Normal Pressure Hydrocephalus

Mrs. Z, a 45-year-old female, was admitted to the hospital for a scheduled third ventriculostomy for the congenital hydrocephalus. Four years ago, Mrs. Z had a fall and hit her head. The computer tomography (CT) scan at that time revealed hydrocephalus and cerebrospinal fluid (CSF) flow study indicated decrease flow to the 4th ventricle. She was diagnosed with congenital hydrocephalus due to cerebral aqueduct (aqueduct of Sylvius) stenosis. She was referred to a neurosurgeon for follow up. As she had no signs or symptoms of hydrocephalus or increased intracranial pressure (ICP), conservative treatment and close monitoring of the progress or changes of hydrocephalus was recommended.

Mrs. Z was a non-smoker and she drinks one bottle of beer every day. She has chronic low back pain and is on acetaminophen with codeine to control her pain. She has had a few surgeries in the past, which include removal of cervical lymph node, excision of benign breast tumor, laparotomy for endometriosis, tubal ligation, caesarean section, and repair of a left shoulder injury.

Since last year, Mrs. Z started to have problems with short term memory. She stated she is forgetful and unable to focus or concentrate. She also has problems with walking and had two falls recently. Another issue, she developed urgency and incontinence of urine. She denied any dysuria, hematuria, or fever (these ruled out a urinary tract infection as the cause of her urgency and incontinence of urine). Mrs. Z also stated that she has bilateral blurriness of vision even with corrective glasses. Magnetic resonance imaging (MRI) (see below diagram) and CSF flow study indicated decreased flow from the foramen Luschka and foramen Magendie. Mrs. Z’s hydrocephalus was classified as obstructive hydrocephalus..

Because Mrs. Z had not shown any signs of increased ICP and also presented with the typical triad presentation (unsteady gait, dementia, and urinary incontinence) of normal pressure hydrocephalus. Mrs. Z was diagnosed with normal pressure hydrocephalus due to aqueduct stenosis. An endoscopic third ventriculostomy was suggested by the neurosurgeon and consented by the patient.

On examination, Mrs. Z’s Glasgow Coma Scale was 15/15, she has strong motor power (5/5), and no changes in sensation and coordination. Her blood pressure was 144/89, pulse 78/min, respiratory rate 14/min, SpO2 97% on room air. As well, the laboratory results were all within normal ranges.

On June 3, an endoscopic third ventriculostomy was performed under general anesthesia. The operation was uneventful with minimal blood loss. Post-operative orders included morphine and acetaminophen with codeine for pain control, metoclopramide and ondansetran as required for nausea, and sequential compression devices for deep venous thrombosis prophylaxis. Mrs. Z’s post operative neurological signs and vital signs were stable. She complained of headache and incisional pain post-operative, which was well controlled with morphine. She experienced some nausea with vomiting with ambulation, and the nausea was relieved with anti-emetic agents.

Post-operation day 1, Mrs. Z stated that her blurry of vision was much improved. Her gait was steadier. She stated she had not noted any improvement for her short term memory or urinary incontinence and she hoped it would improve overtime. The post-operative MRI was performed and showed good CSF flow through the third ventriculostomy (see below MRI).

![MRI showed significant dilation of the lateral ventricles](image1)

![MRI showed good CSF flow through the 3rd ventriculostomy (arrow)](image2)
Post-operative day 2, Mrs. Z was discharged home. She stated that her mother and uncle would be taking care of her when she returned home. She was advised not to do any heavy lifting and no driving until clear with the physician. She was notified if she developed fever, pain, swelling, and/or drainage from the incision site, she should seek medical attention immediately. She was also advised to follow up with the neurosurgeon in 6 weeks.

Case Highlight

Mrs. Z has been diagnosed with congenital hydrocephalus related to cerebral aqueduct stenosis. This type of hydrocephalus is usually classified as obstructive hydrocephalus or non-communicating hydrocephalus.

