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Isolated Fourth Ventricle Causes a Mass Effect in the Posterior Fossa. Difficulties of Making a Diagnosis in the Intensive Care Unit in Comatose Patients: Clinical Overview

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Abstract

Isolated fourth ventricle (IFV) is a rare radiological and clinical complication of the surgical treatment within the cerebral fluid pathways. It may occur as a result of a neurosurgical procedure when post resection space communicates with the fourth ventricle, be a complication of shunt therapy, external drainage or endoscopy of the ventricular system. Neuroinfection or subarachnoid hemorrhage can predispose to the development of the syndrome. The pathomechanism is associated with disturbances in the patency of the Sylvian aqueduct and the outflow routes from the fourth ventricle. An important role in pathology is attributed to the vascular plexus of the fourth ventricle and its physiological secretion of cerebrospinal fluid. The resulting IFV reservoir causes a mass effect in the posterior fossa and the development of compression symptoms in the brainstem. Imaging tests reveal the signs of retention of the cerebrospinal fluid (CSF) inside the surrounding white matter of the posterior fossa. This causes deterioration of the clinical state which is dependent on the compression on the brainstem. At the same time, the supratentorial space of the brain is effectively treated with ventricular drainage or using a shunt. Previous publications mainly concentrated on pediatric patients. In adult patients, IFV is a rare complication that develops few months after the first surgical procedure. The mass effect increases consciousness disorders and brainstem symptoms. We present the diagnosis of IFV in a patient who was unconscious and subjected to long-term hospitalization in the ICU. We pointed out the importance of a detailed examination of brainstem symptoms and the necessity to perform the sequence of MRI images with the cine option after specific neurosurgical procedures. Some neurosurgical procedures may predispose to the development of IFV.

Keywords: Fourth ventricle; Posterior fossa; Isolated ventricle; Complication; Ventriculostomy; Shunt therapy; Intensive care unit

Introduction

Isolated fourth ventricle (IFV) is a rare complication of neurosurgical treatment. It may occur after surgical procedures performed within the posterior fossa, after implantation and replacement of valves, external drainage of the supratentorial ventricular system and endoscopic ventriculostomy of the third ventricle. IFV may be caused by the mechanical or functional obstruction of the reversed flow within the Sylvian aqueduct and obstacles of outflow from the fourth ventricle through the anatomical foramina of Luschka and Magendi. Cerebrospinal fluid (CSF) is produced in the choroid plexus of the lateral ventricles and third ventricle; according to the views of some clinicians the production of CSF in the fourth ventricle is small and does not play any role.

According to our observations and literature reports, the production of CSF by the plexus of the fourth ventricle plays specific role especially for the different pathology in this region. The lack of communication with the supratentorial and infratentorial CSF spaces causes clinical symptoms, such as radiological "isolation" of the fourth ventricle (IFV) and the mass effect in the posterior
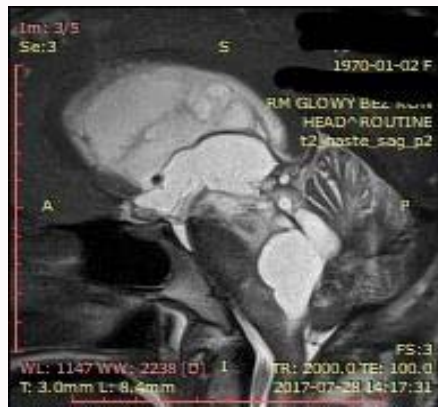
Figure 1: The cystic form with the CSF in the posterior fossa located in the midline with a mass effect and hypodense zones around the fourth ventricle.
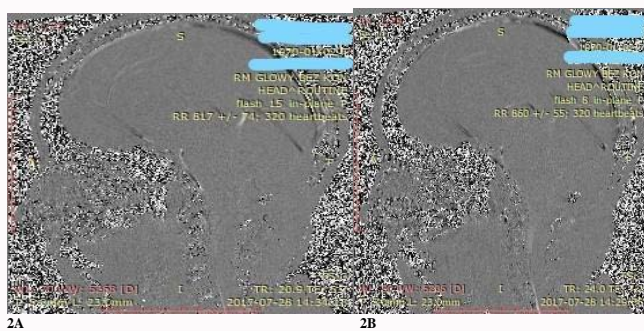

Figure 2A and 2B: Lack of the flow in the Sylvian aqueduct was observed in the systolic and diastolic phase both at the speed of 8 cm/s and 15 cm/s.

fossa. Extension of the fourth ventricle dimensions after posterior fossa surgical procedures, which was described in literature, has even gained the term "postoperative Dandy - Walker syndrome" or "trapped fourth ventricle".

