Case 2309

Primary amenorrhea due to Müllerian duct anomalies
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Section: Genital (female) imaging
Imaging Technique: Ultrasound
Imaging Technique: MR
Case Type: Clinical Cases
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Patient: 15 years, female

Clinical History:

A 15-year old female patient was referred to our institution because of bilateral lumbar and pelvic pain.

Imaging Findings:

A 15-year old female patient was referred to our institution because of bilateral lumbar and pelvic pain. Pain was lasting since 10 months and was continuous, with variable intensity, but no relationship with the menstrual cycle, that was normal. Gynaecological examination demonstrated a large palpable pelvic mass, displacing the left lateral wall of the vagina, and extending superiorly until the transverse umbilical plane. The patient underwent ultrasound and MR imaging of the pelvis (the exams were performed during the menstrual period). The diagnosis was confirmed by surgery.

Discussion:

Uterus didelphys with an imperforate hemivagina is an embryonic malformation of the genitourinary system of the female that occurs between the 12th and 16th week of pregnancy. It is caused by the failure of fusion of the paramesonephric (Mullerian) ducts. This defect may manifest itself as a duplication of all or part of the female reproductive system. The obstruction of one hemivagina will block outflow, resulting in complications such as hematocolpos, hematometra and hematosalpinx. The persistence of this situation also may be complicated by the occurrence of endometriosis as a result of blood reflux into the abdominal cavity. These anomalies frequently are accompanied by kidney and urinary tract malformations (i.e., kidney agenesis and dysplasia, double collecting system, ectopic ureter) on the same side as the defect. The manifesting symptoms usually appear only after menarche and consist of dysmenorrhea, severe abdominal pain, and the presence of an intraabdominal or pelvic mass. Ultrasonography is the first choice technique: after a careful anamnesis and physical examination, transabdominal US can confirm suspicions in most cases revealing the existence of two hemiuteruses, hematocolpos and eventually the presence of unilateral renal agenesis. After either an inconclusive US or an US examination confirming the suspect, the MR imaging is mandatory in order to obtain a precise diagnosis or to have a definitive preoperatory confirmation and a multiplanar spatial balance. MR imaging completes the diagnostic sequence in an non-invasive way and allows an early and limited surgical treatment, substituting explorative laparotomic or laparoscopic procedures and assuring the best fertility prognosis. The aim of surgery is preservation of normal fertility. Excision of the vagina septum is recommended as the procedure of choice.
**Differential Diagnosis List:** Hematocolpos with uterus didelphys and vaginal duplication.

**Final Diagnosis:** Hematocolpos with uterus didelphys and vaginal duplication.

**References:**


**Figure 1**

**a**

Description: Transverse US image shows the presence of a mass (*) located posterior to the urinary bladder (B), with disomoegenous liquid content. Origin:

**b**

Description: US transverse image shows the mass (*) located posterior to the urinary bladder (B) and allows to visualize the uterus with two endometrial cavities uniformly separated (U). Origin:
Description: The sagittal acquisition confirm the presence of a liquid filled formation whose content has signal characteristics of blood (*); these images allow to identify the finding as a hematocolpos in a blind left and diluted hemivagina (*). B= bladder. Origin:
Description: The sagittal acquisition shows the hematocolpos in a blind left and dilated hemivagina (*) in a different plane.
B= bladder. Origin:
Description: The coronal acquisition allows to visualize the hematocolpos (*) and the didelphic uterus with complete separation of uterine horns (arrow). Origin:
Description: In this different plane the arrow indicates also the presence of the two different cervices. Hematocolpos (*) is always well appreciable. Origin:
Description: Axial T1wi MR imaging shows the hemocolpos (*) in left hemivagina (L); the right hemivagina is compressed (R). Origin: