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MANAGEMENT OF MULTIPLE, LATE ONSET COMPLICATIONS IN A 33-YEAR-OLD FEMALE, WITH A VENTRICULOPERITONEAL SHUNT AND CROHN'S DISEASE

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ABSTRACT

Numerous complications can occur after the placement of a venticuloperitoneal shunt. The late onset of an autoimmune disease such as Crohn's disease can be the disruptive factor for a previously well functioning shunt. A 33-year-old female with a ventriculo-peritoneal shunt since the age of 7, as well as Crohn's disease since the age of 25, presented in the ER with dysuria, long-lasting fever and intermittent severe headache. The patient underwent a CT scan of the chest, abdomen and brain. An enlargement of the ventricular system was revealed, suggesting malfunction of the shunt. Simultaneously, the abdominal scan revealed an abnormal course of the peritoneal catheter of the shunt in the lower abdomen, inside the bladder. The existence of the catheter inside the bladder was confirmed and filmed during a cystoscopy and was attributed to the several inflammations and surgeries performed for the treatment of Crohn's disease.The absence of an obvious stenosis of the aqueduct, the early immunodeficiency due to Azathioprine and the multiple abdominal surgeries made the treatment selection a complex algorithm for the neurosurgeon.

Keywords: hydrocephalus, complications, ventriculoperitoneal shunt, Crohn's disease

INTRODUCTION

Usually, the selection of a ventriculoperitoneal shunt depends on several criteria such as the type of hydrocephalus and the presence of other diseases that would probably affect the shunt performance. The late onset of numerous diseases could be able to obstruct the function of the shunt obligating the neurosurgeon to find alternative ways to override the CSF circulation. The appearance of parallel pathologies such as infections or autoimmune diseases is critical. A third venticulostomy should be considered only in cases with aqueductal stenosis as a primary cause. Otherwise, the revision of the shunt is obligatory. In every case, the treatment of late onset complications of ventricu loperitoneal shunts should be customized.

CASE REPORT

A 33-year-old female presented in the ER with severe intermittent headache, long lasting fever and dysuria. The patient's medical history included the placement of a venticuloperitoneal shunt at the age of 2, with unknown \prine since the age of 25. The shunt had been reviewed at the age of 7. The patient had given childbirth with cesarean section twice and had, also, undergone enterectomy twice, for complications of the Crohn's disease. A brain CT scan revealed acute hydrocephalus due to ventriculoperitoneal shunt malfunction. The computerized tomography of the abdomen showed multiple abscesses along the course of the peritoneal part of the shunt due to a relapse of the Crohn's disease, as well as part of the peritoneal catheter inside the bladder. Her 8-year history of Crohn's disease under Azathioprine, two enterectomies for enterocutaneous fistulas and cesarean section twice in her life explained the multiple abdominal complications and the shunt dysfunction. The shunt was removed and an external ventricular drainage was placed instead. The peritoneal part of the shunt was removed by a second cystoscopy. The blood cultures indicated an E-coli strain and the proper antibiotics were administered.

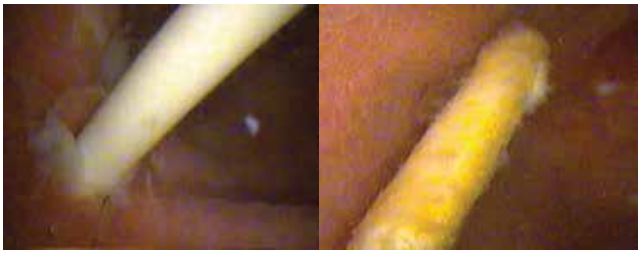

Image 1: The cystoscopy revealed the peritoneal part of the VP shunt inside the bladder. The entrance as well as the tip of the catheter can be seen above.

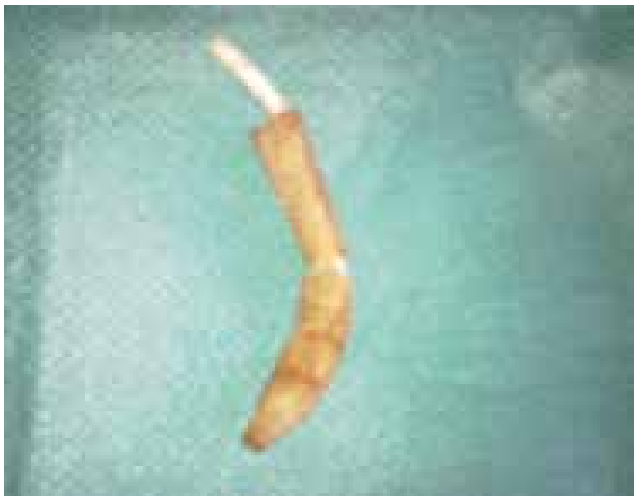

Image 2: The tip of the catheter after the removal during the second cystoscopy. A calcium oxalate stone had been formed around the catheter preventing the neurosurgeon to pull it otherwise.