The production of CSF by the vascular plexus causes enlargement of the dimensions fourth ventricle, local mass effect, dislocation of the posterior fossa structures towards the rostral and caudal direction, compression and failure of the brainstem, cranial nerve dysfunction and the symptoms of cerebellar syndrome.

The imaging studies revealed the areas of sub-ependymal leakage into the white matter similar to PVL around the enlarged fourth ventricle. The rarity of the syndrome (IFV) in adults may cause difficulties in the adequate interpretation of post-operative imaging examinations. We believe that the publication is a good training resource for all colleagues who assess patients after operations within the fluid space of the brain.

The routine imaging evaluation of the effectiveness of treatment in patients after drainage and ventricular shunts is carried out on the basis of the morphology of the lateral ventricles and the third ventricle; the assessment estimates the effectiveness of eliminating supratentorial mass effect. The radiological features of the fourth ventricle enlargement do not always raise a suspicion of IFV. We pointed out that the majority of literature publications relates to the topic of IFV pediatric patients, mainly after complicated ventricular-peritoneal valve implantations. A characteristic feature of IFV is that development of symptoms takes place long time after the first operating procedure performed within the fluid system. The course

of the condition in adults is the same; the publications concern outpatient observation of patients after the ventriculo-peritoneal shunts and are very scarce. Our study presents a unique situation when the syndrome of symptomatic mass effect caused by IFV occurred delayed after the first surgical procedure developed in the unconscious patient.

A female patient after the operation of the posterior fossa was treated for a long time in the ICU, initially by implantation of shunt into lateral ventricle. After the complications and shunt removal the external ventricular drainage was implanted. The neurological examination revealed the weak reactivity of brain stem and the appearance of the symptoms cranial nerves dysfunction. This examination prompted us to perform control radiological assessment and adequate diagnosis of deterioration.

Case Presentation

A 47-year-old patient (A.N) admitted to the ICU was treated surgically due to cavernous angioma of the vermis. Due to obstructive hydrocephalus diagnosed in the postoperative course first external drainage and then ventriculo-peritoneal shunt system was implanted. Unfortunately, the infection of central nervous system was observed. The culture revealed the presence of Staph epidermidis MRCNS. The valve system was removed; external ventricular drainage was applied along with targeted antibiotic therapy. After recovery, the valve system was re-implanted. Unfortunately, inserted shunt led in CT to the asymmetrical dilation of the lateral ventricle on the side opposite to the introduced valve drain due to the impeded communication on the level of Monroe foramen. Deterioration of the clinical condition and the impairment of consciousness assessed at 5 GCS points created the need to apply external drainage of the asymmetrically enlarged lateral ventricle and replacement of the shunt with external drainage. Then, one of the drains was removed and endoscopic septostomy and third ventriculostomy were performed. The post-operative CT scan revealed that the ventricular system was temporarily symmetrically narrowed. The patient was introduced into analgosedation and transferred to the Intensive Care Unit. However, after discontinuation of the analgosedative medication, the patient's condition did not improve, consciousness was assessed at 5 GCS points, clinically, respiratory insufficiency was reported, and artificial ventilation was continued using a ventilator.

The diagnostic and therapeutic period lasted a total of 3 months of hospital stay in the Department of Neurosurgery and Intensive Care Unit (ICU) counting from the moment of surgery on cavernous angioma. During this time, the ventriculo-peritoneal shunt was implanted and removed, the first neuroinfection was treated. Then, both implantation and removal of the shunt were repeated, and another external drainage was established followed by endoscopy procedure. Second neuroinfection was cured.

Despite intensive treatment the patient's condition did not improve. Because of respiratory failure the patient was connected to the ventilator. Ventricular drainage worked properly. The values of intracranial pressure monitored continuously in the drain were in the range from 2 mmHg to 12 mmHg. The neurological examination performed prior to the administration of analgosedation revealed that the pupils were equal (4 mm in a diameter) with a slow reaction to the light, circulatory instability manifested as tachycardia alternating into bradycardia. The examination of the state of consciousness showed the reaction of flexion of the left upper limb after pain stimulus. The

Figure 3 and 4: The MRI examination showed the displacement and tonsillar herniation.

