ENDOSCOPIC TREATMENT OF *IN UTERO* DIAGNOSED MULTILOCULATED INTERHEMISPHERIC CYST IN A NEWBORN: CASE REPORT

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SUMMARY – Interhemispheric cysts, often associated with agenesis of corpus callosum, are rare lesions. The optimal treatment is still controversial. Placement of cystoperitoneal shunt and open microsurgery are traditional treatments. Neuroendoscopy in children is due to its minimal invasiveness a new emerging option. There have been a few published cases on neuroendoscopic treatment of interhemispheric cyst in children. The authors document the youngest reported child with multiloculated interhemispheric cyst that was treated with neuroendoscopy. The cyst was detected in a male fetus in 35th week of gestation and *in utero* magnetic resonance imaging was performed in 37th week of gestation. After delivery, progressive macrocrania with signs of raised intracranial pressure developed. Endoscopic cystoventriculocisternostomy was performed 28 days after the birth. There was a marked symptom relief. One month after the surgery, magnetic resonance showed shrinkage of the cyst and expansion of the brain parenchyma. After a 2-month follow up period, the child showed normal neurologic development and head circumference increased by only 0.5 cm. The created fenestrations enabled the brain to expand. Neuroendoscopic treatment of interhemispheric cysts should be considered the operative technique of choice in newborns. Although the intracranial pressure and the size of the cyst have decreased, long-term follow up is necessary and future studies on more cases are needed.

Key words: *Arachnoid cyst; Neuroendoscopy; Newborn; Ventriculostomy*

Introduction

Interhemispheric cysts are rare lesions, often associated with complex brain malformations such as agenesis of corpus callosum and hydrocephalus[1-5]. Only a limited number of cases have been reported[2,4-6]. Therefore, there is still controversy regarding optimal treatment. Open microsurgery and placement of cystoperitoneal (CP) shunts are associated with good results, but also have some disadvantages[2,4-6]. Pure endoscopic treatment provides a minimally invasive method to achieve cyst decompression by establishment of communication with the physiological cerebrospinal fluid (CSF) pathways[4,7,8]. To the best of our knowledge, there have been only few published cases describing neuroendoscopic treatment of interhemispheric cysts in children, only one in newborn[2].

We report the first case of a newborn with multiloculated interhemispheric cyst that was treated neuroendoscopically.

Case Report

A large multiloculated interhemispheric cyst was detected on ultrasound in a male fetus in 35th week of gestation in a 24-year-old primipara. Intrauterine magnetic resonance imaging (MRI) performed in 37th week of gestation revealed a large (3.5x7.2x6 cm) multiloculated interhemispheric right-sided parasagittal

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cyst with agenesis of corpus callosum and gray matter heterotopy (Fig. 1A and C). There was no communication between the cyst and ventricular system. A midline shift towards the left side was present. According to Barkovich *et al.*[1], it was classified as a type 2c cyst.

The male infant was vaginally delivered with no complications in the 37th week of gestation (Apgar 9/9, weight 3880 g and length 53 cm). His head circumference (HC) was 37 cm. In the next weeks, he developed signs of raised intracranial pressure (ICP) and HC increased rapidly (3 cm in the first week).

The newborn underwent pure endoscopic surgery in general anesthesia at the age of 28 days. A rigid endoscope with four channels was used (Ausculap AG, Tuttlingen, Germany). A single burr hole was made 1 cm laterally from the midline and just in front of the anterior fontanel on the right side. The large interhemispheric cyst was reached neuroendoscopically a few millimeters below the cortex. The cyst was filled with CSF and it was septated. At first septostomy inside the cyst was performed and then the wall of the cyst was widely fenestrated anteriorly and basally, towards the right lateral ventricle. Fenestration was performed using bipolar coagulation and scissors. Dilatation of the fenestration was performed using grasp forceps and Fogarty balloon catheter. In this way, a wide area of communication was created between the cyst and the anterior horn of the right lateral ventricle. The right lateral ventricle was pushed laterally. The foramen of Monro was then visualized and the third ventricle was reached endoscopically. Thalamostriatal vein, choroid plexus and anterior vein of septum pellucidum were identified around the foramen of Monro. Third ventriculostomy was made in front of the corpora mammillaria in the standard way. Basilar artery and brain stem were shown. Areas of communication were confirmed by direct visualization during surgery. During the procedure, no significant hemorrhage was noticed.