Several days later, the shunt seemed to malfunction again due to abdominal adhesions and symptoms of high ICP occurred to the patient who was readmitted to the operating room and a right ventriculo-jugular shunt was performed. Although the symptoms of hydrocephalus were well controlled, one month later low grade fever occurred and a cardiac ultrasound revealed the existence of a 3-4 cm vegetation inside the right jugular vein. In front of the imminent septic embolism the jugular catheter was slowly retracted under ultrasonic guidance. The tip of the catheter was used for multiple cultivations and an external ventricular drainage was placed. After 10 days of combined antimicrobial therapy and the helpful assistance of expert abdominal surgeons, a new ventriculoperitoneal shunt was performed by placing the peritoneal part in the left paracolic sulcus. The patient had a full recovery after 3 months of antimicrobial therapy. The concern for future Crohn's disease attacks and new possible complications still exists. A psychological evaluation and support of the patient is also needed.

DISCUSSION

Complications of ventriculoperitoneal shunts Most problems associated with shunting occur weeks or even years after the surgery. Although, shunt complications can be very serious and become life threatening, they can almost always be treated successfully when they are discovered early.Many of the complications associated with shunt placement, occur during childhood and cease once the patient has reached adulthood. Most of these require immediate shunt revision (the replacement or reprogramming of the already existing shunt). The common symptoms often resemble the new onset of hydrocephalus such as headaches, nausea, vomiting, double-vision, and an alteration of consciousness. The shunt failure rate 2 years after implantation has been estimated to be as high as 50% [1].

Shunts are very durable, but the components of the shunt can become disengaged or fractured as a result of wear or as a child grows, and occasionally they move from where they originally were placed. More rarely, a valve will fail because of mechanical malfunction. Shunt malfunction is usually a problem of partial or complete blockage of the shunt at either the proximal or distal end. Shunt obstruction can occur in any part of the shunt. At the proximal end, the shunt valve can become blocked due to the buildup of excess protein in the CSF. The extra protein will accumulate at the point of drainage and slowly clog the valve. Most commonly in children, the ventricular catheter becomes obstructed by tissue from the choroid plexus or ventricles. In adults it is more often that the distal catheter becomes blocked. This can happen, if the shunt is pulled out of the abdominal cavity (in the case of VP shunts), or from similar protein build-up. The catheters or the valve may, also, become blocked with blood cells or bacteria. In this case, the fluid backs up from the site of the obstruction and, if the blockage is not corrected, almost always results in recurrent symptoms of hydrocephalus.Symptoms of shunt malfunction vary considerably from person to person, but tend to be similar each time for a particular person. In infants, signs include a full and tense fontanel, bulging of the scalp veins and swelling or redness along the shunt tract. Unusual vomiting, irritability, sleepiness and decreased interest in eating may also occur. It is noteworthy that medication with a side effect of drowsiness can mimic or mask signs of shunt malfunction and should be used with caution in those with hydrocephalus, especially infants and young children. Older children and adults may experience headaches, vomiting, irritability and tiredness. Swelling along the shunt tract occurs less frequently. In the event of an abrupt malfunction, a child may develop symptoms very rapidly, in a matter of hours or days. Without treatment, coma, and even death, may occur. People who were diagnosed and treated in adulthood, including those with NPH, tend to revert to the symptoms they experienced before initial treatment during a malfunction.Shunt infection, which may happen with or without a shunt obstruction, is a common problem and can occur in up to 27% of patients with a shunt[2]. It is, usually, caused by a person's own bacterial organisms, rather than acquired from exposure to other children or adults. Infection can lead to long-term cognitive defects, neurological problems, and in some cases death. Common microbial

