CASE REPORT

MULTIMODALITY IMAGING OF ANTERIOR SACRAL MENINGOCELE WITH TETHERED CORD AND NEUROGENIC BLADDER: A CASE REPORT

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ABSTRACT

A dural sac herniation through an anterior sacral defect is the hallmark of anterior sacral meningocele (ASM), a rare congenital form of spinal dysraphism that frequently manifests clinically as an abdominal or pelvic mass. Constipation, urinary disturbances, obstetric difficulties, headaches, or changes in lower limb sensation are the most common symptoms that arise from a mass effect on nearby pelvic structures. Less than 300 cases have been documented; they are clearly more common in women, typically manifest in late childhood or early adulthood, and are often linked to connective-tissue diseases, especially Marfan syndrome. In rare cases, a tethered cord may be present. MRI is the preferred modality to diagnose anterior sacral meningocele . Surgery is the recommended treatment for symptomatic or growing anterior sacral meningoceles We present rare case report of a young female with anterior sacral meningocele, tethered cord and neurogenic bladder to highlight its imaging features, clinical implications, and diagnostic challenges.
Keywords: Anterior sacral meningocele, tethered cord, neurogenic bladder, rare , spinal dysraphism.

Introduction

Anterior sacral meningocele (ASM) is a rare congenital form of spinal dysraphism characterized by herniation of the dural sac through a defect in the anterior aspect of the sacrum.[1] In most cases, the spinal cord and the caudaequina are normal in ASM, but in rare cases, a tethered cord may be present.[2] ASM is frequently misdiagnosed as a gynecologic, urologic, or gastrointestinal pathology because of its deep pelvic location and nonspecific clinical presentation.[3] There is a definite female predominance, and the majority of patients arrive in late childhood or early adulthood.[3,4] Constipation, urinary dysfunction, obstetric complications, headaches, and lower-limb sensory symptoms are among the clinical manifestations that are mainly caused by the mass effect on nearby pelvic structures.[3] ASM is often linked to connective tissue disorders, especially Marfan syndrome, and dural ectasia.[1] Imaging-especially magnetic resonance imaging-plays a crucial role in diagnosis by demonstrating communication with the thecal sac and associated sacral defects.[1] Early detection and effective treatment are crucial due to the possibility of severe consequences like meningitis, sepsis, and progressive pelvic organ dysfunction.[5]

Surgery is the recommended treatment for symptomatic or growing anterior sacral meningoceles because these lesions do not resolve spontaneously and the risk of complications increases over time.[3]

History and Treatment

A 22 year old girl presented in the Emergency department of Liaquat National Hospital with the presenting complaint of hematuria, vaginal bleeding, epistaxis, and

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an episode of hematemesis few hours back. Her past medical history included fever two days ago, stress and urge incontinence one year back that got treated under the line of UTI. She also had a history of amenorrhea for one year. Upon ultrasound moderately echogenic kidneys and moderate to severe hydronephrosis with proximal hydroureter was seen bilaterally however distal ureter was not visualized due to gases. The urinary bladder was inadequately filled and showed heterogenous area with no vascularity on CDI.

Her CT scan of the whole abdomen with contrast showed a pear shaped Urinary bladder with significantly thick walls and trabeculated margins and multiple diverticulae (Fig.1). Mild to moderate bilateral hydronephrosis and hydroureter was seen. Cortical irregularity was seen bilaterally and cortical thinning on left side likely representing chronic pyelonephritis secondary to vesicoureteric reflux (Fig.2). The sacrum was deformed and tilted with anterior sacral defect on left side and herniation of dura matter from this defect into retrorectal region forming a bilobed cystic structure without any solid component or thick enhancing walls (Fig.3). Atro-

