

## Asymptomatic large anterior sacral meningocele and bicornuate uterus: Imaging findings

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**Section:** Neuroradiology

**Area of Interest:** Pelvis Bones

**Procedure:** Diagnostic procedure

**Imaging Technique:** Ultrasound

**Imaging Technique:** CT

**Special Focus:** Cysts Congenital Case Type: Clinical Cases

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**Patient:** 17 years, female

### Clinical History:

A 17-year-old female patient presented at the outpatient urology department, with urinary retention which started a few hours ago. There were no other related complaints and the clinical history was unremarkable. A transurethral bladder catheter was placed and the patient was sent for imaging studies to further investigate the case.

### Imaging Findings:

A pelvic ultrasound revealed a large cystic lesion situated at the lesser pelvic cavity. The lesion was well-defined, except for a small part of its posterior margin, and was displacing the adjacent organs. A note was also made of a bicornuate uterus which was anteriorly displaced (Fig 1). A CT was then requested, and revealed a 121x117x119mm space-occupying lesion with cystic characteristics, smooth margins, located in the pre-sacral space. The cystic lesion was causing a mass effect to the pelvic organs with anterior displacement of the uterus, the sigmoid and the urinary bladder. The cystic lesion appeared to originate from the level of the enlarged third anterior sacral foramen and the sacral bone had a sickle-shaped configuration (scimitar sacrum), with a defect below the third anterior foramen onwards. The bicornuate configuration of the uterus was also confirmed (Fig. 2). A volume rendering technique reconstruction was used to visualise the sacral bone deformity (Fig. 3).

### Discussion:

Anterior Sacral Meningocele (ASM) is a rare congenital abnormality affecting 1 in 40.000 individuals [1]. ASM is an anomaly associated with various degrees of sacral bone agenesis allowing the herniation of a CSF-filled meningeal sack into the pre-sacral pelvic space [2]. ASM was first described by Bryand T. and reported in the Lancet in 1837. Since then, fewer than 300 cases have been reported in medical literature [3, 4, 5, 6]. The congenital form of ASM is a presentation of spinal dysraphism and a minority of cases show an inherited predisposition [7, 8, 9, 10]. In the case of spinal dysraphism, ASM results from failure in fusion of the sacral bone, which allows the protrusion of the

meningeal sac through the sacral hollow [11]. ASM can also appear without the presence of a dysraphic vertebral anomaly, usually associated with Marfan syndrome, neurofibromatosis type NT1 or Ehlers Danlos syndrome [11, 12, 13]. ASM can be associated with a wide range of other malformations like lipomas, teratomas, imperforated anus, duplication of uterus and vagina, kidneys and ureters, as it was the case in our patient where ASM coexisted with bicornuate uterus [2, 14]. AMS exists also in the context of Caudal Regression Syndrome (Caudal Agenesis), as part of the Currarino triad [15, 16]. As opposed to posterior meningoceles, which are protruding from the posterior body surface and therefore are easily recognised, ASM is hidden in the pelvis (truly occult spinal dysraphism [15]) and can remain undiagnosed until it is incidentally discovered or when it causes symptoms like constipation, urinary retention or incontinence. Other complaints include dysmenorrhea, dyspareunia, sciatica, intermitted positional high or low headache due to fluid shifts between ASM and the spinal subarachnoid space. It can rarely present with superinfection and meningitis [9].

The clinical manifestation of ASM is often nonspecific, making imaging essential for diagnosis. Conventional radiography may reveal the "Scimitar Sign" referring to the sacral deformity of the unilateral sickle-shaped sacral defect with smooth margins, produced by the ASM. The scimitar sign is highly suggestive for ASM and is present in at least half of the cases. Grayscale U/S may visualise the presacral hypoechoic mass. A CT accurately assesses the osseous defect and the meningocele neck. CECT might show nerve roots transversing the sacral defect and might help differentiation from lipomas or dermoids. A MRI can provide much more information regarding cystic and thecal sac community, cord tethering and differential diagnosis demonstrating nerve roots transversing a sacral defect and CSF composition of the fluid [1, 9, 17, 18].

