An Unusual Presentation of Anterior Sacral Meningocele: A Case Report and a Review of the Literature

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Abstract

Background
Anterior Sacral Meningocele (ASM) is an extremely rare congenital anomaly which was first described in 1837 by Bryant.1,2 Anterior Sacral Meningocele is a unilocular or multilocular extension or herniation of the dura mater and arachnoid out of the sacral spinal canal through a defect in the sacrum into the retroperitoneum and infraperitoneal space, either anteriorly through the body of the sacrum or anterolaterally through an enlarged intervertebral foramen or coalesced foramina. It contains cerebrospinal fluid (CSF), which in some reported cases, has been discoloured (slightly yellow), and has had an elevated protein content. It may also contain neural tissue.2,3,4.

Most cases usually occur before the third decade of life. The incidence of a posterior meningocele has been reported to be about 1/1,000 live births, but, the actual incidence of Anterior Sacral Meningocele is unknown due to the asymptomatic characteristics. Since it was first described by Bryant, only approximately 245 cases of ASM have been reported in the world literature.5, 6, 7-20.

Even though Anterior Sacral Meningocele occurs sporadically, its inheritance has been proposed to be autosomal dominant or x-linked dominant.14. 21. 22. 23. 24. 25. 26.

Anterior Sacral Meningocele has been stated to be more common in females, ranging from a male to female ratio of 1:4 to 31:1.16.2, 3, 4, & 10.

In view of the fact that Anterior Sacral Meningocele is an uncommon condition, the diagnosis could be missed easily by the unaccustomed practitioner. A case of an Anterior Sacral Meningocele in an adult male which was inadvertently missed after ultrasonography is reported with a review of the literature.

Case Report

A 31 year old man was referred from another hospital with an 8 month history of gradual abdominal distension. There was no history of weight loss,
anorexia, nausea or vomiting. He also did not have urinary or bowel disturbances. He did not have any significant past medical history.

On inspection he was found to have massive distension of abdomen mainly involving the lower part of the abdomen. The umbilicus was pushed up. All quadrants of the abdomen moved equally with respiration. Dilated veins were visible in both groins. On palpation there was evidence of a huge mass in the supra-pubic region extending up to the umbilical region, non tender, non mobile, dull on percussion. On auscultation the bowel sounds were present. His rectal examination did not reveal any abnormality. The systemic examination was normal.

The provisional radiological diagnosis based upon the CT scan was: bilateral hydronephrosis and polycystic right kidney with a huge right renal cyst. The bilateral hydronephrosis was considered to be due to the large intra abdominal and intra pelvic cyst.

The radiology consisted initially of CT scan of abdomen and pelvis with intravenous contrast injection and scanning in the portal venous phase. The initial scannogram was typical of a sacral defect affecting the distal sacrum but greater involvement of the right side. The subsequent scan showed bilateral severe hydronephrosis particularly severe on the right with severe cortical thinning of the right kidney. Arising out of the pelvis and extending into the abdomen was a huge homogeneous cyst without any enhancement or septation. The Hounsfield unit measurement was between -5 and 20. The ureters were displaced anteriorly indicating that the cyst must be retroperitoneal and posteriorly placed. The urinary bladder was displaced anteriorly and to the left of midline. There was a large anterior defect of the sacrum as well as absence of the distal sacrum. The cyst was in communication with the sacral defect and therefore diagnosed as representing anterior sacral meningocele. (See figures 1, 2 & 3 which respectively show: the meningocele; bilateral hydronephrosis and the meningocele; the displaced bladder, the Anterior Sacral Meningocele and the sacral defect). The sigmoid colon was stretched over the under surface of the meningocele. No other abnormalities were detected in the rest of the abdomen.

Following this an MRI scan was also performed. The MRI scan of the spine showed no abnormalities of the cervical, thoracic or lumbar vertebrae or the cord and a huge anterior meningocele. (See figures 4, 5 & 6). There was also cerebellar tonsilar herniation representing the type 1 Chiari malformation due to a large sacral defect.

In view of the CT scan and MRI scan confirmation of the presence of Anterior Sacral Meningocele, the neurosurgeons accepted the patient with a provisional plan to repair the meningocele by the anterior approach.

**Discussion**

Anterior Sacral Meningocele is an uncommon congenital malformation usually associated with sacrococygeal bony defect. Anterior Sacral Meningocele is a lesion, which consist of a cerebro-spinal fluid sac in the pelvic cavity communicating by subarachnoid fistula through the sacral defect. It is recognised within the heritable Currarino triad, consisting of congenital anal stenosis, scimitar sacrum and a presacral mass (the ASM7, 20, 35.)

However, Anterior Sacral Meningocele may exist without the presence of anal stenosis. Anterior Sacral Meningocele has been associated with a variety of congenital abnormalities of the pelvis. Some of these abnormalities include: anorectal malformations, dermoid cysts, sacrococcygeal teratoma and lipomata, duplication of urogenital tracts as well as epidermoid tumours 2, 4, 36, 37. A number of reports have shown some association with (a) Marfan syndrome 10, 27-30 and with (b) neurofibromatosis2, 28.