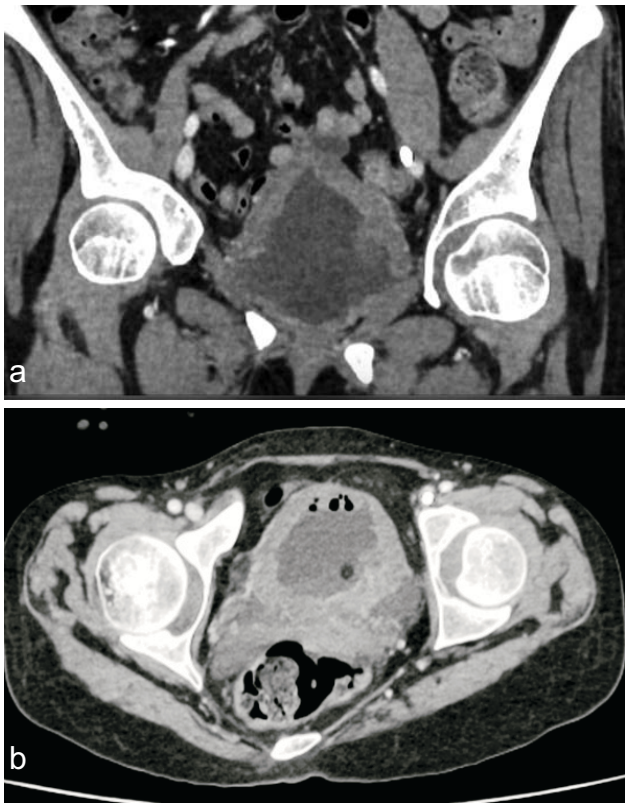


Figure 1(a,b): Shows pear shaped urinary bladder with significantly thick walls and trabeculated margins and multiple diverticulae

Figure 2(a,b): Shows mild to moderate bilateral hydronephrosis and hydroureter. Cortical irregularity bilaterally and cortical thinning on left side likely representing chronic pyelonephritis secondary to vesicoureteric reflux

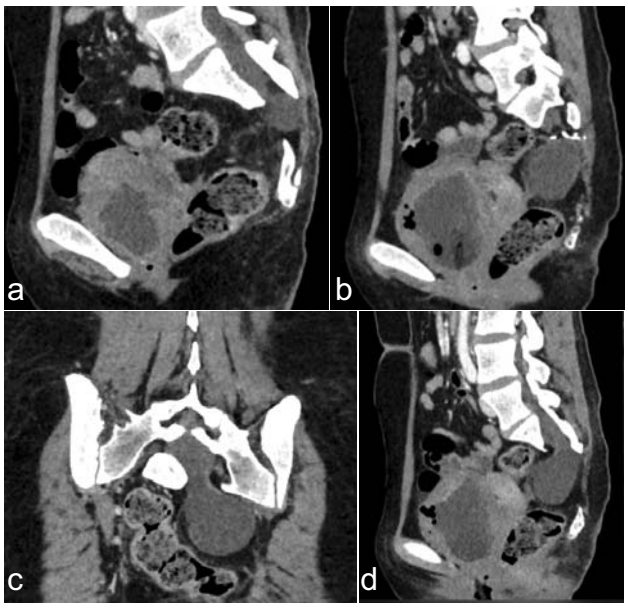


Figure 3(a,b,c,d): Shows deformed and tilted sacrum with anterior sacral defect on left side with herniation of dura mater from this defect into retrorectal region forming a bilobed cystic structure without any solid component or thick enhancing walls.

phy of pelvic bones on the right side was noted resulting in asymmetry (Fig.4). Findings were concluded as sacral spinal dysraphism with anterior sacral meningocele and neurogenic bladder resulting in bladder outflow obstruction and vesicoureteric reflux. The rest of the scan was unremarkable.

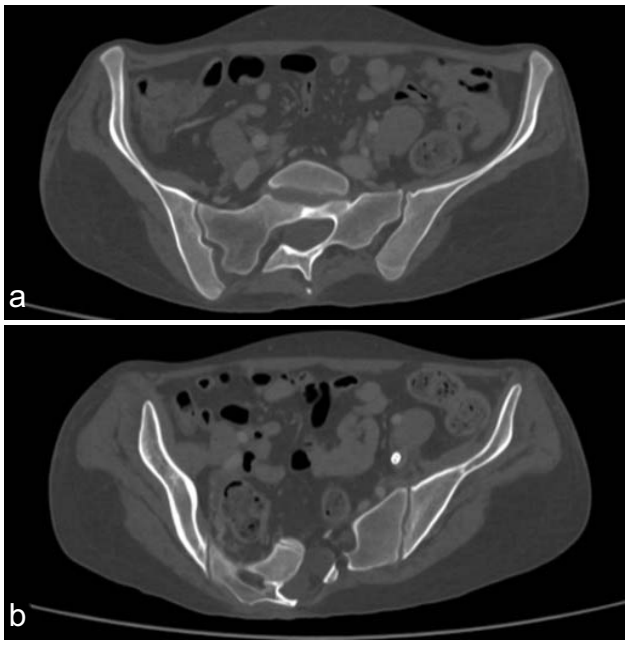


Figure 4(a,b): Shows Atrophy of pelvic bones on the right side resulting in asymmetry.