**Differential Diagnosis List:** Anterior sacral meningocele and bicornuate uterus., Sacrococcygeal teratoma, Sacral chordoma, Neurenteric cyst, Cystic neuroblastoma, Ovarian cyst, Rectal duplication cyst, Abscess, Simple bone cyst, Aneurysmal bone cyst

**Final Diagnosis:** Anterior sacral meningocele and bicornuate uterus.

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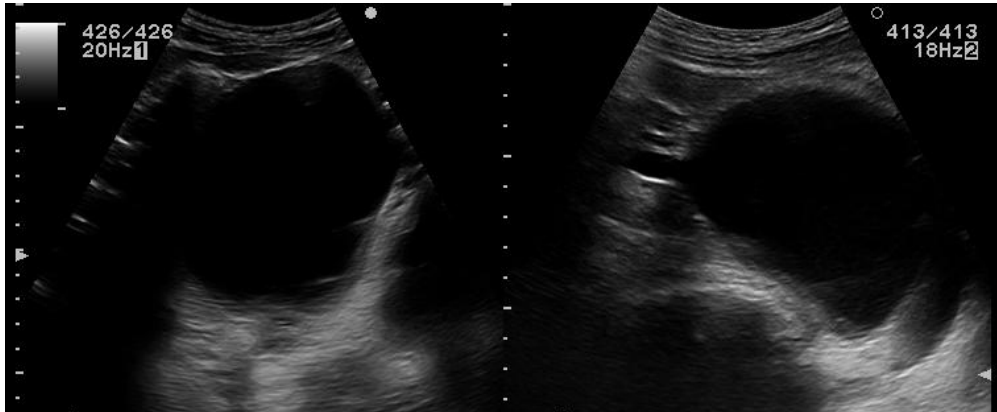
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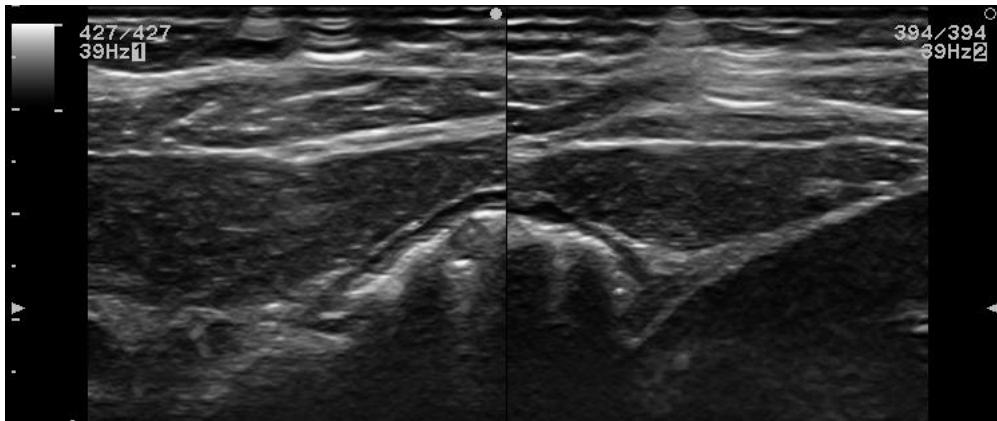
**Figure 1**

**a**



**Description:** A transverse split-screen ultrasound image obtained over the hypogastrium showing the cystic lesion with well-defined borders except for a small ill-defined area in its posterior wall. **Origin:** Radiology Department, "G. Gennimatas" Hospital, Thessaloniki

