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Case Report OMICS International

Diagnostic Difficulties in Slit Ventricle Syndrome

Mateusz Szylberg[1]*, Jacek Furtak[1], Mateusz Szylberg[1], Tomasz Slotala[2] and Marek Harat[1]

[1]*Department of Neurosurgery, 10th Military Hospital, Bydgoszcz, Poland*
[2]*Department of Clinical Neurology at 10th Military Research Hospital and Polyclinic in Bydgoszcz, Poland*

Abstract

Slit ventricle syndrome is a rare complication of the treatment of hydrocephalus in infancy. Symptoms usually appear several years after the placing of the shunt system. They can cause diagnostic difficulties. The diagnosis is usually wrong or delayed. The authors describe a patient with SVS, the course of hospitalization and examinations performed.

Keywords: Slit ventricle syndrome; Normal volume hydrocephalus; Hydrocephalus; Shunt complications; Over drainage

Introduction

Shunt system treats childhood hydrocephalus. This treatment has many complications. The major groups of complications are obstructions, mechanical failure of the shunt, infections and overdrainage of cerebrospinal fluid. Overdrainage of CSF has been reported in 5% to 55% of shunted patients [1,2]. Main clinical symptoms of slit ventricle are: headache, nausea, vomiting, and disturbances of consciousness, hemiparesis, and epileptic seizures [1]. Patients with Slit Ventricle have increased risk for further shunt complications. Slit Ventricle Syndrome has become a possible complication of cerebrospinal fluid shunting procedure. While its etiology and pathophysiology are poorly understood, it appears to be a manifestation of over shunting [3]. Authors reported a case to explain the origin of the symptoms of narrow ventricles [4].

Case Report

A 24-year-old man admitted (30th of January) to the Neurology Clinic because of severe headache. In addition, there were paroxysmal incidents of limbs convulsions with consciousness disorders. The patient was previously (15th of January) hospitalized for a seizure of a generalized nature. The neurologist diagnosed epilepsy and included valproic acid. In medical history, there was ventriculo-peritoneal shunt placed due to hydrocephalus in fifth month of life. At that time CT images of the head revealed a significant dilation of the lateral ventricles (Figure 1). At the age of three controls CT scan showed regression of hydrocephalus (Figure 2). At the age of 12 was qualified for shunt revision, an exchange of peritoneal catheter, due to increased ICP syndrome.

Now due to the traits of the increasing intracranial pressure such as psychomotor slowdown, severe headache, diplopia and meningeal signs, bilateral malfunction of eye movement nerves, paralysis of the facial nerve on the right side and a discreet paralysis on the left. The ophthalmic examination showed papilloedema in both eyes. The patient was submitted to the Department of Neurosurgery (1st of February) to verify the operation of the valve system. The MR imaging of the head found slightly enhanced frontal horns of the lateral

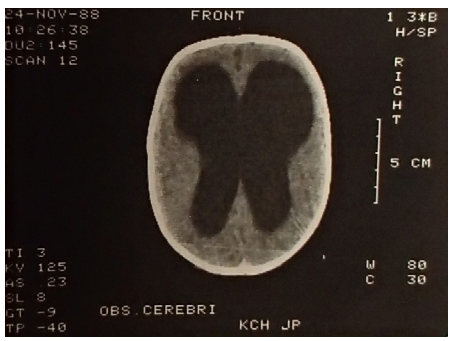

Figure 1: At the age of three control CT scan showed regression of hydrocephalus.

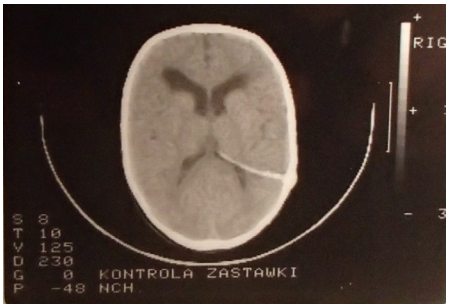

Figure 2: At the age of 12 was qualified for shunt revision, an exchange of peritoneal catheter, due to increased ICP syndrome.

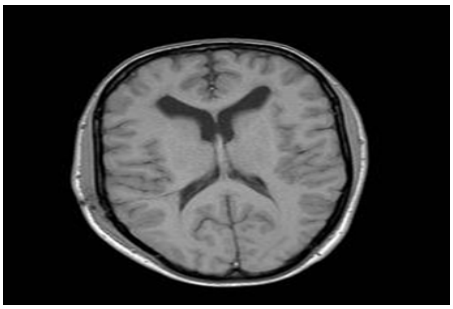

Figure 3: Slightly enhanced frontal horns of the lateral ventricles with CSF penetration traits.