The postoperative course was complicated by transient hyponatremia (121 mmol/L), which resolved after fluid restriction in 3 days. The child's condition improved. One month after the surgery, control MRI showed significant reduction of the cyst volume with expansion of the right hemisphere (Fig. 1B and D). Throughout the 2-month follow up period, the child was symptom-free and without any signs of increased ICP. His weight was 6370 gm, length 58 cm and HC increased only to 40.5 cm.

Discussion

This case represents the first infant with multiloculated interhemispheric cyst being treated neuroendoscopically during the first month of delivery. To the best of our knowledge, only a few cases of interhemispheric cysts treated endoscopically have been described[2]. In only one case, the patient was a newborn, but this interhemispheric cyst was not multiloculated as in our report[2].

There are two types of interhemispheric cysts according to the classification proposed by Barkovich *et al.*[1]. Type 1 cysts are extensions or diverticulations of the third or lateral ventricles, whereas type 2 are loculated and do not communicate with the ventricular system[1,9]. They are separated into subtypes according to associated anomalies such as neuronal heterotopias and gyral malformations[1,9]. In our case, it was a type 2c cyst. Nowadays, interhemispheric cysts can be incidentally discovered prenatally with ultrasonography[10,11]. Additional intrauterine MRI better localizes the cyst and gives better view of the parenchyma as well as of the cortex and CSF space[10]. Interhemispheric cysts are usually large and can distort the CSF pathways leading to asymmetric ventricular enlargement[3,4]. They can be either asymptomatic or accompanied by clinical symptoms and signs such as macrocrania, failure to thrive, irritability, hemiparesis, epileptic seizures and psychomotor retardation[2,4,6,12-15]. The appropriate time and indication for surgery can sometimes be difficult to determine[3-5]. The operation is indisputably indicated when signs of raised ICP appear[2,3,5,11,16]. Apart from this, the significant volume of the cyst and macrocrania were also supporting surgical treatment in our case. Nevertheless, it must also be kept in mind that intracranial cysts can stop increasing in size or even regress but those that are located interhemispherically are more likely to progress than others[10].

The reported cyst was multiloculated, which made endoscopic treatment more challenging because additional fenestrations had to be made within the cyst. In such cases, open microsurgery is usually easier to