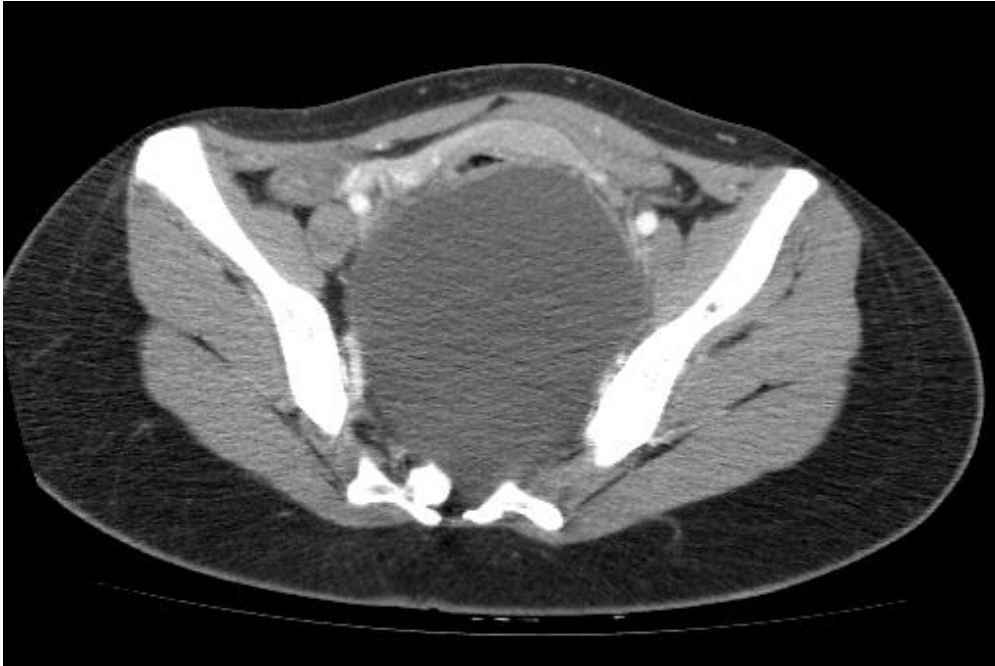
**b**



**Description:** A transverse split-screen ultrasound image obtained over the hypogastrium showing the two solid echogenic structures, corresponding to the two horns of a bicornuate uterus. **Origin:** Radiology Department, "G. Gennimatas" Hospital, Thessaloniki

**Figure 2**

**a**



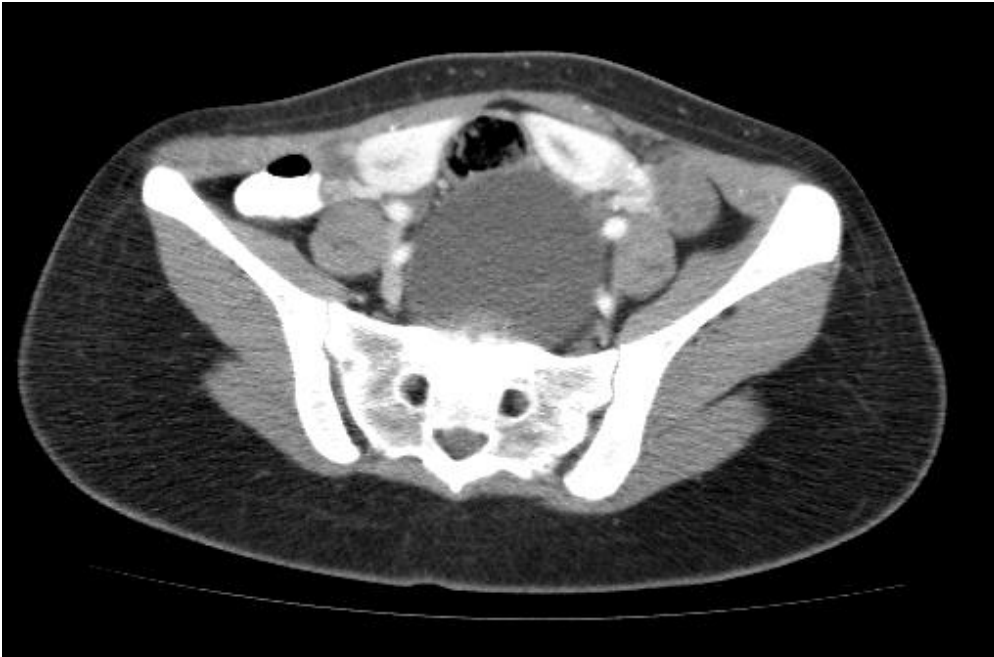
**Description:** Axial CT image showing the cystic lesion originating from the level of the third anterior sacral foramen. **Origin:** Radiology Department, "G. Gennimatas" Hospital, Thessaloniki

