

Case Report

Low-pressure hydrocephalus: A case report

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- Cerebrospinal fluid leak
- Transmantle pressure gradient
- Gravitational valve

Abstract

The underlying pathophysiology in low-pressure hydrocephalus (LPH) includes brain viscoelastic properties, brain turgor, transmantle pressure gradient and cerebrospinal fluid leaks. LPH consists of ventriculomegaly with low or negative intracranial pressure values. It is a rare entity and it can appear after hydrocephaly ventriculoperitoneal shunts, subarachnoid or intraventricular hemorrhage, cranial traumatism, lumbar puncture, meningitis among others. LPH clinical manifestations include: headaches, low level of consciousness, vomiting and cranial neuropathies, similarly to those occurred in high intracranial pressure hydrocephaly. A better understanding of LPH mechanism may help us to optimize treatment, as its diagnosis and management are complex.

ABBREVIATIONS

CSF: Cerebrospinal Fluid; NPH: Normal Pressure Hydrocephalus; LPH: Low Pressure Hydrocephalus; ICP: Intracranial Pressure; VPS: Ventricular Peritoneal Shunt; SAH: Subarachnoid Hemorrhage; ME: Multiple Sclerosis; CT: Computed Tomography; TBI: Traumatic Brain Injury; EVD: External Ventricular Drain; EC: Ear Canal; GSC: Glasgow Coma Score

INTRODUCTION

Hydrocephalus is a pathological increase in the CSF volume, regardless of hydrostatic or barometric pressure [1]. Normal pressure hydrocephalus (NPH) triad includes gait disorder, cognitive impairment and urinary incontinence associated with ventriculomegaly. The clinical outcome associated with LPH is defined as a distinct disease entity with typical pathophysiological characteristics. LPH was described in 1994 by Pang and Altschuler [2] who mentioned the role of the viscoelastic properties of the brain in the pathogenesis of the disease. Filippidis et al [3] reported that Cerebrospinal Fluid (CSF) fistulae are the main cause of LPH.

LPH ventriculomegaly is characterized by low or negative values of intracranial pressure (ICP) [2, 3]. LPH may occur in 1 in 375 ventricular peritoneal shunt (VPS) by hydrocephalus and has been described in patients with SAH, intraventricular hemorrhage, cranial trauma, spinal tap, meningitis, parasitosis, posterior fossa tumors, congenital aqueductal stenosis, spinal arachnoid cysts, hemispherectomy and NPH [3, 4]. The clinic of LPH includes: headache, decreased level of consciousness, vomiting and cranial neuropathies similar to the symptoms that appear in hydrocephalus with intracranial hypertension [2, 3].

CASE PRESENTATION

50 year old male with a history of Multiple Sclerosis (MS),

diencephalic arteriovenous malformation treated with radio surgery and aneurysm of the anterior communicating artery treated with embolization.

Four years after embolization, the patient had a sudden onset of headache, bradipsiquia and unstable gait. A computerized tomography (CT) showed a hydrocephalus not present in the images of the past two years. An arteriogram revealed recanalization of the aneurysm (15 mm long and 6 mm neck). It was scheduled for the same clipping and evolved satisfactorily during the postoperative period.

Within one month, the patient was admitted in the emergency room due to rhinoliquorrhea, fever, headache and cognitive deterioration. A CT scan showed tension pneumocephalus and after a consult to neurosurgeon he suspected a frontal CSF leak, which required emergency surgery.

One month after discharge, he suffered a traumatic brain injury (TBI) with subarachnoid hemorrhage (SAH) (Figure 1). The CT scan revealed a chronic arrearabsortive ventriculomegaly and a chronic left frontal subdural collection sheet. The neurosurgeon decided to place a low medium pressure VPS CODMAN® type. The surgery was not enough and the clinical course was torpid, with bradipsiquia, fluctuating level of consciousness with occasional episodes of psychomotor agitation and hallucinations, accounted for psychiatry treatment. On the tenth day after the placement of the VPS, the patient presented a fever peak of 39.5°C and stupor us state. Suspecting meningitis it was decided to puncture the reservoir obtaining a turbid CSF. The CT showed a small brain hematoma in the insertion of ventricular catheter, a clot in the tip and an increased hydrocephalus compared to the previous CT. Therefore, the VPS was removed and contra lateral external ventricular drainage (EVD) was placed instead (Figure 2a). In the cultivation of the explanted material a *Staphylococcus hominis*,



Figure 1 Computerized tomography performed at hospital admission. Triventricular dilatation with bilateral frontal transependymal reabsorption findings suggesting hydrocephalus, perimesencephalic subarachnoid hemorrhage (arrow). Left temporofrontal subdural collection (dotted arrow).