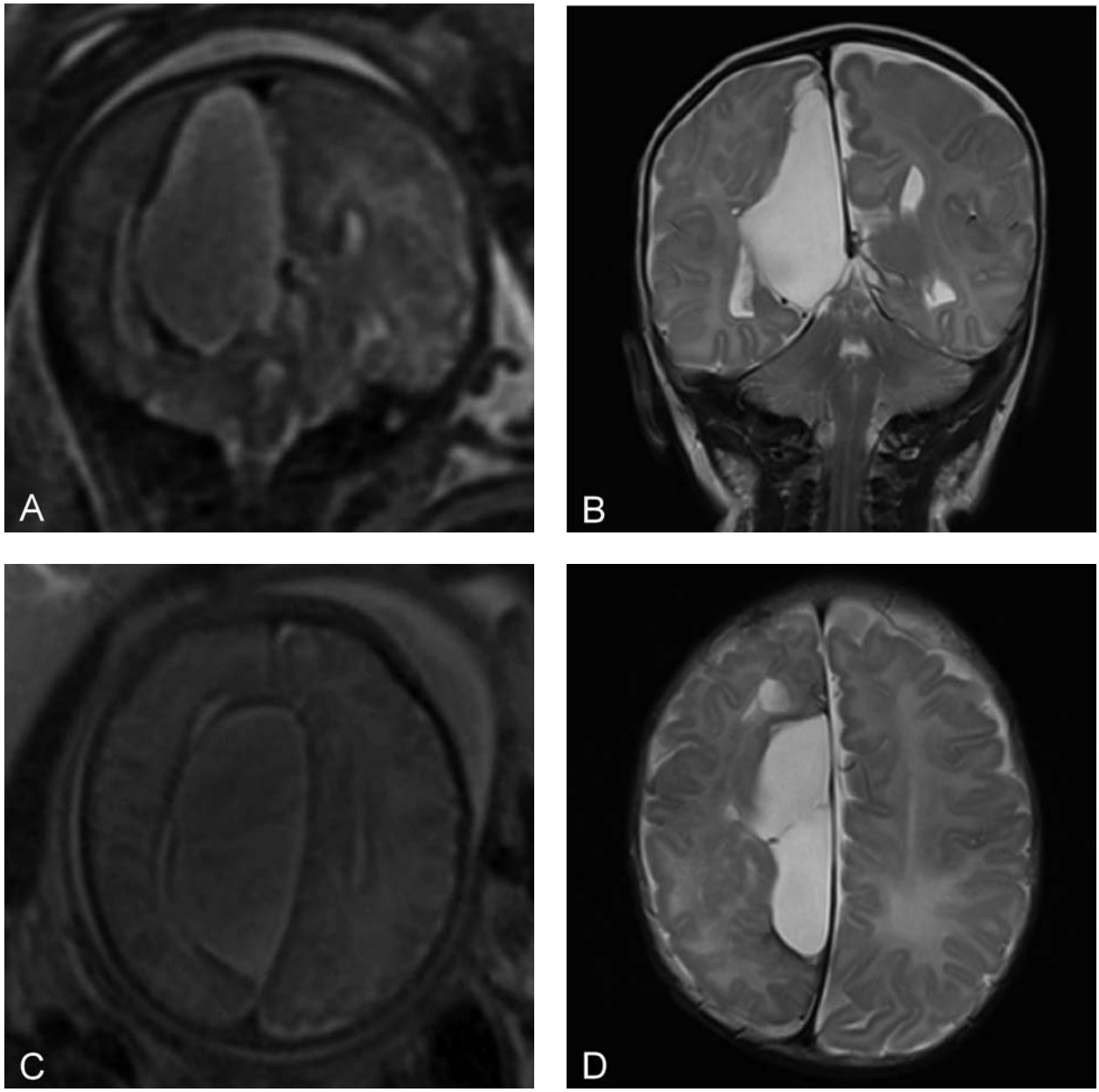

Fig. 1. A and C: Prenatal coronal and axial magnetic resonance images revealing a large multiloculated interhemispheric cyst; B and D: follow-up coronal and axial magnetic resonance images acquired 1 month after cystoventriculocisternostomy revealing reduction in cyst volume and expansion of brain parenchyma.

perform and broader excision of the cyst walls can be achieved[4,6,17]. Calderelli and Di Rocco[6] argue that the direct craniotomic approach to the interhemispheric cyst has more consistent results than CSF shunting devices and therefore CP shunts should be considered a second-choice treatment[6]. On the other hand, open microsurgery in newborns carries increased morbidity and mortality. Therefore, minimally invasive methods such as placement of a CP shunt and endoscopic treatment might be a better alternative[6]. Ulu *et al.*[5] report on a series of seven patients with interhemispheric cysts being treated with CP shunts. Cysts were completely resolved in five patients[6]. Furthermore, Calderelli and Di Rocco[6] report on a 50% success rate

with CP shunts. In all cases of shunt failure, the proximal end was entrapped by the collapsed cyst lining[6]. The drawbacks of this treatment are also infections, postoperative seizures, subdural hygromas, and shunt dependency[4-6.11]. Moreover, given the multiloculated nature of the cyst, recurrence after the placement of a CP shunt is not exceptional[4,5,18,19]. Moriyama *et al.*[4] describe an enlargement of another cyst component after CP shunt placement, which resulted in an additional open surgery. Therefore, a communication inside the cyst has to be performed and endoscopy appears to be a promising solution[4]. Endoscopic fenestration of the cyst wall is a relatively minimally invasive technique with a reported success rate of 71% to 81%[2,13,20]. However, the number of series of endoscopic procedures involving interhemispheric cysts is limited, the first being reported by Cinalli *et al.*[2] in 2006. Out of seven patients, complete success was achieved in five[2].