Figure 5: The control MRI performed on the following day revealed the correct position of the drain in the lumen of fourth ventricle and decompression of anatomical structures of the posterior fossa.

analgosedation was stopped after 3 days and the neurological condition was reassessed. External drainage worked properly maintaining the pressure in the normal range. There was no improvement of the clinical condition, the patient was still unconscious, and the pupils were dilated with the slow reaction to the light and inefficient breath. Reactions to pain stimulus as before. Because of the deterioration of brainstem reactivity, an imaging examination was ordered. The computed tomography showed efficiency of lateral ventricular drainage, narrowing of the supratentorial dimensions of the ventricular system. The consulting neurosurgeon noted the cystic form with the CSF in the posterior fossa located in the midline with a mass effect and hypodense zones around the fourth ventricle (Figure 1). A suspicion was made of either the presence of pseudo-cystic enlargement of the site after the removed cavernous angioma of the vermis or disturbance of cerebrospinal fluid outflow from the fourth brain ventricle. The next MRI examination performed with the cine option revealed the enlarged fourth ventricle and lack of outflow from this structure. The occlusion of Sylvian aqueduct and isolated fourth ventricle (IFV) were detected. No flow in the Sylvian aqueduct was observed in the systolic and diastolic phase both at the speed of 8 cm/s and 15 cm/s (Figure 2a and 2b).

Additionally, Periventricular Lucency (PVL) around the fourth ventricle and atrophy of the cerebellar vermis were observed. The MRI examination showed the displacement and tonsillar herniation (Figure 3 and 4). The presence of mass effect, compression on the brainstem resulted in a decision to perform suboccipital decompression and making the connection between the IFV and the subarachnoid space. The patient was operated in a lying position. After craniectomy and posterior arch of atlas removing, dura was opened and the tightly adhering cerebellar tonsills were microsurgically separated. During this maneuver lack of free outflow of CSF from Magendie was noticed. The medial foramen was obstacled and due to the lower medullary velum was cut to improve the outflow. Post-inflammatory changes were observed within the fourth ventricle lining as an additional membrane blocking the communication with subarachnoid space. An incision was made in the midline at the distance of 7 mm long within the Magendi foramen, the lower medullary velum and the post-inflammatory membrane. The CSF flowed out under the elevated pressure, the hemispheres of the cerebellum clearly diminished their volume, they "collapsed" and began to pulse. The incision was made at a length of 7 mm to avoid of spontaneous closure caused by arachnoidal adhesions. A soft, perforated drain was also inserted through the cut line into the fourth ventricle to a depth of 8 mm, was placed and fixed followed by
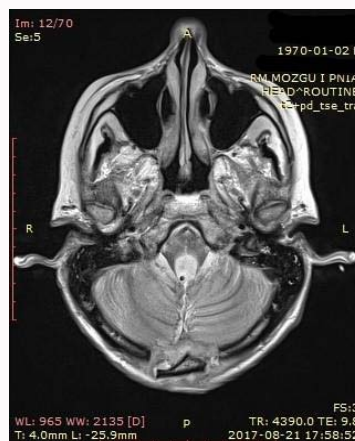
suturing to the arachnoid membrane around the Magendi foramen. The total length of the drain was 15 mm. The distal end of the drain was placed within the anatomical cerebellomedulary cistern. The free outflow of CSF via the IFV-ventriculostomy and through drain was observed. The wound was tightly stitched. The operative procedure was uneventful and patient was transported to the Intensive Care Unit. The next day after stopping the analgosedation, pupils reactivity *i.e.* light reflexes was improved and reactivity of pain stimulus reported as intensification of the facial response (facial grimace), which has not been observed so far. The control MRI performed on the following day revealed the correct position of the drain in the lumen of fourth ventricle (Figure 5) and decompression of anatomical structures of the posterior fossa. External drainage of the lateral ventricle was left. It could not be internalized due to the high levels of protein in the cerebrospinal fluid. The ICP in the range of 4 mmHg to 10 mmHg was measured. In the following days, the examination of CSF taken from the lateral ventricle revealed the next neuroinfection - *S. epidermidis* MRCNS, therefore, targeted antibiotic therapy was implemented. Unfortunately, suddenly the clinical condition significantly deteriorated and the patient died due to cardiorespiratory failure.

Discussion

Certain types of neurosurgical operations within the cerebral fluid pathways may predispose to the development of Isolated fourth ventricle (IFV). This applies to the operation of ventriculo peritoneal shunt implantation, revision and removal of shunt to infectious complications. Especially it was observed in pediatric patients. The use of time extended external ventricular drainage and the surgical procedure performed within the structures of the posterior fossa predispose to IFV as well. The postoperative condition complicated by neuroinfection, formation of arachnoid adhesions within subarachnoid spaces and post-inflammatory obstruction of the Sylvian aqueduct can play important role [1-7]. There are reports of the possible development of IFV after endoscopic ventriculostomy of the third ventricle too. Reversal flow in Sylvian aqueduct seems the reason of IFV formation [8].

IFV or "trapped" fourth ventricle encountered in pediatric neurosurgery can be a serious consequence of intracranial hemorrhage, infection or post-inflammatory central nervous system complication. The development of IFV may also occur after properly performed

ventricular -peritoneal shunt implantation. It is characteristic feature of IFV that the clinical and radiological symptoms can appear in the course properly shunting and draining supratentorial ventricles few months after the effective treatment. The development of radiographic and clinical features of IFV responsible for the mass effect in the posterior fossa and the symptoms of brainstem compression were observed after several months. The set of symptoms does not occur immediately after the surgical procedure [1-8]. IFV was described for the first time by Dandy [9].

The previous publications about adult patients with radiological feature and symptomatic IFV were very scarce and concerning outpatient clinic after ventricular shunt. Our publication presents the very rare case of IVF feature developed during long lasting in meaning several months hospital stay.

The treatment of patient after surgery in posterior fossa and shunt therapy was complicated by infection and finally admitted to ICU. The patient was in a severe clinical condition during the stay in ICU. The insight clinical examination was difficult due to deep unconsciousness and brainstem dysfunction. We observed the increasing of brainstem dysfunction and the lack of improvement clinical status during hospitalization. Our suspicion of deterioration was related to the possibly direct compression of the brainstem in the IFV mechanism. Therefore, we decided to extend the scope of diagnostic imaging. In our opinion, this was unique in this publication to perform the Cine Sequence MRI to confirm the lack of CSF flow through the Sylvian aqueduct in the systolic and diastolic phase of examination. This observation revealed the important factor of the IFV pathogenesis. It should be emphasized that in unconscious patients the lack of improvement or deterioration after above mentioned surgical procedures can be caused by IFV. Therefore, so it is very important to carefully observe brainstem symptoms.

During the debate about the pathophysiology of CSF in the context of IFV is present a view about low significance of vascular plexus in fourth ventricle for the production of CSF. In our opinion, this is a myth, especially when radiologically and clinically IFV is developed. In certain situations, the secretory activity of the plexus leads to the features of mass effect in the posterior fossa. The vascular plexus of the fourth ventricle has rich vascularization and is supplied by AICA to lower and lateral segment and PICA to the medial segment [10]. In our opinion, it is important the knowledge of the operational anatomy of this area because this may prevent from the formation of postoperative CSF reservoirs connected with the fourth ventricle, causing Pseudo Dandy-Walker syndrome. This was described in the 60's of the last century [6,7].

Some neurosurgeons, who did not face the problem of IFV due to the rarity of the syndrome, can believe that the production of CSF in the fourth ventricle is not relevant. The literature and clinical practice provide the information that consultants performing control CT scan after ventricular shunt therapy pay attention mainly to the assessment of supratentorial space. They analyze the efficiency of draining the ventricular system by evaluating only the dimensions of the lateral ventricles and the third ventricle, and ignoring the presence of IFV development. In this way they can ignore the possibility of fourth ventricle enlargement and incorrectly evaluate the neurological symptoms from posterior fossa and brain stem. The isolated fourth ventricle can be incorrectly interpreted as a site after the surgery in the area of the posterior fossa too. We recommend that different consultants should pay attention to this in their work, especially when

they base only on descriptions of radiological imaging. Sometimes, before a neurosurgeon final decision, the cumulative symptoms of IFV are incorrectly interpreted as post-traumatic syndrome or toxic effect of drugs [4,11,12]. Symptoms of brain stem compression and mass effect of the posterior fossa caused by IFV requires surgical treatment. In adults, this pathology is very scarce, but treatment strategies developed for pediatric patients can also be used in adults [13,15-19].