Even though Anterior Sacral Meningocele can present at any age, it usually occurs in the second or third decades and child bearing years in females; but in males it is usually diagnosed in the first decade 2, 3, 4, 8, 38.

Patients' symptoms are usually attributable to a pelvic mass (for example, chronic constipation, urinary incontinence, and dysmenorrhoea / dyspareunia) or chronic low back ache. 1, 4, 38, 39-43.

In females, Anterior Sacral Meningoceles have been reported to have caused problems in labour and they have caused obstructed labour which resulted in deliveries by caesarean section 44, 45.

Other presentations of Anterior Sacral Meningocele include:

a) Abdominal, gluteal, pelvic or groin masses. 1, 2, 3, 4

b) Bacterial meningitis. 9, 46, 47, 48, 49.

c) Head aches. - Increased pressure headaches due to compression of the anterior sacral meningocele by increased abdominal pressure such as occurs during defaecation with emptying of the contents of the ASM back into the cerebro-spinal fluid spaces. On the other hand, low pressure head aches have also been reported when the patient stands and the ASM refills4.

d) Local neurological symptoms in the form of sacral
numbness, lower extremity paresthesia or difficulty of anal sphincter control.11
There is disagreement with regards to the presence or absence of spina bifida with Anterior Sacral Meningocele. Wilkins in 1996 stated that the association with spina bifida is rare but can be present, with Posterior Sacral Meningocele associated with Anterior Sacral Meningocele being rarer still.2 Some authors are in agreement with Wilkins’ view 3 46. On the other hand others have stated that all cases of Anterior Sacral Meningocele are associated with spina bifida, at least in a paediatric population10.
With regards to radiological diagnosis, there is the general acceptance that a scimitar shaped sacrum (a concave defect in one side of the sacrum) is often diagnostic on plain radiography (plain X-ray of the pelvis).4, 35. There is variation in the bone defects that have been associated with Anterior Sacral Meningocele ranging from an enlarged foramén to complete sacral agenesis2, 39. Myelography, CT scan and MRI scan are useful in delineating the anatomy of the stalk and in planning of a surgical operation for the meningocele. CT scan and MRI are the investigation of choice46. The cord and the cauda equina have been reported to be usually normal 3, 40. It has nevertheless, been reported that there may be tethering and abnormalities of the nerve supply to the bladder 4, 39, 43. In our case CT scan and MRI scans clearly confirmed the diagnosis of Anterior Sacral Meningocele as well as bilateral hydronephrosis (What was considered to be a huge renal cyst at the referring hospital was found to be rather a huge anterior sacral meningocele).12
Conservative management of Anterior Sacral Meningocele 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 tumours14. Nevertheless, surgical closure is the treatment of choice, because Anterior Sacral Meningocele does not have spontaneous regression and generally progresses its enlargement with a corresponding increase in the risk of complications. In the surgical treatment care is required to avoid precipitating bacterial meningitis 2, 8, 31, 46. The goal of the treatment of Anterior Sacral Meningocele is the obliteration of communication between the spinal subarachnoid space and the meningocele. A posterior approach to perform a sacral laminectomy and tie off the neck of the Anterior Sacral Meningocele is favoured by some surgeons2, 12, 31, 48. Nevertheless, trans-abdominal (‘anterior abdominal approach’ with over-sewing of the neck of the meningocele) and perineal approaches have been reported2, 16, 32, 33. Other surgical approaches that have been reported in the treatment of Anterior Sacral Meningocele include endoscopic and laparoscopic techniques6, 13, 30 & 34. It has been stated that meningocele excision is not deemed necessary2. Hino et al (1993) reported that failure to close the neck of the Anterior Sacral Meningocele resulted in a recurrence31. It has also been reported that lumpo-peritoneal shunt insertion is an alternative treatment, which could improve the symptoms of the high-pressure sacral meningocele, especially in a large fistula with unsuccessful surgical closure5.

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Illustrations

Illustration 1

Figure 1: CT scan Block Arrow showing Meningocele

Illustration 2

Figure 2: CT scan Black Arrow - Bilateral hydronephrosis (Right > Left); Block Arrow - Meningocele
Illustration 3

Figure 3: CT scan - Black Arrow Bladder; Block Arrow Meningocele; Stripped Arrow - Defect in spine

![CT Scan Illustration](image)

Illustration 4

Figure 4: MRI scan - Arrow indicates the sacral defect leading to large meningocele

![MRI Scan Illustration](image)
Illustration 5

Figure 5: T2 weighted sagittal MRI scans of lumbar spine - Arrow indicates the sacral defect anterior to which is a large meningocele. Incidental degenerate L5/S1 disc.

Illustration 6

Figure 6: T1 weighted axial MRI scan through the sacral level - Arrow points to the large sacral defect.
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