**b**



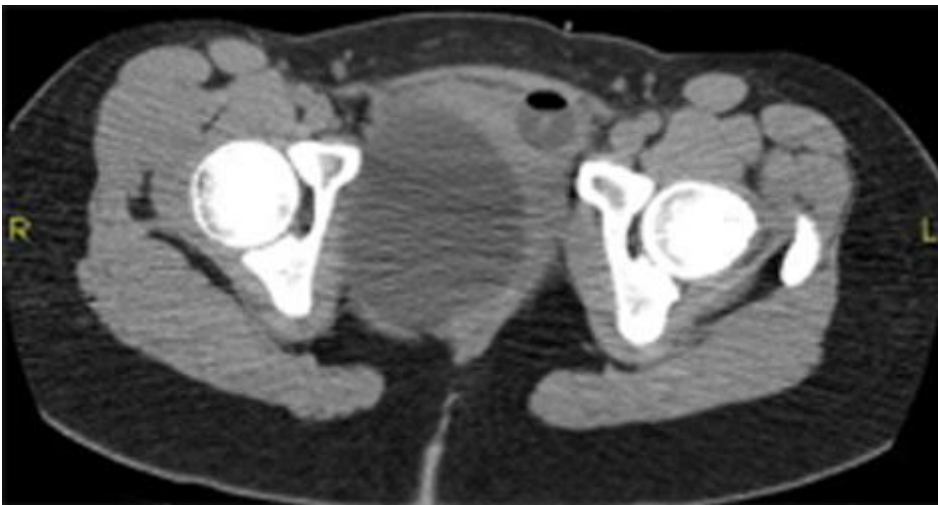
**Description:** Oblique multi-planar reconstructed CT image showing the communication between the cystic lesion and the spinal canal. **Origin:** Radiology Department, "G. Gennimatas" Hospital, Thessaloniki

c



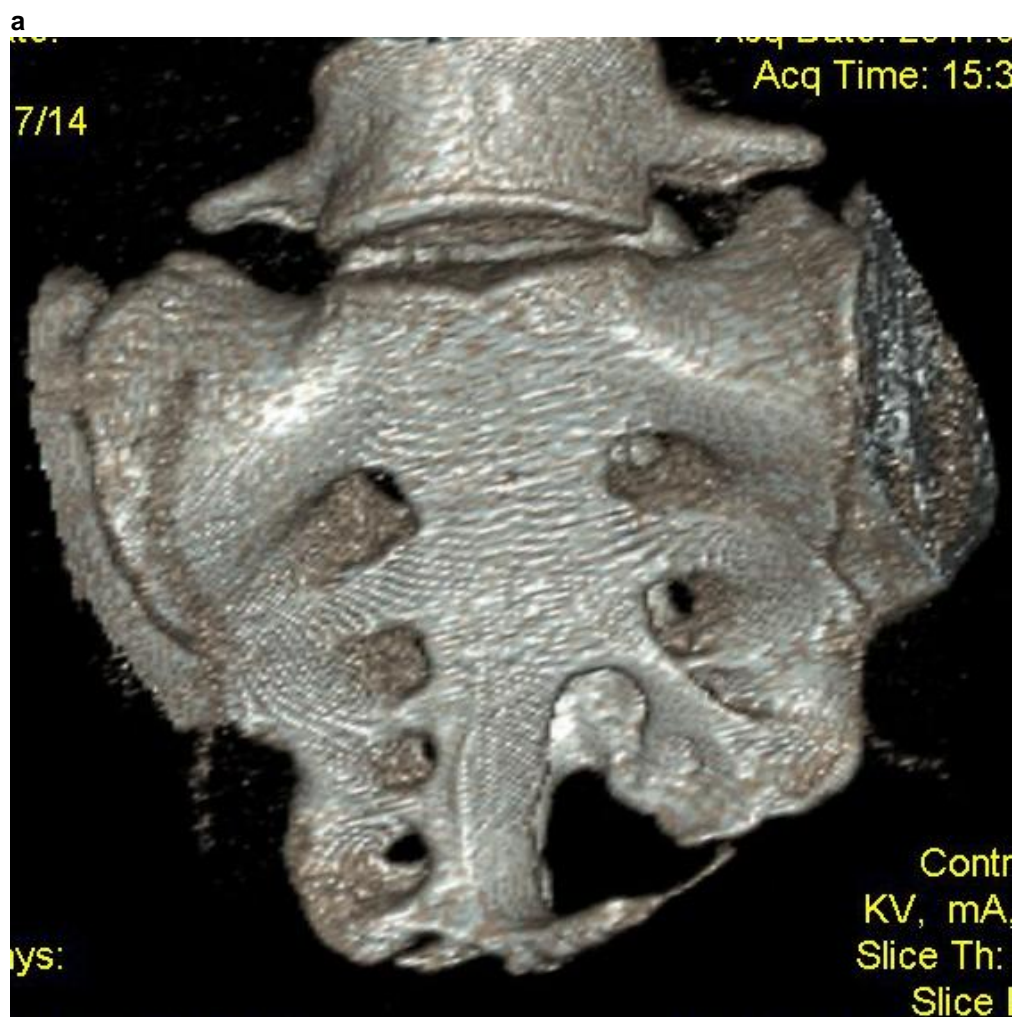
**Description:** Axial CT image showing the two horns of the bicornuate uterus. **Origin:** Radiology Department, "G. Gennimatas" Hospital, Thessaloniki

d



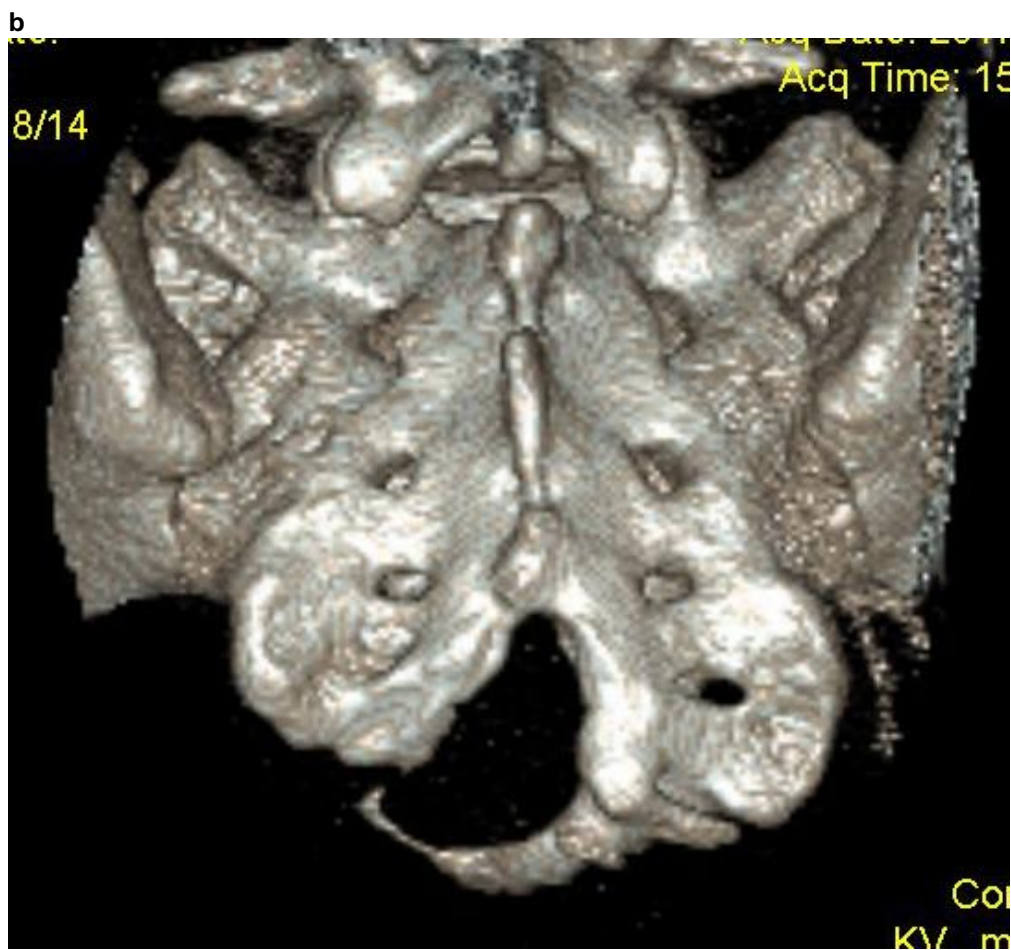
**Description:** Axial CT image showing the anteriorly displaced urinary bladder containing the catheter. **Origin:** Radiology Department, "G. Gennimatas" Hospital, Thessaloniki

**Figure 3**



**Description:** Volume rendering technique CT image showing the anterior surface of the sacral bone with its deformities. **Origin:** Radiology Department, "G. Gennimatas" Hospital, Thessaloniki





**Description:** Volume rendering technique CT image showing the posterior surface of the sacral bone with its scimitar sacrum configuration. **Origin:** Radiology Department, "G. Gennimatas" Hospital, Thessaloniki