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Original Investigation

Third Ventricle Floor Variations and Abnormalities in Myelomeningocele-Associated Hydrocephalus: Our Experience with 455 Endoscopic Third Ventriculostomy Procedures

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ABSTRACT

AIM: To evaluate the incidence of anatomical variations and abnormalities of the third ventricle floor encountered during the endoscopic third ventriculostomy (ETV) procedure in myelomeningocele-associated hydrocephalus (MAH) cases.

MATERIAL and METHODS: A retrospective analysis was performed on 455 pediatric MAH cases that had been treated with ETV. This case series consisted of the patients who were initially treated with ETV and also those who were treated with ETV for the management of cerebrospinal fluid shunt dysfunction. Variations and anomalies of the third ventricle floor were determined by reviewing the video records of the ETV procedures.

RESULTS: The analysis of the data revealed that the rate of the MAH cases with variations and abnormalities of the third ventricle floor was 41.1%. The most common anatomical features were "thick and prominent massa intermedia" (37.1%) and "narrow tuber cinereum" (33.1%).

CONCLUSION: This study documents the most common anatomical variations and abnormalities of the third ventricle floor in cases with MAH. Various anatomical situations and specific ventricular configuration of MAH cases may add an operative factor of difficulty which should be well recognized by the neurosurgeon who plans and executes an ETV procedure in this patient population.

KEYWORDS: Endoscopic third ventriculostomy, Hydrocephalus, Myelomeningocele

INTRODUCTION

Myelomeningocele is the most common form of spinal dysraphism and affects the whole central nervous system (10). Most patients with myelomeningocele present with associated hydrocephalus (7). The hydrocephalus in myelomeningocele patients has an incidence of 15 to 20% at birth; however, the incidence reaches 80 to 90% after closure of the defect (10).

Surgical management by shunt implantation is a widely used and an effective treatment for hydrocephalus (13). However, consistent follow-up is necessary after shunt implantation, taking into consideration several shunt-related complications including obstruction, fracture, and infection (13).

The endoscopic third ventriculostomy (ETV) procedure is well recognized as an effective alternative to cerebrospinal fluid (CSF) shunt placement and has been an ideal treatment of

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choice for obstructive hydrocephalus in recent years (10). Although, myelomeningocele-associated hydrocephalus (MAH), which can be congenital or acquired after the myelomeningocele closure, has an obstructive basis, the use of ETV for the management of hydrocephalus in these patients is still controversial (9,10,13). Anatomical variations and abnormalities of the ventricular system and the confusing topography of the floor of the third ventricle are of utmost importance in the ETV procedure, since all these factors play an important role in the limitations and current risks of surgery.

In this study, we aimed to evaluate the incidence of anatomical variations and abnormalities of the third ventricle floor during the ETV procedure in MAH patients.

MATERIAL and METHODS

The data of 455 MAH patients who were treated with ETV procedure at two pediatric neurosurgery centers (Kocaeli University and Selcuk University) between 2004 and 2016 were retrospectively analyzed. The patients who were treated with ETV in the first-line setting and those with a mechanical or dynamic shunt failure who were later treated with ETV as an alternative to shunt revision were included in the study. Since previous CSF infection or hemorrhage may affect anatomical development and configuration, cases with a history of previous CSF or shunt infection or intraventricular hemorrhage were excluded from the study. A written informed consent was obtained from each patient. The study was conducted in accordance with the principles of the Declaration of Helsinki.

The incidence and the types of encountered variations and the abnormalities of the third ventricle floor in patients with MAH were analyzed by reviewing all the video recordings of ETV procedures performed in 455 cases.

RESULTS

Among the 455 MAH patients who treated with ETV, 187 patients had "third ventricle floor variations and abnormalities" based on video recordings of the procedures. The rate of MAH patients with anatomical variations and abnormalities of the third ventricle floor was 41.1%. The most common anatomical features of the third ventricle floor and the rate of those features were as follows: thick and prominent massa intermedia (37.1%), narrow tuber cinereum (33.1%), parenchymatous, opaque and/or thick floor of the third ventricle (27%), hollow/steep third ventricle floor (18%), small anterior chamber of the third ventricle (17.1%), the presence of interhypothalamic adhesions (14.9%), vascular floor of the third ventricle (14%) and the presence of adhesions or bridges between mammillary bodies (13.1%). Two or more types of anatomical features were documented in most of the cases.

