Endoscopic treatment of cavum vergae cyst: case report and review

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SUMMARY: Gazioğlu N, Kafadar AM, Abuzayed B. Endoscopic treatment of cavum vergae cyst: case report and review. Turk J Pediatr 2011; 53: 590-594.

A 3.5-year-old boy was admitted with febrile convulsion for the first time. The patient had a medical history of *in vitro* fertilization (IVF) and premature delivery in the 21st week of pregnancy. Further, he suffered from neonatal germinal matrix hemorrhage and intraventricular hematoma (IVH). The patient was treated in the pediatric intensive care unit. Cranial magnetic resonance imaging (MRI) showed resorption of the hematoma with the presence of cavum vergae (CV) variation. The new brain MRI revealed progression of the cavum to a large CV cyst compared with the previous imagings, causing pressure upon both lateral ventricles. Endoscopic fenestration of the lateral walls of the cyst with lateral ventricles was performed. The postoperative period was uneventful and the patient was discharged the next day. Follow-up MRI one year after surgery showed normal ventricle size, shrinkage of the cyst and cerebrospinal fluid (CSF) flow between the cyst and the lateral ventricles. This is an interesting demonstration of the progression over years of a CV anatomical variation to a large CV cyst in a premature birth case that experienced germinal matrix hemorrhage.

Key words: cavum vergae cyst, intraventricular hemorrhage, neuroendoscopy, seizure.

First described by the Italian anatomist Vergae in 1851[1], cavum vergae (CV) -also called the sixth ventricle- is an intraventricular cyst located between the corpus callosum above and the commissure of the fornix below. It is found frequently in preterm infants and term neonates, and most disappear soon after birth. However, persistent CV is detected in 15% of the asymptomatic adult population[2]. Some cases become symptomatic with expanding cysts causing obstruction of cerebrospinal fluid (CSF) flow through the foramen of Monro, leading to increases in intracranial pressure (ICP) and hydrocephalus[3]. Symptomatic cases require surgical intervention, which includes open surgical procedures, ventriculo-peritoneal shunting, stereotactic fenestration, and neuroendoscopic cysto-ventriculostomy[3-5]. Today, endoscopic cyst wall fenestration is the method of choice in many neurosurgical centers. The technique is minimally invasive, permits direct visualization of the cyst wall and avoids the need to place a shunt. While the incidence of the symptomatic CV is rare, endoscopic treatment of these cysts is even more rarely reported in the literature[5-6].

In this report, the authors present a case of progressively enlarging CV cyst in a child, who was successfully treated by endoscopic fenestration of the cyst, together with a brief review of the literature on this issue.

Case Report

A 3.5-year-old boy was admitted with febrile convulsion for the first time. The patient had a medical history of in-vitro fertilization (IVF) and premature delivery in the 21st week of pregnancy. During the follow-up in the neonatal intensive care unit, the patient suffered from neonatal germinal matrix hemorrhage and intraventricular hematoma (IVH). The hematoma was observed and follow-up brain magnetic resonance imaging (MRI) showed resorption of the hematoma and the presence of a CV cyst (Fig. 1A, B). Thereafter, the patient was followed in the pediatric neurology clinic with a regular physiotherapeutic program and MRI (Fig. 1C, D).

Seizure intervention was done with intravenous phenobarbital and the patient was unconscious for 20 minutes in the postictal period. Physical examination and chest X-ray revealed upper respiratory tract infection (URTI). Neurological examination after the postictal period showed general minimal motor retardation with more significant spastic paresis of the lower extremities. Mental and language development were found to be normal. Brain computerized tomography (CT) scan and brain MRI revealed a large CV cyst compared to the cyst volume in the previous imagings, causing pressure upon both lateral ventricles (Fig. 1 E-H). Surgical intervention was planned after completing the antibiotic trial and resolving the infection.

