trapped fourth ventricles (FTFVs) were performed in 12 patients. In 6 patients with prominent arachnoid thickening, a stent was left from the opened fourth ventricle into the spinal subarachnoid space. One patient underwent a second FTFV 21 months after the initial procedure. No perioperative complications were encountered. All 12 patients (100%) showed clinical improvement after FTFV. Radiological improvement was seen in only 9 (75%) of the 12 cases. The follow-up period ranged from 2 to 9.5 years (mean 6.11 ± 2.3 years) after FTFV [18].

2004

Fritsch et al., retrospectively reviewed the medical histories of 18 patients with an isolated fourth ventricle. Surgical procedures included endoscopic aqueductoplasty, endoscopic aqueductoplasty with a stent, endoscopic interventriculostomy (lateral ventricle or third ventricle to fourth ventricle), and endoscopic interventriculostomy with a stent. Operations were performed between July 1997 and June 2002. The mean age of the patients at the time of surgery was 3 years. The mean follow-up was 29 months. All patients had a supratentorial ventriculoperitoneal shunt.

Clinical symptoms (impairment of consciousness, tetraparesis, and ataxia) improved in all patients. Reduction of the size of the fourth ventricle was observed in all patients. Seven patients required reoperation because of restenosis (39% revision rate). Restenosis occurred between 2 weeks and 7 months after surgery (average, 3 mo). Four patients underwent reoperation with stent placement, and three patients underwent reaqueductoplasty. We had the following complications: one infection, one asymptomatic subdural hygroma, one transient oculomotor paresis, and one permanent oculomotor paresis (4 [22%] of 18 patients).

The significant failure rate of fourth ventricle shunts has led to the development of alternative treatment methods. Endoscopic aqueductoplasty or interventriculostomy presents an effective, minimally invasive, and safe procedure for the treatment of isolated fourth ventricle in pediatric patients. Compared with suboccipital craniotomy and microsurgical fenestration, endoscopic aqueductoplasty is less invasive, and compared with fourth ventricle shunts, it is more reliable and effective [19].

1997

Between January 1986 and December 1995, Eder et al., treated 292 children younger than 16 years for hydrocephalus: 7 (2.4%) developed an isolated IV ventricle, and 5 of these were symptomatic with posterior fossa signs. These 5 patients required posterior fossa shunting, after which their neurological status improved. However, 1 week and 6 weeks after surgery, respectively, 2 patients developed new cranial nerve deficits related to a slit-like IV ventricle with secondary irritation of the brain stem by the IV ventricular catheter. Shortening the catheter and replacing the valve eliminated the cranial nerve palsies, implying that these complications were not caused by direct injury of the brain stem during placement of the shunt. Alternative surgical techniques and the use of different (flow-regulating) valves may avoid such complications [20].

1980

Isolated fourth ventricles were diagnosed by computed tomography (CT) in 16 children in a 3 year period. They all had shunting of the lateral ventricles for hydrocephalus, and all needed subsequent shunt revisions. Seven patients without signs of raised intracranial pressure clinically had new posterior fossa signs at different intervals after lateral ventricular shunting. The clinical findings in the other nine patients were much less specific and in some cases the isolated fourth ventricle was an incidental finding. CT is essential for the diagnosis. The isolated fourth ventricle needs to be differentiated from posterior fossa cysts and cystic tumors. Shunting of the fourth ventricle improved the clinical condition in six of 14 children [21].

1978

Signs of cerebellar dysfunction combined with signs suggestive of shunt malfunction developed in three children with obstructive hydrocephalus. Shunt function was normal. In all cases, the cerebellar signs persisted and computerized tomography scans revealed enlargement of the fourth ventricle. Shunting of the fourth ventricle returned the patients to normal function [22].

Case reports

2016

Trapped Fourth Ventricle With Vasogenic Edema [23].

2013

A 28-year-old female who had previously undergone treatment of intracerebral hemorrhage and meningitis developed a hydrocephalus with TFV. After 3 years she developed disturbance of walking and coordination. Cranial-CT revealed an enlargement of the shunted fourth ventricle as a result of shunt dysfunction. Furthermore a cervical syringomyelia developed. The patient underwent a revision of a failed fourth ventriculo-peritoneal shunt. Postoperatively, syringomyelia resolved within 6 months and the associated neurological deficits improved significantly. An insufficiency of cerebrospinal fluid

draining among patients with TFV can be associated with communicating syringomyelia. An early detection and treatment seems important on resolving syringomyelia and avoiding permanent neurological deficits. Ventriculo-peritoneal shunt in trapped fourth ventricles can resolve a secondary syringomyelia [24].

2009

A 4-year-old girl with a ventriculoperitoneal shunt presented with complaints of ataxia and altered consciousness. These symptoms were subacute at onset and progressive in nature.

Radiological evaluation revealed a trapped fourth ventricle with brainstem compression, associated with abnormal diffuse diencephalic signal changes compatible with edema. The entrapment was managed by foramen magnum decompression, resulting in complete symptom resolution and improvement in the abnormal magnetic resonance findings.

While trapped fourth ventricle is a well-described entity, we could not find a similar reported case where such an acute clinical syndrome was associated with such a distinct radiological picture [25].

A 20-year-old man with complex hydrocephalus and trapped fourth ventricle underwent a suboccipital placement of a VP shunt. Postprocedure patient developed double vision. Magnetic resonance imaging showed that the catheter was penetrating the dorsal brainstem at the level of the pontomedullary junction. Patient was referred to our Neuroendoscopic Clinic. Physical exam demonstrated pure right VI cranial nerve palsy. Patient underwent flexible endoscopic exploration of the ventricular system. Some of the endoscopic findings were severe aqueductal stenosis and brainstem injury from the catheter. Aqueductoplasty, transaqueductal approach into the fourth ventricle, and endoscopic repositioning of the catheter were some of the procedures performed. Patient recovered full neurological function. The combination of endoscopic exploration and shunt is a good alternative for patients with complex hydrocephalus. A transaqueductal approach to the fourth ventricle with flexible scope is an alternative for fourth ventricle pathology [26].

2005

Cranial nerve palsy is rarely seen after shunt placement in an isolated fourth ventricle. In the few reports of this complication, neuropathies are thought to be caused by catheter injury to the brainstem nuclei either during the initial cannulations or after shrinkage of the fourth ventricle. The authors treated a child who suffered from delayed, progressive palsies of the sixth, seventh, 10th, and 12th cranial nerves several weeks after undergoing ventriculoperitoneal shunt placement in the fourth ventricle. Magnetic resonance imaging revealed the catheter tip to be placed well away from the ventricular floor but the brainstem had severely shifted backward, suggesting that the pathogenesis of the neuropathies was traction on the affected cranial nerves. The authors postulated that the siphoning effect of the shunt caused rapid collapse of the fourth ventricle and while the cerebellar hemispheres were tented back by adhesions to the dura, the brainstem became the only mobile component in response to the suction forces. Neurological recovery occurred after surgical opening of the closed fourth ventricle and lysis of the basal cistern adhesions, which restored moderate ventricular volume and released the brainstem to its normal position [27].

1975

The first reported case was a patient with cysticercosis meningitis and communicating hydrocephalus in whom signs of a posterior fossa mass developed a few months after shunting of the lateral ventricles . Air studies and posterior fossa exploration demonstrated an encysted fourth ventricle due to occlusion of its outlets as well as of the aqueduct [28].

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