The distribution of anatomical variations and abnormalities in our series are summarized in Table I. Intraoperative endoscopic images of the most common anatomical features of the third ventricle floor in MAH patients are shown in Figure 1A-F.

DISCUSSION

Although the incidence of MAH at birth is 15 to 20%, it increases up to 80 to 90% after the closure of the defect (10). However, the exact etiology of MAH still remains unclear. According to McLone and Knepper (8), hypoplastic development of the nervous system structures and the caudal descent of the hindbrain occur due to insufficient mesenchymal induction during the embryonic life at the level of the posterior fossa, leading to hydrocephalus. The Chiari II malformation with abnormality of the fourth ventricle floor and the cervicomedullary junction represents the main factor of the obstructive nature of the MAH (3,8).

The standard surgical treatment of MAH still remains the insertion of a ventriculoperitoneal shunt; however, it has significant associated complications such as infection (3). On the other hand, ETV is the procedure of choice in the treatment of obstructive hydrocephalus (11,12). It is safe and efficient with a success rate of 70 to-90% (11,12). Major complications of ETV are low when the procedure is performed by experienced surgeons. Kulkarni et al. (6) reported that the incidence of complications such as new neurological deficit or meningitis was less than 2%. Although the recent development of ETV

Table I: Distribution of Anatomical Variations and Anomalies in Our Series. Among 455 Cases, 187 Had One or More Anatomical Variation Types

Anatomical Variation	Number of Cases	Percent (%)
Thick and prominent massa intermedia	169 of 455 cases	37.1
Narrow tuber cinereum	151 of 455 cases	33.1
Parenchymatous, opaque and/or thick floor of the third ventricle	123 of 455 cases	27
Hollow/steep third ventricle floor	82 of 455 cases	18
Small anterior chamber of the third ventricle	78 of 455 cases	17.1
Existence of interhypothalamic adhesions	68 of 455 cases	14.9
Vascular floor of the third ventricle	64 of 455 cases	14
Existence of adhesions or bridges between mammillary bodies	60 of 455 cases	13.1

has provided a challenging way of treatment, it still has a very limited role in the first-line treatment of MAH patients due to the immature CSF reabsorption system and anatomical difficulties in these patients; therefore, this method should be suggested only in selected cases (3,10).

Furthermore, the alterations or anatomical variations of the anatomy of the floor of the third ventricle play an important role in limitations of the ETV procedure and these can also complicate the surgical procedure (10). Perez da Rosa et al. reported the structural changes as malformation of the corpus callosum (hypoplasia, dysplasia, focal posterior thinning), enlarged massa intermedia, gray matter heterotopias (medial occipital lobes), absence of the septum pellucidum, stenosis of the foramen of Monro, fusion and thickening of the forniceal columns, thickening of the choroid plexus, interhypothalamic adhesions, thickening or distortion of the floor of third ventricle (which can complicate visualization of anatomical structures such as the infundibulum or the mammillary bodies), large and posteriorly displaced occipital horns, interhemispheric cyst and protrusion of the roof of the third ventricle in a 'diamond' shape up towards the velum interpositum and the quadrigeminal cistern (10).