The patient was operated under general anesthesia. The endoscopic approach was performed through the right frontal burr hole, and the endoscope was introduced into the right lateral ventricle. The cyst was identified and was found covering the foramen of Monro. The right wall of the cyst, which was covered with a thin ependymal layer, was fenestrated just posterior to the septal vein. The cyst was entered and the opposite (left) wall was identified and also fenestrated. The procedure was completed without complication. The postoperative period was uneventful and the patient was discharged the next day. The last follow-up MRI one year after the surgery showed normal ventricle size with shrinkage of the cyst and communication of the CSF between the cyst and the lateral ventricle (Fig. 1 I-L).

Discussion

Cavum vergae (CV) is considered a normal variant that usually does not require surgical treatment. However, some cases become symptomatic by causing obstruction of CSF flow through the foramen of Monro, leading to various symptoms including headache, papilledema, emesis, loss of consciousness, and visual and sensorimotor disturbances caused by increased ICP and hydrocephalus[3-7]. CV is classified as a non-communicating or communicating cyst, depending on whether or not the cyst communicates with the cerebral ventricular system. The communicating type is the most common and is considered to be asymptomatic[8]. In contrast, non-communicating cyst, which could originally be asymptomatic, may enlarge, block the foramen of Monro, and cause symptomatic hydrocephalus. A communicating cyst may become a non-communicating cyst through various mechanisms. For example, the cyst might secrete fluid, possibly through the presence of migrated ependymal cells[3-7]. The check valve phenomenon between the cyst and the subarachnoid cavity may be implicated based on immunohistochemical analysis[9]. The involvement of minor head injury was also implicated in the expansion of these cysts[10]. However, the exact mechanism of CV expansion remains uncertain, since most reported cases were only identified after becoming symptomatic. In our case, the presence of CV can be related to the prematurity of the patient, complicated by IVH.

An enlarging cyst may obliterate the foramen of Monro and/or the aqueduct of Sylvius and create intermittent hydrocephalus that causes symptoms, therefore justifying surgery. In these reports, the enlargement of the cavum has been attributed to a one-way valve mechanism[9]. Moreover, the absence of secretory ependymal

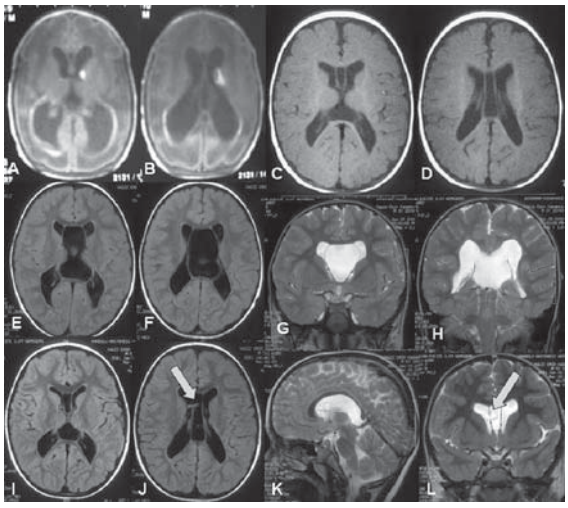

Fig. 1. Brain MRI of our case in different life periods demonstrating progressive expansion of the cyst and the postoperative results. A-B: The first MRI during follow-up in the neonatal ICU showing the resolved IVH and formation of CV. C-D: The second MRI at the age of 1 year showing the classical CV with minimal expansion compared to the first imagings. E-H: The third MRI at the age of 3.5 years showing the massive expansion of CV compressing the lateral ventricles bilaterally. I-L: Postoperative MRI 1 year after the operation showing regression of CV, decompression of the lateral ventricles and the communication of CSF flow between the cyst and the lateral ventricles *(arrows)*.