Interhemispheric cysts are accessible with a neuroendoscope through a thin layer of brain parenchyma. Once the cyst has been penetrated, a significant brain shift occurs. Therefore, the neuronavigation may not be accurate and it was not used in our case. Nevertheless, it could be helpful to identify the suitable point of perforation. In our case, the cyst was large, situated medially and superficially, and it was easily accessible without neuronavigation. Once in the cyst, the neurosurgeon might face some orientation difficulties due to the brain shift and distorted anatomy; recognizing visible landmarks is therefore crucial. On the other hand, direct visualization of structures enables detection of additional intraventricular pathology associated with the cyst[11,17]. After fenestrations had been made, the pressure within the cyst decreased to the level of the intraventricular pressure, but the cyst stayed distended. This technique prevents the cyst walls from collapsing and blocking the areas of communication created endoscopically. With more radical resection, patches of the cyst membrane would be created and could block the communications in the CSF space. Large cysts adjacent to the CSF pathways interfere with normal CSF flow and/or compress adjacent neuronal structures leading to intracranial hypertension[2,5,17]. It has also been described that interhemispheric cysts can be associated with some additional intraventricular abnormalities such as aqueductal stenosis[10,17]. Therefore, ventriculostomy of the third ventricle was made to minimize the risk of hydrocephalus[2,10,11]. Using neuroendoscopy to create communications between the cyst, the ventricular system and the subarachnoid space, the basic surgical strategy of cyst marsupialization is maintained, while invasiveness of open microsurgery and complications caused by shunting, which are especially common in very young children, are avoided[3,6,17,18]. The postoperative course was complicated by transient hyponatremia, which was probably caused by endoscopic third ventriculostomy performed in the close proximity of the supraopticohypophyseal tract[21,22]. Neurosurgeons should be aware of the possible electrolyte disbalance, which can cause neurologic deterioration in the patient. Other complications that can be expected in neuroendoscopic procedures are bleeding, subdural collection of CSF, and diabetes insipidus[2,13,22]. Comparison of preoperative, postoperative and follow up clinical status and radiologic images is of major importance. However, reduction in the cyst size after endoscopic procedure is variable. Permanent clinical improvement has been reported despite only minimal reduction of the cyst volume based on imaging[2,3,17]. Therefore, additional surgical interventions are indicated based on the patient's symptoms and not only on radiologic findings[17]. In our case, expansion of the displaced structures was visible and there was rapid resolution of the symptoms. It seems that the cyst is communicating well with the ventricles and the subarachnoid space. Nevertheless, there is always the risk of re-occlusion, therefore long-term follow up is needed.

It has been shown that reduction of the cyst size is more evident in children under the age of two years due to brain plasticity[14,18]. The current possibility of diagnosing these lesions *in utero* or in early infancy gives the neurosurgeon an opportunity of eliminating or at least reducing the cause of possible interference with cerebral growth prior to the establishment of irreversible brain damage. This gives a growing brain room to expand and helps prevent developmental delay[6]. In the case reported, the HC increased by only 0.5 cm. This indicates that the brain is expanding to the expense of cyst shrinkage.

It seems that neuroendoscopy, when possible, presents an ideal way between invasiveness and effectiveness. With advancing technique and more ex-

perienced neuroendoscopists, more complex cases are being treated this way and we believe neuroendoscopy will become the first line treatment for interhemispheric and other intracranial cysts in newborns. Even though it is recognized as a safe technique, we should be aware of the possible complications. Despite favorable initial results, long-term follow up and further studies comparing endoscopic treatment with other methods are needed.

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Sažetak

ENDOSKOPSKO LIJEČENJE UNUTARMATERNIČNO OTKRIVENE MULTILOKULARNE INTERHEMISFERIČNE CISTE U NOVOROĐENČETA: PRIKAZ SLUČAJA

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Interhemisferične ciste, često s pridruženom agenezom korpusa kalozuma, rijetko se pojavljuju. Optimalno liječenje je još uvijek sporno. Cistoperitonealna drenaža i otvoreni mikrokirurški zahvat su tradicionalne metode. Neuroendoskopija u djece je obećavajuća metoda zbog svoje minimalne invazivnosti. Dosad je objavljeno svega nekoliko slučajeva neuroendoskopskog liječenja interhemisferične ciste u djece. U ovom članku su autori opisali slučaj najmlađeg djeteta s multilokularnom interhemisferičnom cistom operiranog neuroendoskopskom tehnikom. Cista je bila otkrivena u muškog fetusa u 35. tjednu trudnoće, a u 37. tjednu je učinjena magnetska rezonancija unutar maternice. Nakon poroda se razvio ubrzan rast glave i znaci povišenog intrakranijskog tlaka. Učinjena je endoskopska cistoventrikulocisternostomija 28. dana nakon poroda i nastupilo je značajno poboljšanje. Mjesec dana nakon zahvata kontrolna magnetska rezonancija je pokazala smanjivanje ciste i širenje tkiva mozga. Nakon dvomjesečnog praćenja utvrđen je normalan neurološki razvoj djeteta, a opseg glave je porastao za samo 0,5 cm. Otvori u membrani ciste su omogućili širenje mozga. Neuroendoskopsko liječenje interhemisferične ciste bi trebalo smatrati metodom izbora u novorođenčadi. Međutim, iako su se intrakranijski tlak i volumen ciste smanjili, potrebna su daljnja istraživanja na većem broju bolesnika i uz dugoročno praćenje.

Ključne riječi: *Arahnoidna cista; Neuroendoskopija; Novorođenče; Ventrikulostomija*