The discussion on the IFV treatment referring to microsurgical restoring patency to the outflow routes from the fourth ventricle [13,14] or endoscopic fenestration of IFV, surgery of the Sylvian aqueduct aqueductoplasty, introduction of the stent into the aqueduct or drainage IFV peritoneal shunt. Due to the high risk of complications, surgical techniques are still improved. Actually in the management most often is used the IFV peritoneal shunt and aqueductoplasty of Sylvian aqueduct with a stent [20-23]. The aqueductoplasty without stenting may cause re-occlusion or obliteration [24], additionally navigated endoscopy techniques guarantee the precision of stenting [16,25].

I our clinical situation due to serious symptoms of mass effect in posterior fossa, we decided to perform suboccipital decompression and microsurgical patency restoring to the outflow routes from the fourth ventricle and create the connection between IFV and fluid spaces. The aim of our decision was to decompress the brainstem following radiologically diagnosed brain herniation and directly eliminate the cause of IFV. In this condition the method of IFV peritoneal shunt, should rule out. On the other hand the high level of protein in CSF can disturb the patency of drainage and can irritate the peritoneum additionally.

During the operation I decided to leave a small fragment of the drain inserted into the IFV lumen. I stitched it with the arachnoid membrane to avoid the dislocation. I measured the length of drain to avoid symptomatic irritation of the bottom of the fourth ventricle. The management to establish IFV peritoneal shunt has the risk of complications, such as over drainage, occlusion of the shunt, infection, mechanical irritation of the brainstem (fourth ventricle floor) and the symptoms of cranial nerve and brainstem dysfunctions.

The aqueductoplasty maneuvers are associated with complications, such as post-inflammatory or post-infection occlusion and may require revision. The cranial nerves alterations observed after IFV peritoneal shunt are evoked by direct irritation of the floor of fourth ventricle by the tip of inserted drain and as a result of brainstem dislocation and require revision [11,15,26,27]. The above mentioned mechanism of cranial nerve dysfunctions and brain stem alteration may also result of the overdrainage due to siphon effect of the inserted shunt in the both situation supratentorial ventricle-peritonal and IFV peritoneal as well. This phenomenon can provoke to collapse of the Sylvian aqueduct walls as well and deformation of the brainstem due to changes of the pressure [26-29].

The pathomechanism of the obstacle for reverse flow of CSF through the Sylvian aqueduct from the fourth to third ventricle followed the lateral ventricle-peritoneal shunt can refer the suction mechanism and is associated with CSF flow disturbances on the level of the cisterna ambiens and the upward herniation of the vermis of the cerebellum into the tentorium edges. The observed mechanism of positive feedback causes that the increasing IFV volume creates additional compression around the Sylvian aqueduct followed by herniation into the tentorium edges. The difficulties of CSF outflow

from IFV via anatomical foramina causes that the expanding dimensions fourth ventricle creates the mass effect leading to "upward herniation" and collapse Sylvian aqueduct followed compression and dislocation of anatomical structure within tentorial edges.

The functional mechanism of the IFV development due to the pulsating wave is described in the literature. According to the dynamic theory of CSF circulation, the flow depends on pulsation of the arterial vessels and giving the speed of the flow of CSF in a given direction. The pulsating wave is mechanical and reverberates along the ventricular system. The fourth ventricle is located at the end of this route; the mechanical wave is reflected as a result of pulsation and occurs constructive interference which has an impact on the ventricular walls being anatomical structures of the brainstem.

It may be possible the mechanism of constructive interference of the pulsating mechanical waves is responsible for the development of so called "functional trapped fourth ventricle" described in the literature and characterized by no mechanical obstacle inside of Sylvian aqueduct observed during aqueductoplasty [8,30,31].

Conclusion

The presented clinical case of IFV which was diagnosed during the long-term ICU stay is the only one in the literature. The mechanism of the alteration can have a significant impact on the lack of success in therapy due to brain stem compression and mass effect in posterior fossa. The adequate diagnosis of IFV is crucial. It is very important to observe the unconscious patient for possible brainstem dysfunction during long lasting hospital stay in ICU and perform sequential brain image monitoring. According to publications, we can anticipate development of IFV after neurosurgical activity in CSF space, CNS infections, post inflammatory hydrocephalus and external drainage, bleeding to the posterior fossa, complications after the ventricle-peritoneal shunt operation or after prolonged external drainage in supratentorial spaces. In ICU the monitoring of the brainstem function and cranial nerve symptoms is especially important, in the context of pharmacotherapy, including sedation, the use of opioids and muscle relaxants.

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