Case Report

Relocation of ventricular catheter trough ventriculostomy due to congenital unilateral hydrocephalus: Nine year follow-up

Milenkovic J. Zoran, Stevanovic S. Biljana[1], Markovic P. Ivana[2]

Department of Surgery, General Hospital "Sava Surgery" Bulevar Zorana Djindjica, [1]Ophthalmologic Clinic , Clinical Center Niš , Bulevar Zorana Djindjica 48, [2]Center for Radiology, Clinical Center Niš , Bulevar Zorana Djindjica 48 ,Serbia

E-mail: *Milenkovic J. Zoran - zoran@ni.ac.rs; Stevanovic S. Biljana optokontakt@yahoo.com; Markovic P. Ivana - ivanadidi@yahoo.com
*Corresponding author

Abstract

Background: Congenital unilateral hydrocephalus is an uncommon entity occurring almost exclusively in children. Atresia, stenosis, membranous occlusion and even functional obstruction of the foramen of Monro have been described to be the main cause of this type of hydrocephalus. There are two options available in the surgical management of unilateral hydrocephalus: one is the placement of shunt CSF diversion from the dilated ventricle and the other is fenestration of the occluded foramen of Monro or septum pellucidum by endoscopy or by stereotactic method. Migration of the ventriculoperitoneal (VP) shunt in or out of ventricles is not so uncommon, but the relocation of the ventricular tip of a catheter from the ventricle into the quadrigeminal cisterns and superior vermis in association with ventriculostomy is extremely rare. Spontaneous ventriculostomy is a rare event and results from spontaneous rupture of a ventricle into the subarachnoid space.

Case Description: A 5½-month-old baby with a right-sided congenital unilateral hydrocephalus underwent a VP shunt andhad experienced an uneventful outcome. Four years later on an MR imaging examination, the tip of the ventricular catheter passing through the medial wall of the ventricle and the quadrigeminal cistern was found to be situated in the superior vermis. During the follow-up period, there were no neurological difficulties. The cognitive and motor skill development corresponded well with the child's age. It transpired that the hydrocephalic ventricle reduced its size dramatically to normal.

Conclusion: We have described the extremely rare site of the relocation of the ventricular catheter after the treatment of the congenital unilateral hydrocephalus by VP shunting. Spontaneous ventriculostomy as a rare phenomenon may be the explanation of the relocation of the ventricular catheter.

Key Words: Congenital unilateral hydrocephalus, ventriculoperitoneal shunt, ventriculostomy

INTRODUCTION

Migration of the ventriculoperitoneal (VP) shunt in or out of ventricles is not so uncommon, but the relocation of the ventricular tip of a catheter from the ventricle into the quadrigeminal cisterns and superior vermis in

association with ventriculostomy is extremely rare and we have not been able to find any case in the currently available literature. We are presenting the case of an infant harboring a unilateral congenital hydrocephalus who was treated with a VP shunt. Almost 4 years later, the tip of the ventricular catheter, passing through the tiny ventricular wall, was found to be situated in the superior vermis. On the other hand, spontaneous ventriculostomy is a rare event and results from the spontaneous rupture of a ventricle into the subarachnoid space. Thereare only four reports regarding this phenomenon at the level of the lateral ventricle.[1,4,7,12]

CASE REPORT

A 5½ month-old infant was referred to a neurosurgeon for consultation due to congenital unilateral hydrocephalus detected on MR imaging investigation. From the discharge list of neonates issued by the Pediatric Clinic of the Clinical Center of Nis, Serbia, we were able to find out the course of the disease, outlined below. At 37 weeks of gestation, a boy weighing 3759 g, with an Apgar score of 9 at 1 min, was delivered after prolonged labor on 24th April 2002. Several ultrasound examinations had been performed during pregnancy and all were described as normal except for the last one that was done in the 37th week of gestation, which showed an enlargement of the right lateral ventricle compressing the left one and moving the interhemispheric fissure to the left. The third and forth ventricles were found to have a normal size. The second ultrasound done immediately after the delivery was identical. After birth, his head circumference was found to be 36 cm. The neonate was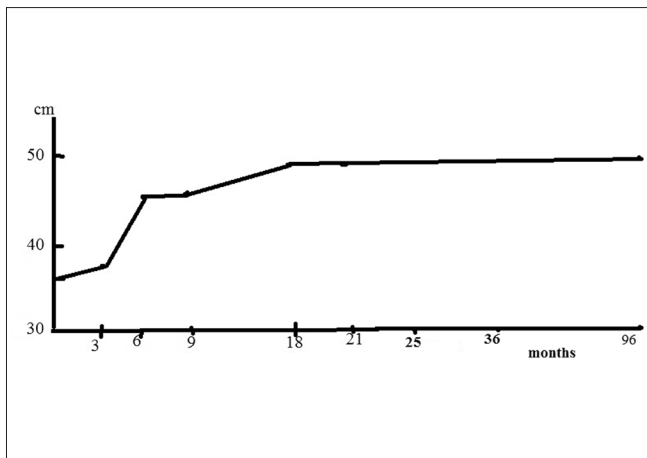 under thesupervision of a neonatologist and was transferred to the Pediatric clinic due to high fever with signs and symptoms of pneumonia. The infant spent more then 1½ months, being closely observed and treated by pediatrician and neurologist. The neurosurgeon suggested the operation, but the child's parents refused it, believing that the intervention would not bring any benefit. The child was discharged as a severely handicapped hydrocephalic baby with poor prognosis and was referred to pediatrician and neurologist in the outpatient clinic. For the next 4 months, the infant was followed up by them. The parents changed their opinion and the child was referred to the neurosurgeon for further treatment.

On admission, there were slight delays in cognitive and motor development. The neurological examination revealed an inconclusive left side weakness. The MR investigation showed a severe dilatation of the right ventricle, with the atrial diameter over 34 mm and evident midline shift to the left [Figure 1]. There were initial signs and symptoms of hydrocephalus slightly tense anterior fontanelle and splayed sutures. Fundoscopy was normal. The head circumference was 47 cm at the beginning of the sixth month. Having had no endoscopy at that time, the VP shunt was accepted as a reasonable option and the baby was operated at 6 months after delivery. The NMT neurosciences' gravity-compensating lumboperitoneal valve system consisting of the original Hakim valve with a ball-in-cone mechanism in a silicon elastomer chamber was used. The postoperative course was uneventful. The size of the skull corresponded well with the infant age [Figure 2], as well as the cognitive and motor development. The ultrasound examination performed 1 month after VP shunt revealed an enlargement of the right ventricle of 27 mm in size with the ventricular
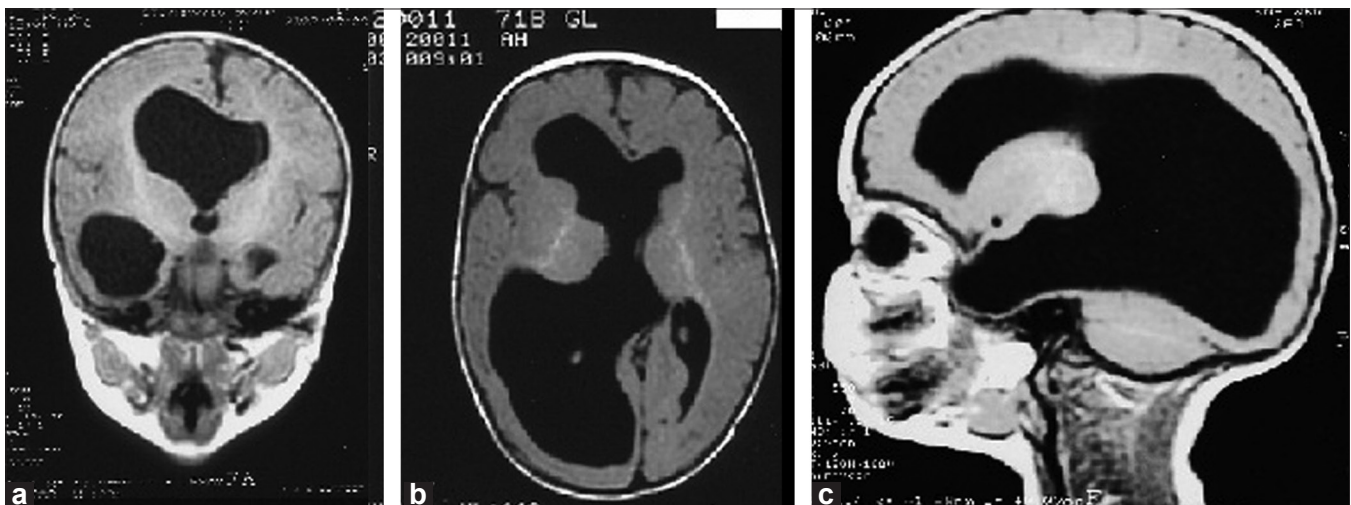

Figure 1: Coronal T1w image SE 540/14 (a) shows the severe dilatation of right lateral ventricle and consequent compression and loss of brain tissue in the right temporal and occipital region. Axial FLAIR T1 2200/35 (b) presents the enlargement of right atrium and occipital horn with reduced volume of periventricular and subcortical white matter. Parasagittal T1w image SE 550/20 (c) depicts the marked enlargement of the right lateral ventricle with prominent compression and lack of brain parenchyma volume, predominantly in the temporo-occipital region

drainage system in the right ventricle. Both the third and fourth ventricles were normal in size. In the follow-up period, there were not any major or mild neurologic disturbances. The evident reduction in ventricular size was clearly presented in the first postoperative MR done 6 months later. The atrial diameter was 23 mm [Figure 3]. For 1 year or so, the child was under pediatric and neurosurgical observation that registered normal cognitive and motor development and no neurologic deficit. The VP shunt was functioning well. The head circumference was corresponding with the child's age [Figure 2]. After this, the referring neurosurgeon was away for 2 years, and therefore had no opportunity to observe the patient. During this period of time, the child was under pediatric and neurologic care. His mental and physical capabilities were completely normal and they perfectly fitted to the child's stature. Surprisingly, MR images taken 5 years after the operation showed the tip

of the ventricular catheter to be situated in the superior vermis. The right atrial diameter was evidently smaller measuring 17 mm [Figure 4]. In the following period, the child was doing fine without any mental or neurological disturbances. After that, the MR was performed three times which presented the evident, dynamic shrinkage of ventricle, and the last one was performed 2 months ago which showed the normal-sized ventricles with the catheter's tip in the same position [Figures 5 and 6].

DISCUSSION

Spontaneous ventriculostomy is a rare event and results from a spontaneous rupture of a ventricle,

Figure 3: (a, b) Coronal, axial and sagittal T1w SE 500/14 images show initial/incipient decompression (and decreased size) of the right lateral ventricle after shunting (right transparietal catheter *in situ*). Rim of brain parenchyma in the right temporo-occipital region is thicker, consequent to the mild increase of brain volume. Also, there is consequent mild atrial diverticulum and displacement of mediosagittal structures. Right frontal horn is slightly rounded, while the left lateral ventricle is completely decompressed

Figure 2: The head circumference evolution: at birth was 36 cm; at 3 months -37 cm; at 6 months -47 cm; 9 months later -47,5 cm; at 21 months -49,5 cm. Now is 50 cm
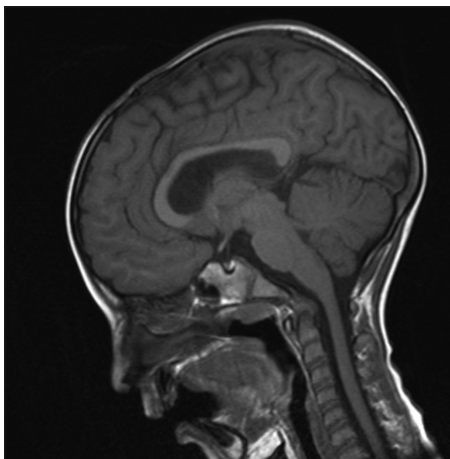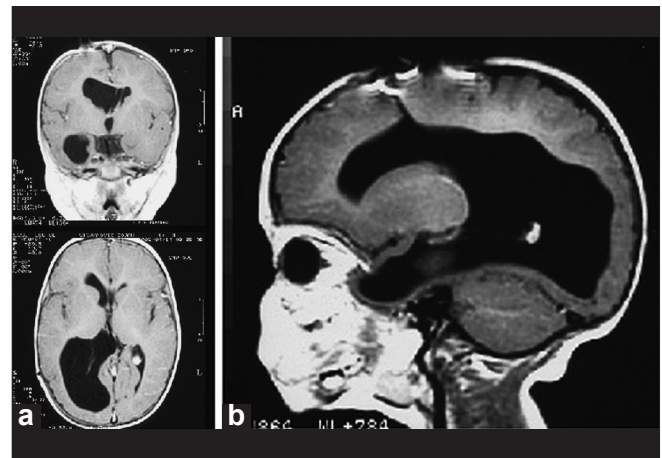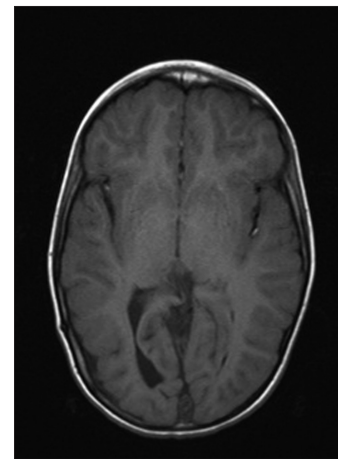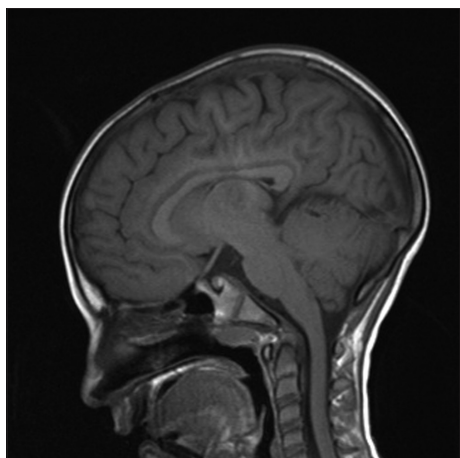

Figure 4: The second and the third postoperative MRIs present further decompression of lateral ventricle and increase in brain volume

Figure 5: The position of catheter seen in axial image (SE 500/7.8) is better visualized on sagittal and coronal planes within different sequences

Figure 6: Sagittal (SE 500/7.8) image shows catheter as a short inferior pericallosal signal void with a tip in the anterior cerebellar lobe and vermis

generally at the level of the medial wall of the trigone or at the pineal recesses of the third ventricle into the subarachnoid space, but is usually seen in patients with chronic obstructive hydrocephalus. This rupture results in direct communication of the ventricular system with the subarachnoid space.[1,8-10] Previously reported cases were noted to have occurred secondary to obstructive hydrocephalus caused by neoplastic disease or by benign stenosis of the aqueduct, mostly in adults.[2,5,7,8,10,11] There were a few reports of spontaneous ventriculostomy in infants due to aqueductal stenosis involving the third ventricle[3,6] and only one occurring as a consequence of congenital stenosis of the foramen of Monro.[1] Spontaneous ventriculostomy may occur in a number of locations, but only four reports refer to the atrium of the lateral ventricle.[1,4,7,12]

Only two children with congenital unilateral hydrocephalus due to stenosis of the foramen of Monro had paraventricular diverticula and spontaneous ventriculostomy. Both patients had a cystic space in the quadrigeminal and supracerebellar cistern.[1] Spontaneous ventriculostomy can be demonstrated by ventriculography, cine MRI, and endoscopic procedures.[4,7,8,10,12] In our case, the migration of the ventricular tip of catheter into the upper vermis, passing through the quadrigeminal cistern, has established the functional communication between the ventricle and subarachnoid space. It has been stated that spontaneous ventriculostomy usually occurs in patients with chronic obstructive hydrocephalus.[8,10] The importance of chronic, longstanding, high-pressure CSF pulsation has been the explanation in establishing the ventricular stoma,[8] although there were no signs and symptoms of chronic obstructive hydrocephalus in our patient. The explanation might be similar where the constant pulsating motion of ventricular CSF as the result of the hearth action serves as a favorable factor in the

rupture of the thinnest segment of the ventricular wall into the subarachnoid space. The tip of the ventricular catheter simply passed through the acquired opening.

The possibility of the overdrainage as a pathophysiological mechanism of the relocation of the catheter cannot be excluded. Overdrainage can occur normally with postural changes, REM sleep and straining. The gravity effect increases the differential pressure across the valve, keeping it open and allowing more CSF drainage. The effects of siphoning depend on multiple factors, occurring to a greater or lesser degree depending upon the type of valve implanted and present conditions. Ventricular overdrainage occurs when siphoning happens chronically and may in turn cause the brain to pull away from the inner surface of the skull, tearing the bridging scalp veins and causing associated bleeding and brain compression, debilitating headaches and slit ventricles and may also induce symptomatic orthostatic hypotension, subdural CSF collections, slit ventricle syndrome, craniosynostosis, loculation of the ventricles and so on. But we were not able to confirm any signs or symptoms related to overdrainage in our patient.

The clarification may lay in application of the NMT neurosciences' gravity-compensating lumboperitoneal valve system with a ball-in-cone mechanism. This type of valve adds resistance when an individual stands through the weight of several stainless steel balls on a ball-in-cone mechanism. Flow is not restricted when an individual is lying down. Ball-in-cone valves are less prone to the effects of the aging of materials than are miter or slit valves, and they have been demonstrated to handle higher CSF protein levels. This advantage reduces the siphoning effects of the valve. The relocation of ventricular catheter is supposed to be followed by its occlusion which is usually followed by increasing ventricular size confirmed on CT scan or MR imaging. But there has not been increasing ventricular size in our case, in spite of the migration of the catheter outside of the ventricle. Moreover, the shrinkage of the ventricle had taken place during the following period. The communication between the right lateral ventricle and the supracerebellar cistern can be one of the reasonable contexts. This is the reason why we have postulated the option of existing ventriculostomy which enables direct communication of the right ventricle with subarachnoid cisterns, providing dynamic CSF circulation in the brain.

To our knowledge, this is the first report describing the migration of the ventricular tip of the catheter into the upper vermis through ventriculostomy.

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