Lumbo-peritoneal Shunting Improved Spinal Cord Compression Due to a Large Anterior Sacral Meningocele

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Objective: *Lumbo-peritoneal shunt was used as an alternative treatment in a large anterior sacral meningocele, a rare congenital malformation, is illustrated.*
Clinical Presentation: *A 48-year-old female patient presented with a two-month history of progressive paraparesis, leg pain and numbness of both legs. The investigation demonstrated spina bifida with a huge sac of meningocele, protruding anteriorly into the pelvic cavity. On two previous surgical operations for closure, the fistula through the posterior transsacral approach failed 20 years ago at another hospital, but her symptoms had subsided spontaneously without explanation. She came to Thammmasat Hospital with an episode of spinal cord compression for 2 months.*
Management: *Because of the old surgical scar and high pressure of the meningocele, a lumbo-peritoneal shunt was selected to drain the cerebrospinal fluid from the meningocele to the peritoneal cavity.*
Conclusion: *Lumbo-peritoneal shunting is an optional treatment for a large sacral meningocele, especially in a large fistula with unsuccessful surgical closure.*

Keywords: *Lumbo-peritoneal shunt, Anterior sacral meningocele*

J Med Assoc Thai 2005; 88(2): 265-8
Full text. e-Journal: *http://www.medassocthai.org/journal*

Anterior sacral meningocele (ASM) is a rare congenital anomaly, which is caused by anterior herniation of the thecal sac through a sacral defect. Most cases usually occur before the third decade of life. The incidence of a posterior meningocele is about 1/1000 live births, but the actual incidence of ASM is still unknown due to the asymptomatic characteristics. Since it was described by Bryant in 1837[1,2], only approximately 200 cases of ASM have been reported in the world literature[3-4].

Although ASM frequently occurs sporadically, the inheritance of ASM has been proposed to be autosomal dominant or x-linked dominant[4-10]. Several reports have shown some association with Marfan syndrome[11-15].

Surgical treatment for ASM is generally recommended because spontaneous regression does not occur. Several surgical techniques have been demonstrated in several reports, such as a posterior transsacral approach with sac ligation[16,17], anterior transabdominal approach with oversewing of the neck of the meningocele[18,19] and endoscopic technique[3,15,20,21]. However, no report has demonstrated any alternative management in the case of failure from surgical closure for ASM. The present report is the first case presentation to use the lumbo-peritoneal shunt to reduce the symptoms of spinal cord compression following the failures of surgical operations.

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Case Report

History

A 48-year-old female patient was admitted to the Neurosurgical Unit of Thammasat University Hospital, Patumthani, Thailand. She presented with a two month history of progressive paraparesis, pain and numbness of both legs, from hip to toe, as well as progressive headaches during the two months before admission. She had more symptoms of numbness in the morning, which improved when she walked. She also had several episodes of urinary incontinence.

20 years ago, she was admitted to a department of gynecology in a hospital in Bangkok, Thailand,

with symptoms of dysfunctional uterine bleeding. The diagnosis of an ovarian cyst was made by pelvic ultrasonography. She was, therefore, operated upon by a gynecologist, but both ovaries were revealed to be normal and no abnormal cyst in the pelvic cavity was demonstrated. Two weeks later, she underwent a physical examination with manual compression of the lower abdomen. After a while, she had sudden paraparesis and numbness of both thighs and legs and severe headaches. She was immediately further investigated by lumbar myelography and magnetic resonance imaging (MRI) scan. The result demonstrated spina bifida with a huge sac of meningocele, protruding anteriorly into the pelvic cavity. On two occasions surgical operations through the posterior transsacral approach failed, because the neck and orifice of the meningocele were too large to perform a simple ligation. After the last operation, the patient was lost to follow-up for a 20-year period. However, the patient's symptoms spontaneously subsided to nearly normal with only minimal numbness of both legs during the lost to follow-up period until two months before this admission.

Evaluation

On admission, the neurological examination showed good consciousness, no stiff neck and papilledema. Motor function of upper extremities were normal. The positive finding was paraparesis of both thighs and legs with grade IV/V motor power. Pain and temperature sensation revealed impairment of sensory function from the level of the hip to the toe. Propioceptive and touch sensation were impaired only in a small area of the dorsum of both feet.

Plain x-rays demonstrated a spina bifida of the lower lumbar spine and a scimitar-shaped sacral bone defect (Fig. 1). She was investigated by MRI scan, which revealed a large lumbosacral meningocele with no significant compression of the spinal cord (Fig. 2). Cerebral computed tomography (CT) scan was performed and showed no hydrocephalus.