agents for shunt infection include Staphylococcus epidermidis, Staphylococcus aureus and Candida albicans, which can be found on the surface of the person's skin and in the sweat glands and hair follicles deep within the skin. Infections of this type are most likely to occur one to three months after surgery but may occur up to six months after the placement of a shunt. Further factors leading to shunt infection include shunt insertion at a young age (<6 months old) and the type of hydrocephalus being treated. There is no strong correlation between infection and shunt type[2]. People with ventriculo-peritoneal (VP) shunts are at risk of developing a shunt infection secondary to abdominal infection, whereas people with ventriculo-atrial (VA) shunts may develop generalized infection, which may rapidly deteriorate. In either case, the shunt infection must be treated immediately to avoid life-threatening conditions or possible cerebral damage. The symptoms of a shunt infection are very similar to the symptoms seen in hydrocephalus but can also include fever and elevated white blood cell counts[3]. Sometimes shunt infection also produces reddening or swelling along the shunt tract.Treatment of a CSF shunt infection generally includes removal of the shunt and placement of a temporary ventricular reservoir until the infection is resolved[4][5]. There are four main methods of treating ventriculoperitoneal (VP) shunt infections: (1) antibiotics; (2) removal of infected shunt with immediate replacement; (3) externalization of shunt with eventual replacement; (4) removal of infected shunt with external ventricular drain (EVD) placement and eventual shunt re-insertion. The last method is the best with over 95% success rate[6].Other shunt complications may include the shunt system draining CSF at the wrong rate. Underdrainage of the ventricles can fail to relieve the symptoms of hydrocephalus. On the other hand, overdrainage of the ventricles, which occurs when a shunt has not been adequately designed for the particular patient, can cause the ventricles to decrease in size to the point where the brain and its meninges pull away from the skull or the ventricles become like slits. If blood from ruptured vessels in the meninges becomes trapped between the brain and skull, resulting in a subdural hematoma, further surgery is required. This is most common in older adults with normal pressure hydrocephalus (NPH).Slit-like ventricles, also called slit-ventricle syndrome (SVS), is an uncommon disorder associated with shunted patients, which occurs when CSF slowly overdrains, over several years and it results in a large number of shunt revisions. It is most commonly a problem in young adults who have been shunted since childhood. The condition is often thought to occur during a period where overdrainage and brain growth occur simultaneously. In this case the brain fills the intraventricular space, leaving the ventricles collapsed. Furthermore, the compliance of the brain will decrease, which prevents the ventricles from enlarging, thus reducing the chance for curing the syndrome. The collapsed ventricles can also block the shunt valve, leading to obstruction. Since the effects of slit ventricle syndrome are irreversible, constant care in managing the condition is needed[7][8]. The most common symptoms are similar to normal shunt malfunction, but there are several key differences. First, the symptoms are often cyclical and will appear and then subside several times over a lifetime. Second, the symptoms can be alleviated by lying prone. In the case of shunt malfunction neither time nor postural position will affect the symptoms.Recent studies have shown that overdrainage of CSF due to shunting can lead to acquired Chiari I malformation[9]. It was previously thought that Chiari I Malformation was a result of a congenital defect but new studies have shown that overdrainage of cysto-peritoneal shunts used to treat arachnoid cysts can lead to the development of posterior fossa overcrowding and tonsillar herniation, the latter of which is the classic definition of Chiari I malformation. Common symptoms include major headaches, hearing loss, fatigue, muscle weakness and loss of cerebellum function. To restore a balanced flow of CSF it may be necessary to place a new shunt containing a more appropriate pressure valve. For those who have externally adjustable or programmable valves, the balance of flow can often be restored by re-setting the opening pressure. An intraventricular hemorrhage is another complication that can occur at any time during or after a shunt insertion or revision. The hemorrhage can cause an impairment in shunt function which can lead to severe neurological deficiencies[10]. Studies have shown that intraventricular hemorrhage can occur in nearly 31% of shunt revisions[11].The peritoneum is the most popular site for the distal catheter implantation. Although, ventriculoperitoneal (VP) shunts do not have fewer complications than ventriculoatrial shunts, the complications are less severe and associated with a lower mortality rate. However, the peritoneum is not immune to specific complications. Several intra-abdominal complications following ventriculoperitoneal procedures, have been reported. These include: intra-abdominal cerebrospinal fluid (CSF) cysts (peritoneal pseudocysts), catheter disconnection and lost distal catheters, knotted catheter, intestinal obstruction due to volvulus around the shunt, bowel or bladder perforations and previously unrecognized inguinal hernias that require special attention[12]. Abdominal x-ray is often diagnostic; it may demonstrate the VP-shunt catheter in the scrotum in patients with otherwise unrecognized inguinal hernia, and it will also reveal cases of knotted shunt, shunt disconnection, bladder perforation, CSF cysts, and intestinal obstruction. In addition, contrast studies

in instances of shunt dysfunction documented bowel perforation and CSF cyst formation[12]. CSF ascites is a very rare complication of ventriculoperitoneal (VP) shunt procedure. No definite explanation has been offered for the inability of the peritoneum to absorb the CSF. The treatment of choice is conversion of the VP shunt to a ventriculoatrial shunt[13]. There have also been reports of asymptomatic perforation of the large bowel and the urinary bladder with prominence of the VP shunt tip from the anus and from the urethral orifice, respectively[14][15][16]. Furthermore, vaginal perforation has also been reported[17]. These data indicate that intra-abdominal complications, following VP-shunt operations, are not uncommon and close observation is advised.

CONCLUSION

Autoimmune, cardiac, inflammatory and other diseases can affect the shunt function.The treatment of ventriculoperitoneal shunt's complications should always be under the evaluation of a multidisciplinary medical team. Numerous therapeutic pathways should be examined and the final decision should always be customized upon the special needs and complex pathologies of the patient.

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Dr. Gkolemis: Study concept and design, protocol writing, data collection, data analysis, manuscript writing, manuscript review

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Dr. Kokkalis: Protocol writing, data collection, data analysis, manuscript writing, manuscript review

Dr. Stamatopoulos: Study concept and design, protocol writing, data collection, data analysis, manuscript writing, manuscript review

Dr. Syrmos: concept and design, protocol writing, data collection, data analysis, manuscript writing, manuscript review

Dr. Paleologos: Data collection, data analysis, manuscript writing, manuscript review