***Corresponding author:** Mateusz Szylberg, Department of Neurosurgery, 10th Military Hospital, Bydgoszcz, Poland, Tel: +86159-2806-3038; E-mail: mateuszszylberg@op.pl

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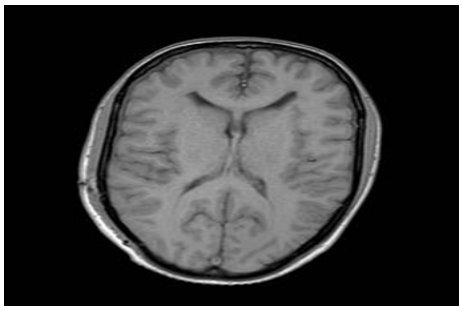

Figure 4: Regression of dilation of the lateral ventricles.

ventricles with CSF penetration traits (Figure 3). Moreover, the other ventricles were narrowed. The patient was qualified for the removal of ventriculo-peritoneal shunt because of suspected infection (2nd of February). The cerebro-spinal fluid was taken for microbiological examination, the result was negative. After vetriculo-peritoneal shunt removal, the condition of the patient was significantly deteriorated (3rd of February). The neurologist observed severe headache with a medium response to pharmacological treatment. Moreover, there were increasing signs of meningeal syndrome and numerous simple partial seizures of convulsions evolving to secondarily generalized seizures. The neurologist administered intensive antiedematous treatment and antibiotics. CT and MR imaging (5th of February) of the head after shunt removal did not reviewed any radiographic changes compared to previous studies. The ventricles were narrowed. In further hospitalization patient revealed signs of brain stem syndrome with malfunction of nerve VI, right VII, left hemiparesis and consciousness disorders. After exclusion of infection of shunt system, the patient was immediately qualified for surgical implantation of ventriculo-peritoneal shunt to the front right corner of the lateral ventricle (10th of February).

After next surgical procedure, a significant improvement in neurological status was observed. The patient fully recovered from headache, gradually withdrew malfunction of eye movement nerves, decreased diplopia. After few days the left hemiparesis withdrew, patient moves independently and efficiently. In a control CT after implantation of a ventriculo-peritoneal shunt image of the brain and the width of the ventricular system did not change significantly. The MR in comparison with the previous study showed regression of dilation of the lateral ventricles (Figure 4). There was no evidence of increase of intracranial pressure.

Discussion

Authors present a case of a male patient with unusual increase intracranial pressure with slightly enhanced lateral ventricles and problems connected with this diagnosis. Intracranial hypertension with nondistending ventricles was originally described by Engel et al. They called it normal volume hydrocephalus (NVH) [5]. In the publications of Engel et al., there were patients with signs and symptoms of increased ICP without enlarged ventricular system. They recommended exploration of these shunts that were malfunctional [5]. These patients eventually have progressive symptoms of increased ICP such as morning headaches leading to headaches throughout the whole day, papilledema, loss of vision and diplopia. If this condition is not treated early, it may possibly lead to a neurological deterioration, which can lead to blindness. This is a common problem in cases of congenital hydrocephalus [6]. Normal volume hydrocephalus is defined as elevated ICP without enlargement of ventricles when the valvular system does not work properly. Normal volume hydrocephalus develops as a complication of shunt implantation in childhood. NVH does not present when the hydrocephalus occurs in adulthood [3]. Numerous complications after shunt implantation require from neurosurgeons a lot of experience and a constant readiness to shunt revision in the event of malfunction. The surgeon must deal with infections, obstruction and overdrainage. The overdrainage among some patients can appear as slit ventricle syndrome [7,8]. In the author's case, ventricles were enlarged only before the first shunt implantation. After that all imaging studies presented narrow ventricles despite the obvious symptoms of intracranial hypertension. In some cases, the authors observed symptoms of high intraventricular pressure, while CT images of the head revealed that the ventricles were narrowed. The reason of this phenomenon may be a chronic change within the lining of the brain causing the stiffness [9-11].

Symptoms may suggest encephalitis but the CSF test excluded this diagnosis. The patients who had hydrocephalus in infancy may develop pseudotumor cerebri in adulthood [12]. Pseudotumor cerebri is a state with raised ICP and small ventricles that can be managed by CSF draining. Thus, older children and adults with shunt failure whose ventricles stay narrowed they no longer have hydrocephalus [3]. In some opinions, they have a form of pseudotumor cerebri that may be dangerous and more difficult to manage than pseudotumor cerebri that begins in adulthood [12]. In fact, slit ventricle syndrome is often misdiagnosed or the diagnosis is delayed [12].

Conclusion

The level of recognition of this state among general neurosurgeons, radiologists and neurologists is relatively low. Therefore, these patients are at high risk before the final proper diagnosis of Slit Ventricle Syndrome. Imaging studies (CT scans) have been interpreted as "no evidence of hydrocephalus" or "no evidence of shunt failure". The finding of small ventricles in a shunted patient can be misinterpreted as a properly working shunt. The patients with complete shunt failure often have prolonged periods of suffering and occasional blindness with no evidence of ventricular dilatation [12]. Slit ventricle syndrome is a rare symptomatic condition and adequate treatment may be delayed [13]. Slit ventricle syndrome can be dangerous condition for the patient. It should be kept in mind by neurosurgeons.

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