Hydrocephalus can be diagnosed by using the Evans ratio. Evans ratio is dividing the maximum width of the anterior horn (black arrow) divided by the maximum width of the calvarium (white arrow) at the level of foramen of Monro on the diagnostic images (see Mrs. Z’s MRI on below diagram), and if the ratio is or greater than 0.3, hydrocephalus is confirmed (Factora, 2006). The Evans ratio for Mrs. Z was 0.48.

Mrs. Z’s MRI

Congenital hydrocephalus has an incidence of 0.1% of live births (Munch et al., 2012). Aqueduct stenosis is the most common cause of congenital hydrocephalus (Lee et al., 2012).

The typical presentations of normal pressure hydrocephalus include three presenting symptoms, which are unsteady gait, dementia, and incontinence of urine. It is also referred to as Adam’s triad (Factora, 2006), some also called it “Hakim triad” (Kiefer & Unterberg, 2012). However, some patients with normal pressure hydrocephalus may only present with one or two symptoms (Paranathala et al., 2013). A detailed assessment must be performed to confirm the diagnosis, as these presentations can be easily mixed up with other diagnosis such as Alzheimer’s disease (dementia), Parkinson’s disease (unsteady gaits), and degenerative changes related to aging (dementia and incontinence of urine).

Most patients with non-communicating hydrocephalus are diagnosed in their younger age. There are some possible reasons for the delayed presentation of hydrocephalus. It may be due to the late-onset idiopathic aqueduct stenosis or compensated obstructive hydrocephalus (Wilson & Williams, 2007). Because of the compensation mechanism, symptoms of normal pressure hydrocephalus usually develop slowly (Paranathala et al., 2013). Mrs. Z’s hydrocephalus was diagnosed four years ago and she remained asymptomatic until recently. She was able to accommodate gradual increases in CSF volume without any symptoms until the “excessive” CSF caused stretching of the periventricular nerve fibers. At this point she started to present with signs and symptoms of hydrocephalus (Factora, 2006). The symptoms of normal pressure hydrocephalus are usually reversible when the excess CSF is “drained” by an external ventricular drain, a shunt or third ventriculostomy (Gelling et al., 2004; Kahn, 2013; Paranathala et al., 2013). Unsteady gait is usually the first presentation of normal pressure hydrocephalus and it is also the earliest symptoms improved after surgery (Kiefer & Unterberg, 2012). As shown in this case study, Mrs. Z stated that her gait is steadier and the blurry vision was resolved immediately after surgery.

Reference


**Normal Pressure Hydrocephalus Quiz**

1) Where is the most common blockage location in obstructive hydrocephalus?
   a) Foramen Monro  
   b) Lateral ventricle to third ventricle  
   c) Third ventricle to 4th ventricle  
   d) Forth ventricle to subarachnoid space

2) The triad presentation for normal pressure hydrocephalus include:
   a) Burry of vision, unsteady gait, dementia  
   b) Incontinent of urine, confusion, unsteady gait  
   c) Urgency of urination, unable to concentrate, ataxia  
   d) Unsteady gait, dementia, incontinent of urine

3) Cerebral aqueduct is the connection between:
   a) The lateral ventricles  
   b) Lateral ventricles to 3rd ventricle  
   c) 3rd ventricle to 4th ventricle  
   d) 4th ventricle to subarachnoid space

4) What causes the unsteady gait in normal pressure hydrocephalus?
   a) Mass effects of the enlarged ventricles  
   b) Stretches to the priventricular nerve fibers  
   c) Increased intracranial pressure  
   d) Damages to the motor neurons

5) What are the possible treatments for patients with normal pressure hydrocephalus?
   a) Ventriculo-atrial shunt, endoscopic 3rd ventriculostomy, ventriculoperitoneal shunt  
   b) External ventricular drain, steroid therapy, endoscopic 3rd ventriculostomy  
   c) Craniotomy, ventriculoperitoneal shunt, endoscopic 3rd ventriculostomy  
   d) Lumbar puncture, ventriculoperitoneal shunt, endoscopic 3rd ventriculostomy

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