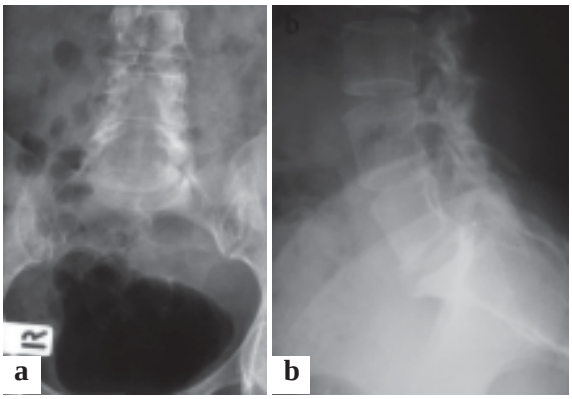

Fig. 1a,b Plain x-ray of lumbosacral spine in AP and lateral views, demonstrating the spina bifida of lower lumbar spine and the dysgenesis of sacrum

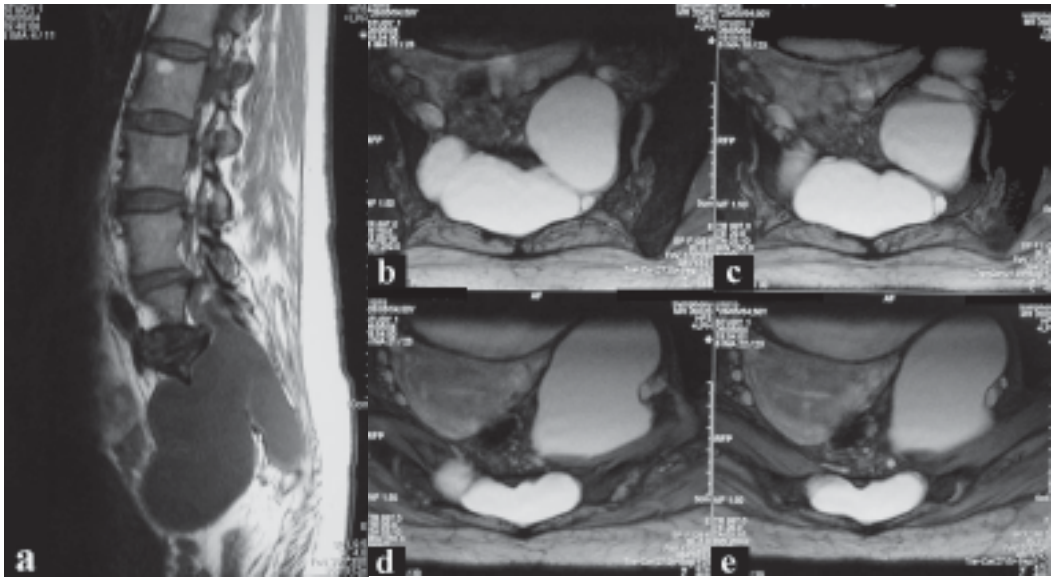

Fig. 2a-e Lumbosacral MRI: **a** sagittal T1-weighted lumbosacral MRI reveals a large sacral meningocele with a large fistula; **b-e** T2-weighted lumbosacral MRI (axial views) shows a large sacral meningocele with anterior protrusion into the pelvic cavity

During the admission, acetazolamide was given to the patient at a dosage of 250 milligrams twice a day.

Surgical Management

Lumbar puncture was performed by using a 22-gauge Quinke needle with a stylet. It was inserted through the skin and advanced through the cyst of the meningocele. The cerebrospinal fluid (CSF) pressure was measured. The opening pressure was 28 cmH_2O. Because of the high CSF pressure of the meningocele without hydrocephalus, lumbo-peritoneal shunt might benefit the patient to release the CSF pressure. Moreover, because of the fibrotic scar of the previous two operations, re-exploration might be very difficult to close the fistula. Therefore, lumbo-peritoneal shunting might be the best option to perform on the patient.

The lumbo-peritoneal shunt was inserted by making a small mid-line lumbar incision. The spinal catheter was introduced through a trocar and cannula up to 10 cms within the cavity of meningocele and then tunnelled under the abdominal wall to be introduced into the peritoneal cavity by using the percutaneous method using trocar and cannula.

Clinical Course

The symptoms of headaches and leg pain disappeared immediately after shunting. The motor powers of both lower extremities improved post-operatively to grade V/V within one week. Pain and temperature sensation improved to normal in a two week period. No symptoms of urinary incontinence occurred after the lumbo-peritoneal shunt operation. All symptoms recovered to completely normal after three months.