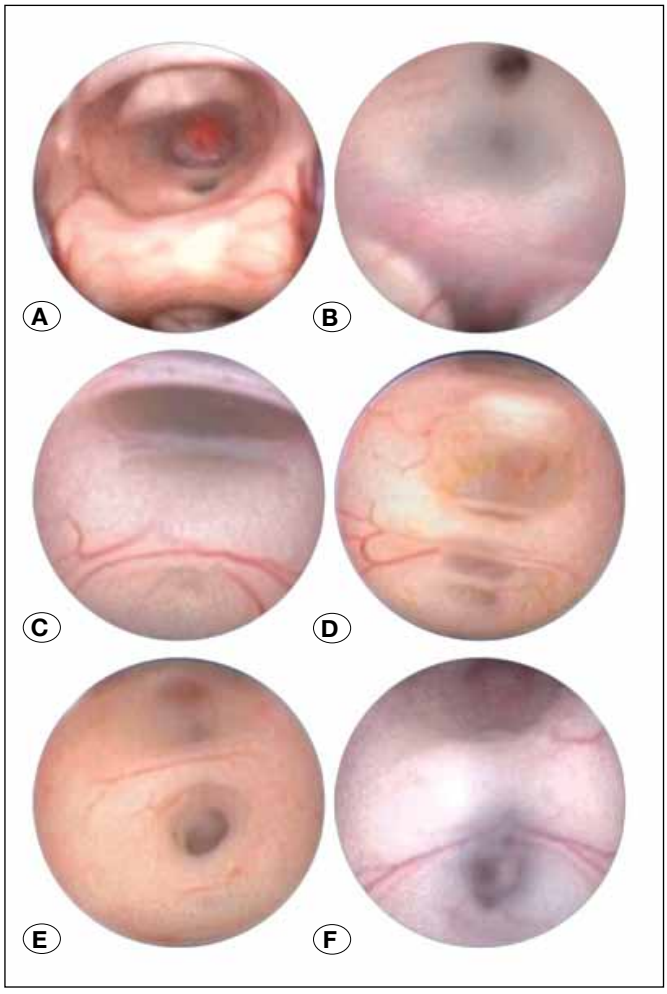

Figure 1A-F: Figures show intraoperative endoscopic images of the third ventricle floor in myelomeningocele-related hydrocephalus during endoscopic third ventriculostomy procedure. The most common anatomical features were as follows:

- Thick and prominent massa intermedia **(A)**
- Parenchymatous, opaque and/or thick floor of the third ventricle **(B,C,D,E)**
- Narrow tuber cinereum **(A, C, E, F)**
- Hollow, steep or vascular floor of the third ventricle **(C, E, F)**
- Small anterior chamber of the third ventricle **(C, E)**
- Presence of interhypothalamic adhesions **(C,D)**
  Presence of adhesions or bridges between mammillary bodies **(D)**

Ventricular system abnormalities are present in most myelomeningocele patients. Gilbert et al. described anomalies in 92% of autopsy exams (4,9). Babcock and Han found a rate of 96% as evidenced by echographic scans, Zimmermann et al. reported a rate of 89% in tomography scans (1,14). In another study Kawamura et al. (5) reported a rate of 8/10 for ventricular abnormalities in magnetic resonance images. Bankole et al. (2) also reported the abandonment of the procedure in 15 of 52 patients due to distorted anatomy or poor visibility in a heterogeneous population of hydrocephalus. In the current study, at least one anatomical variation was seen in 187 cases (41.1%). Thick and prominent massa intermedia was the most commonly encountered anatomical variation as seen in 169 of 455 cases (37.1%). A narrow tuber cinereum, parenchymatous, opaque and/or thick floor of the third ventricle, hollow/steep third ventricle floor, small anterior chamber of the third ventricle, existence of interhypothalamic adhesions, vascular floor of the third ventricle and existence of adhesions or bridges between mammillary bodies were the other anatomical features seen.

Some of our MAH cases have been treated with ETV as an alternative to shunt revision because of a mechanical or dynamic shunt failure. A few of these cases had a history of long-term (over two years) shunt dependence. It is possible that, long-term shunt existence may further affect the anatomical development and configuration especially when the CSF shunt system has been placed in the neonatal period. A future complementary study may help to unveil such effects in MAH cases.

CONCLUSION

ETV is the treatment of choice and an alternative to shunt insertion in selected MAH patients. Our study results suggest that anatomical variants and specific ventricular configuration of MAH may add an operative factor of difficulty that should be well recognized by the neurosurgeon who plans and executes an ETV procedure in this patient population.

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