lining in these cavities has been proven by many autopsy series[9,11]. Oteruelo[12] suggested a different mechanism and claimed that fluid travels passively from the ventricle into the cavum through the septal lamina by a pressure gradient in the absence of any communication. Resorption of the fluid is performed by septal capillaries and veins[13]. In several reports, it was claimed that the underlying pathophysiological mechanism of the symptoms is an intermittent obstruction of the foramen of Monro that results in raised ICP, and not the compression of surrounding structures, that causes limbic dysfunction[9,11,13-15]. Of interest, a dilated cavum septum pellucidum has been associated with psychiatric disorders, mainly schizophrenia[16-17]. On the other hand, Bodensteiner et al.[17] found that CV alone does not identify individuals at risk for cognitive disorders. In our case, the patient was found to have normal mental status but delayed motor development. This was probably secondary to the history of neonatal IVH and the pressure of the enlarging cyst upon the surrounding neural tissue. Due to the variable developmental outcomes, it is important to make the correct diagnosis of CV, which may be confused with cavum septum pellucidum. CV is a cavity within the septum pellucidum, and is located posterior to an arbitrary vertical plane formed by the columns of the fornix[18]. On the other hand, cavum septum pellucidum consists of two thin translucent leaves that extend from the anterior part of the body, the genu and the rostrum of the corpus callosum, to the superior surface of the fornix, forming the medial wall of the lateral ventricles[19]. The other differential diagnoses include a dilated third ventricle, aneurysm of the vein of Galen and an arachnoid cyst.

Bronstein and Weiner[18] first reported prenatally diagnosed cases of cava septum pellucidum et vergae in 1992. These authors reported eight cases diagnosed by midtrimester transvaginal ultrasonography. Among these cases, associated anomalies were detected in five fetuses. As a result, they concluded that prenatal detection of cavum septum pellucidum et vergae should be followed by a detailed search for associated anomalies. Sahinoglu et al.[20] reported ultrasonographic prenatal diagnosis of three fetuses with dilated CV. One of these fetuses had ventriculomegaly and lumbar meningocele. Another patient required placement of a stereotactic cystoperitoneal shunt at six months of age because of intracranial hypertension and progressive enlargement of the CV. The third infant remained asymptomatic.

Historically, CV cysts have commonly been treated by craniotomy with cyst fenestration or removal or by cystoperitoneal shunting. Cystoperitoneal shunting has proven to be a safer, simpler and more effective treatment method than open surgery, and is considered to be the best treatment of a large cyst in an infant or a young child. However, complications related to the shunting procedure such as obstruction, infection, hemorrhage, and life-long shunt dependence are still not uncommon and should be considered[11].

With the high failure rate of shunts[21] and the high morbidity rate after craniotomy-based approaches, such as postoperative neurological deficits, seizures, subdural hygroma, and meningitis[22], there is a trend toward minimally invasive procedures. With the advances in neuroendoscopy and the large experience gained over the last 20 years, many articles advocate the advantages of this procedure to manage certain intracranial pathologies, with identification of the indications and surgical techniques[23-26]. The neuroendoscopic approaches have gained in popularity and have been favored for the treatment of these lesions. This was supported by reports that documented the high success rate (71%-81%) of the neuroendoscopic treatment of intraventricular cysts[21]. Furthermore, seeking more precise localization of the intraventricular cyst to reduce the failures and complications, a combination of neuroendoscopy with stereotaxy[5,14,27,28] or neuronavigation system application[29] has been reported. Now, the role of neuroendoscopy is standard for the treatment of hydrocephalus by endoscopic third ventriculostomy[25], aqueductoplasty[30], intracranial arachnoid cyst fenestration[31,32], intraventricular and pineal tumor biopsy or removal[33], and colloid cyst removal[34]. The neuroendoscopic approaches are considered safe techniques, especially in the hands of highly experienced surgeons in specialized centers[35], permitting direct visualization of the cyst wall and avoiding the need for a shunt. The endoscopic surgical technique includes cysto-ventricular fenestration to achieve

communication between the cyst and both lateral ventricles. This is done by fenestration of both lateral walls of the cyst, as shown in our case. However, the failure and complication rates still range between 2% and 7%[36]. In addition to technical failures, the most common complications are CSF leakage, intraventricular hemorrhage, intraparenchymal hemorrhage, subdural hygroma, and hematoma[37]. These complications were not seen in our case.

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