Her MCUG showed typical christmas tree appearance of urinary bladder representing Neurogenic bladder (Fig.5). Her MRI brain including Cervico vertebral juntion and lumbosacral spine was done next. Brain including CVJ was unremarkable however her lumbosacral spine showed straightening of lumbar spine and abnormal configuration of sacrum with malpositioned sacroiliac joints, low lying cord tethered to thecal sac at S1 level with possibility of lipoma. Anterior meningomyelocele was seen in the pelvic region on the left side at the level

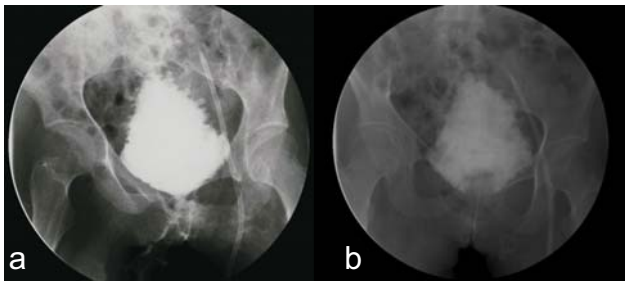


Figure 5(a+b): Shows typical christmas tree appearance of urinary bladder representing Neurogenic bladder

of S3 with thick walls. (Fig. 6 & 7) Rest of the study was unremarkable. Contrast enhanced MRI lumbar and sacral spine was recommended. The patient was conservatively treated and discharged.

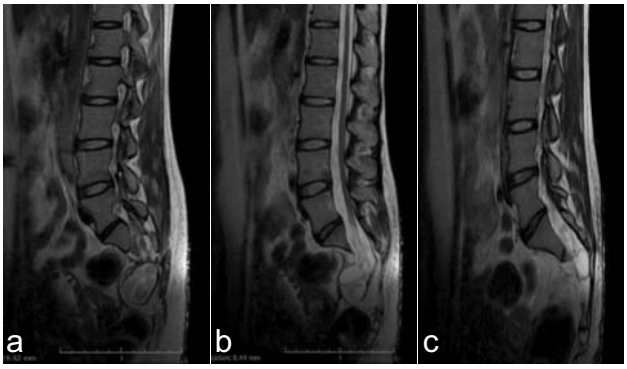
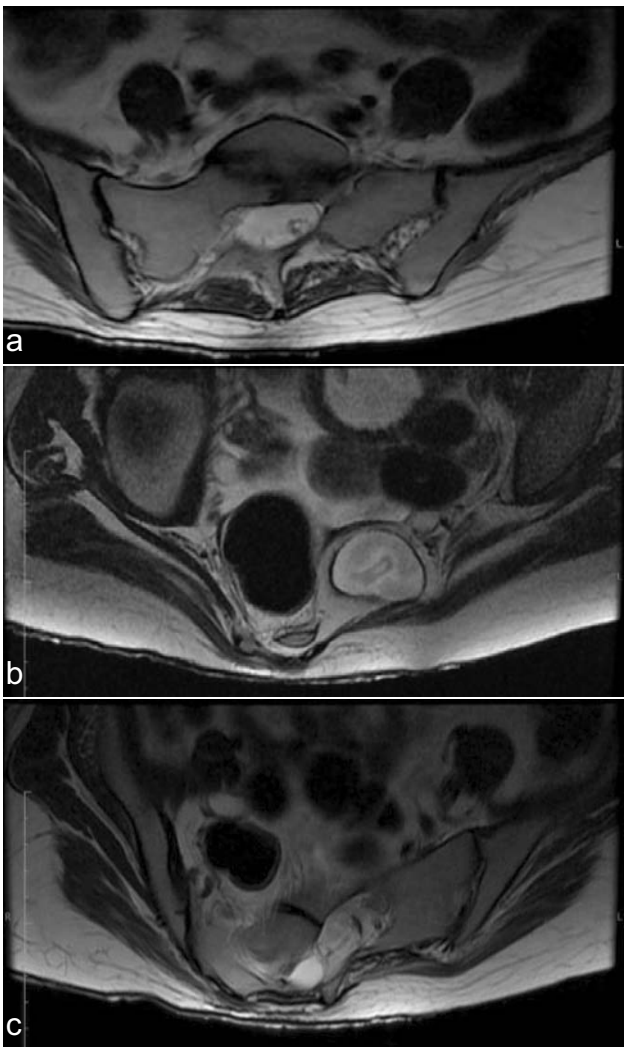


Figure 6(a,b,c): Straightening of lumbar spine ,low lying cord tethered to thecal sac at S1 level with a lipoma. Anterior meningomyelocele was seen in the pelvic region on the left side at the level of S3 with thick walls.