Discussion

ASM is a rare congenital malformation usually associated with sacrococcygeal bony defect. ASM is a lesion consisting of a cerebro-spinal fluid sac in the pelvic cavity communicating by a sub-arachnoid fistula through the sacral defect. Conservative therapy of ASM is limited to children, who are at high surgical risk, and to male patients affected with a small meningocele which doesn't tend to enlarge and with no associated tumors[4]. However, surgical closure is the treatment of choice, because ASM does not have spontaneous regression and generally progresses its enlargement with a corresponding increase in the risk of complications. The goal of ASM treatment is the obliteration of communication between the spinal subarachnoid space and the meningocele. In clinical practice, the surgical approach is via a posterior transsacral approach and transdural ligation of the neck of the meningocele[22]. In addition, a few reports showed successful obliteration by endoscopic techniques[3,15,20,21].

ASM commonly presents with symptoms caused by a local mass effect in the adjacent area, which can lead to urinary incontinence, constipation, dysmenorrhea and/or dyspareunia. Moreover, it can manifest itself with local neurological symptoms in the form of sacral numbness, lower extremity paresthesia or difficulty of anal sphincter control[23].

The initial symptoms of the illustrated case were numbness and paraparesis of both thighs and legs which were caused by spinal cord compression. The lumbar puncture confirmed the high CSF pressure in the cavity of the meningocele, which could compress the spinal cord and spinal nerve root. For the surgical management decision, re-exploration for meningocele closure might be unquestionably difficult due to the fibrotic scaring from the previous two operations. Additionally, theoretically, the fibrotic scaring might lead to loculation of CSF in the meningocele as a sac with a one-way valve opening, producing high pressure to compress the spinal cord. For these reasons, a lumbo-peritoneal shunt was used to release the pressure as an alternative. It is also safe in the protection of spinal headache due to CSF overdrainage because of the one-way valve mechanism.

So far, there have been no reports of lumbo-peritoneal shunts for ASM treatment in the previous literature. The objectives of this presentation were, therefore, to demonstrate an alternative management which might be useful to improve symptoms in some conditions, for instance, failure of surgical closure as well as small ASMs. Although the lumbo-peritoneal shunt is not the ideal management for ASM, it could be beneficial to improve the quality of life in some patients.

Conclusion

Lumbo-peritoneal shunt is an alternative treatment, which could improve the symptoms of the high-pressure sacral meningocele, especially in a large fistula with unsuccessful surgical closure.

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การใส่สายระบายน้ำไขสันหลังสู่ช่องท้อง (lumbo-peritoneal shunt) สามารถช่วยลดอาการจากไขสันหลังถูกกดเบียดโดยถุงน้ำของเยื่อหุ้มไขสันหลังขนาดใหญ่ในอุ้งเชิงกราน

ภัทรวิทย์ รักษ์กุล

รายงานนี้เป็นรายงานครั้งแรกของการทำผ่าตัดใส่สายระบายน้ำไขสันหลังสู่ช่องท้องในการรักษาอาการจากไขสันหลังถูกกดเบียดโดยถุงน้ำของเยื่อหุ้มไขสันหลังขนาดใหญ่ในอุ้งเชิงกรานในผู้ป่วยหญิงอายุ 48 ปี ซึ่งมีประวัติอาการปวดขา 2 ข้าง ชาและอ่อนแรง และมีอาการมากขึ้นในช่วง 2 เดือน โดยเมื่อ 20 ปีก่อนทำการตรวจพบมี spina bifida และถุงน้ำของเยื่อหุ้มไขสันหลังขนาดใหญ่ และยื่นไปทางด้านหน้าเข้าไปในอุ้งเชิงกราน โดยผู้ป่วยได้รับการผ่าตัด 2 ครั้งเมื่อ 20 ปีก่อน และล้มเหลวทั้ง 2 ครั้ง แต่อาการผู้ป่วยดีขึ้นได้เองจนเริ่มมีอาการอีกครั้งเมื่อ 2 เดือน ก่อนมาโรงพยาบาล แต่เนื่องจากพังผืดจากการผ่าตัดทั้ง 2 ครั้ง และตรวจพบความดันภายในถุงน้ำสูง การผ่าตัดใส่สายระบายน้ำ ไขสันหลังสู่ช่องท้องจึงถูกเลือกมารักษา เพื่อเป็นการระบายน้ำไขสันหลังออกเพื่อลดการกดเบียดไขสันหลัง และจากผลการรักษาสามารถสรุปได้ว่า การใส่สายระบายน้ำไขสันหลังสู่ช่องท้องสามารถช่วยลดอาการจากไขสันหลังถูกกดเบียดโดยถุงน้ำของเยื่อหุ้มไขสันหลังขนาดใหญ่ในอุ้งเชิงกราน โดยเฉพาะอย่างยิ่งในผู้ป่วยที่ไม่สามารถทำการผ่าตัดปิดช่องเปิดขนาดใหญ่ได้