Staphylococcus epidermidis and one Propionibacterium were isolated, confirming the diagnosis of polymicrobial ventriculitis. During the following days after the start of antibiotic treatment, the fever subsided and a very low output pressure of CSF was observed, it was necessary to maintain the EVD 10 cm below the ear canal (EC) so it could drain CSF, a fact that coincided with neurological improvement of the patient. Therefore, throughout over more than a month during his stay in the intensive care unit, a test of CSF drainage was performed against gravity with negative pressures beginning from -10 cm from the EC and progressively rising every 2-3 days until the patient was able to tolerate neurologically pressures above zero. During the patient admission five EVD changes were made due to its detachment or obstruction. The EVD manipulation contributed to the air inlet causing the appearance of a pneumo ventricle (Figure 2b). Finally, after finding successive negative CSF cultures and an absence of inflammatory and/or infectious markers, a permanent programmable Miethke proSA® Aesculap EVP was implanted at an initial pressure of 0 cm H₂O, and the former EVP was removed. The patient was discharged from the postoperative care unit 24 hours after surgery.

Ten days afterwards, he suffered from orthostatic headache and clinical over-drainage, requiring an upward adjustment of the EVD programming, rising the pressure 2 cm above the estimated CSF drainage threshold. The subsequent evolution was favorable with a clear neurological and radiological improvement (Figure 3).

At the time of discharge (2 months after implantation of the gravitational valve), the patient had a score of 15/15 on the GCS, he was disoriented and he had a discrete motor aphasia and unsteadiness of gait, but managed to walk with help.

DISCUSSION

Despite being a rare entity [3], the clinical suspicion of a LPH increases as a deterioration appears in a patient with an initial asymptomatic ventriculomegaly, poor drainage of CSF despite the presence of an EVP, and low intracranial pressure (up to -10 cm H₂O). Several mechanisms have been described [3,5-7] in an

attempt to explain the pathogenesis of LPH: from the ventricular hysteresis given by Lesniak et al [4] and brain turgor [8], through the debated roll of transmural pressure [9] and the existence of cortical subarachnoid cortical space [7]. The latest theory has been described by Filippidis et al [3] that justifies LPH by a persistent CSF loss.

The ventricular hysteresis is the property of the ventricular system by which a value of the ICP may correspond to two different states in the ventricular volume. On the one hand, a sharp increase in ICP due to increased CSF volume would gradually produce a ventricular enlargement due to lack of absorption of CSF (disjunction of ependymal cells). Alternatively CSF production would be greater than the leak thereof, which can be or not continuous through the fistula. Anatomical recovery can be achieved thanks to the hysteresis of ependymal tissue and for this reason the height test is performed. When the ICP decreases due to CSF drainage, ICP reaches a lower or normal value within an increased ventricular volume, even requiring negative pressure in the ventricles to regain a normal size. CSF fistulae contribute to development of a LPH causing a very low subarachnoid pressure. The difference between a greater ventricular pressure and this lower subarachnoid pressure generates a transmantle pressure gradient between brain and ventricular surface which progressively determines histological changes that changes the size of the ventricles. Eventually, a new equilibrium with a constant ratio between output and CSF leakage is established, with consequent increase of the ventricles and the reduction of the subarachnoid space, reaching a zero transmantle pressure gradient.

In this case, ventriculomegaly with low ICP would be conditioned by the different processes that suffered this patient (SAH, cranial radiotherapy, ventriculitis and various EVD replacements), and MS. The neurological symptoms resemble hydrocephalus situations with increased ICP. Placing an EVD failed to decrease the ventricular size and neurological symptoms continued to fluctuate. Chronic ventricular dilatation (found in previous studies), suggested ependymal tissue stiffness.

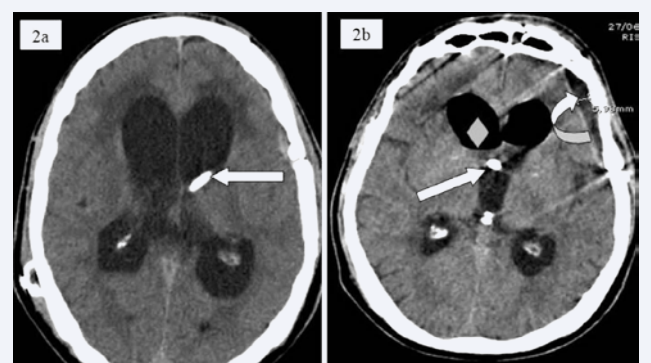


Figure 2 Day 10 after admission a new computerized tomography (CT) showed persistent hydrocephalus (A) despite the ventricular peritoneal shunt (arrow) is observed. Day 20 after admission a sudden low level of conscience and a new CT (B) showed a bilateral pneumoventricle (diamond) and persistence of the subdural collection (curved arrow) and the new external ventricular drainage (arrow) changed due to ventriculitis suspicion and shunt obstruction.