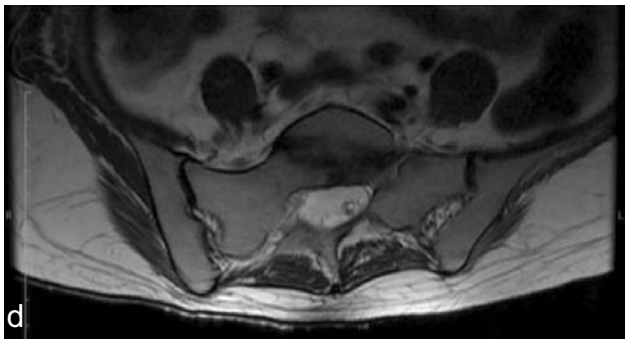


Figure 7(a,b,c,d): Abnormal configuration of sacrum with malpositioned sacroiliac joints, low lying cord tethered to thecal sac at S1 level with a lipoma. Anterior meningomyelocele was seen in the pelvic region on the left.

Discussion

Anterior sacral meningocele (ASM) is a rare subtype of spinal dysraphism in which the dural sac herniates through a defect in the anterior sacrum, sometimes manifesting clinically as a pelvic or abdominal mass.[1,3] The meningocele sac is made up of an inner arachnoid layer and an outer dural layer. It usually contains cerebrospinal fluid, though there are occasionally neural components. Symptoms usually arise from compression of adjacent pelvic structures and may include constipation, urinary urgency or incontinence, pregnancy-related dystocia, lower-limb pain or paresthesia, and headaches.[3] The incidence of ASM is not well established, and since Bryant's initial description in 1937, roughly 300 cases have been reported worldwide.[6] While posterior meningoceles are typically identified and treated during infancy, anterior sacral meningoceles typically manifest later and are most frequently found in female children or young adults.[4] A female predominance is noted, with an approximate 3:1 female-to-male ratio.[3] Although their cause is unknown, anterior sacral meningoceles are thought to be congenital abnormalities that enlarge as dural ectasia progresses under the influence of CSF pulsation. These pulsations promote expansion of the dural sac, which then herniates through a defect in the sacrum.[1,5] While neurological problems are rare, meningitis, sepsis, obstetric complications, and bladder or bowel issues can occur in affected individuals.[5] Weakened dural sac resistance is predisposed by several conditions, including Marfan syndrome, Ehlers-Danlos syndrome, neurofibromatosis types I and II, and prior sacral trauma. Marfan syndrome exhibits the strongest correlation among these.[1,3] Anterior sacral meningoceles are not associated with hydrocephalus or Chiari malformation, in contrast to posterior meningoceles.[4] In most cases, anterior sacral meningoceles do not exceed 5 cm in size.[3]

US is also useful in follow-up of nonsurgical patients. CT may better delineate the bony defects of the anterior sacral wall than any other imaging modality. Injection of intrathecal contrast media can reveal the relationship of the dural sac to the presacral cyst and the solid components of the cystic mass.

Because of its multiplanar imaging capability, MRI is the preferred modality for diagnosing ASM. Simple cysts with no solid components are hypointense on T1W, and hyperintense on T2W, sequences. Contrast media can be used to demonstrate associated pathological conditions, such as a lipoma or a dermoid tumor. In some cases, the neural components may be present in the cyst and are detected with MRI. In most cases, the spinal cord and the caudaequina are normal in ASM, but in rare cases, a tethered cord may be present. The conus medullaris may be well delineated on sagittal MRI views.[2]

In some patients, tethered cord syndrome is asymptomatic until adolescence, a situation that can be explained by clinical and experimental reports, that show a delicate balance between skeletal maturation and traction on the tethered conus.

Because the spinal cord grows more slowly than the spinal column, the cord becomes stretched and stressed over time and causes neurological damage. Bending or stretching the body puts extra tension on the tethered spinal cord.

The study by Kang et al. supports the hypothesis that the early stage of tethering is characterized by spinal cord dysfunction due to mechanical and vascular damage of a spinal cord segment, leading to widespread involvement of the cord with ischemia in a prolonged effect of tethering during growth.[7]

Another case of a 5-year-old boy with incontinence of urine and faeces since birth was reported. His abdominal MRI and sacral CT revealed a cystic presacral mass that communicated with the spinal dural sac through a neck that traversed a sacral bony defect and tethered spinal conus. On neurological examination the patient was found to have neurogenic bladder. Improvement after gradual treatment of his tethered cord neurogenic bladder.[8]

Surgery is the recommended treatment for symptomatic or growing anterior sacral meningoceles because these lesions do not resolve spontaneously and the risk of complications increases over time. Prophylactic surgery may be necessary for women with large lesions in order to avoid obstetric complications.[3,5]

Conclusion

The anterior sacral meningocele is a rare condition that can be hard to diagnose because it is located deep in the pelvis and can cause a lot of different symptoms. In rare cases, a tethered cord may be present. CT scan and MRI together prove to be important tools for diagnosing it.

CONSENT FOR PUBLICATION : The patient's permission has been obtained.

CONFLICT OF INTEREST : The authors declare no conflict of interest.

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