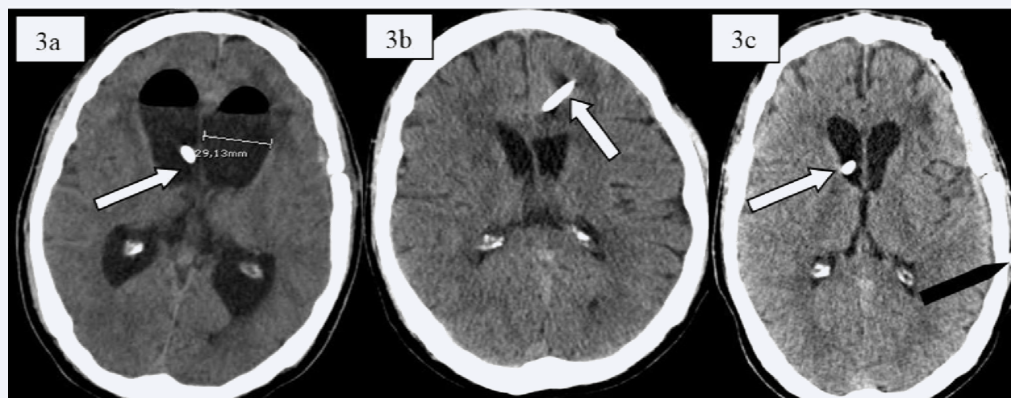


Figure 3 3a) On 27th day after admission, the pneumoventricle persisted and subdural collection was virtually inexistent. 3b) Disappearance of the ventriculomegaly and residual air, being the external ventricular drainage outside of the frontal ventricular horn (arrow). 3c) The anatomical recovery of ventricular size after ventricular (arrow) peritoneal Miethke shunt (black pencil) was placed a month later.

After discarding an obstruction of the EVD, the presence of a CSF fistula (given the surgical history) was suspected which could be responsible for the CSF low pressure? This fistula was not possible to detect in CT imaging studies, probably due to its small size, but it would be significant big enough to cause undetected liquorrhea and produce the clinical symptoms. Some authors [3] recommend a nuclear scanning with Tc-99m infusion to detect such communication.

MS is characterized by foci of inflammation with demyelination, axonal degeneration and reactive gliosis. In the study by Wuerfel et al [10] they demonstrated by magnetic resonance elastography the existence of a decrease in the viscoelasticity of the brain parenchyma in MS patients compared with healthy volunteers. In LPH, cerebral turgor is low due to loss of blood or extracellular fluid. This means that changes in intracranial volume does not increase ICP, because the brain can mold and act like a sponge [2,3]. Both concepts could act antagonistically in this patient, but we might think that in this case, his brain parenchyma had adapted to these changes by developing a progressive stiffness in the ventricles.

In a hydrocephalus situation, clinical and neurological status must be assessed. Imaging tests are used to confirm the diagnosis. The ICP monitoring confirms the diagnosis.

The actions that should be taken in every LPH are [3,6,11]: a) seeking a probable CSF fistula in order to seal it as far as possible; b) finding a possible communication between the ventricle and the subarachnoid space to perform lysis of fibrosis in the subarachnoid space or endoscopic third ventriculostomy in selected cases; c) increasing the pressure of the subarachnoid space, using compression around the neck as a conservative treatment; d) restore cerebral turgor with a low pressure ventricular drain [11].

Furthermore, other intensive care managements provided in this case included the use of a laser pointer in order to register the exact level between the EVD and the EC, and EVD closure during postural changes and while bed-armchair transfers occurred. Finally it was decided to put in a programmable VPS Miethke proSA® Aesculap [12] (pressure of 0-40 cm H₂O) at

zero pressure. This device consists of a programmable pressure hydrocephalus valve whose opening pressure for CSF drainage can be adjusted (in this case a pressure of 0-40 cm H₂O) according to the needs of the patient in supine position and it is coupled to a gravitational unit, which allows adjustment of the opening pressure that increases automatically when the patient is incorporated. Thus, the settings are regulated in different pressures for lying and seated positions, reducing the symptoms when ICP is inadequately low.

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