Herlyn-Werner-Wunderlich syndrome: triad of didelphys uterus, obstructed hemivagina and ipsilateral renal agenesis

Published on 15.09.2014

DOI: 10.1594/EURORAD/CASE.12097
ISSN: 1563-4086
Section: Genital (female) imaging
Area of Interest: Genital / Reproductive system female
Procedure: Diagnostic procedure
Imaging Technique: Ultrasound
Imaging Technique: MR
Special Focus: Congenital Case Type: Clinical Cases
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Patient: 13 years, female

Clinical History:

A 13-year-old female patient presented with a history of severe abdomino-pelvic pain that occurred since the patient reached menarche and irregular menses.

Imaging Findings:

A pelvic ultrasound was performed initially, showing two widely divergent uterine horns with separate, noncommunicating endometrial cavities. Two cervices and duplicated upper vaginas were present. Right haematocolpos was documented. Right renal agenesis, not known before, was revealed. MRI followed for more detailed evaluation, confirming the presence of a didelphys uterus and a double vagina. The right hemivagina was obstructed and considerably dilated by the accumulation of fluid exhibiting high signal intensity on T1 sequences, due to the presence of bloody contents, consisting with haematocolpos. The right uterine cavity, in continuity with the obstructed hemivagina, was slight distended. The ovaries showed normal morphology with several subcentimetre follicles.

Extension of the MRI examination to the upper abdomen showed right renal agenesis and a vicariant left kidney.

Discussion:

Herlyn-Werner-Wunderlich syndrome (HWWS) is a rare congenital anomaly of the urogenital tract involving Müllerian ducts and Wolffian structures, and it is characterized by the triad of didelphys uterus, obstructed hemivagina and ipsilateral renal agenesis. [1]

HWWS is usually discovered at puberty with non-specific symptoms, like increasing pelvic pain, dysmenorrhoea and palpable mass due to the associated haematometrocolpos, which result from retained, longstanding menstrual flow in the obstructed hemivagina. [1]

Endometriosis is a frequent complication of this syndrome. This is because of obstructed outflow of a part of the menstrual blood, causing retrograde menstrual flow, leading to haematometra, haematosalpinx, peritoneal and adnexal endometriosis. [2]

A delay in diagnosis is not uncommon. Menstruation is often regular in the context of an incomplete vaginal outlet obstruction and slow extension of haematocolpos occurs. When patient complains of symptoms of cyclic
dysmenorrhea, anti-inflammatory drugs and oral-contraceptives are usually prescribed, which reduces or eliminate menses; after all, this is an uncommon syndrome, not often thought of as a diagnostic possibility. [1, 3]

US is usually the first imaging modality used and allows a correct diagnosis in most cases. [4] MRI provides more detailed information regarding uterine morphology, the continuity with each vaginal (obstructed and non-obstructed) lumen, and the bloody nature of the contents.

Uterus didelphys is a symmetric anomaly in which two completely separate uterine cavities are identified, each with normal zonal anatomy as well as its own endometrial cavity and cervix. No communication between the two cavities is present. [5]

There are two hemivaginas, one obstructed and another with a normal outflow tract. Obstruction is due to a longitudinal obstructing vaginal septum, which occludes one cervix and isolates it. [1]

Trapped secretions and menstrual blood are seen on MRI, distending the obstructed hemivagina (haematocolpos) and the ipsilateral uterine cavity (haematometra), best appreciated on T1-weighted imaging (with fat suppression) where any blood products remain of high signal intensity.

An important point to note is that renal agenesis is typically ipsilateral to the vaginal anomaly (right side prevalence). [2]

Additionally, MRI can detect associated findings such as adnexal and peritoneal endometriosis, pelvic inflammation and adhesions. [2]

Laparoscopic vaginal septum excision is the treatment of choice for relieving the vaginal obstruction in HWWS. MRI plays an important role in pre-operative planning.

It should be stressed that whenever the pelvic MRI shows a didelphys uterus in association with obstructed hemivagina, the examination should be extended to the upper abdomen to check for agenesis of ipsilateral kidney.

**Differential Diagnosis List:** Herlyn-Werner-Wunderlich syndrome, Unicornuate uterus with renal agenesis, OHVIRA syndrome

**Final Diagnosis:** Herlyn-Werner-Wunderlich syndrome

**References:**


Description: Longitudinal US image show dilated hemivagina containing low-level echoes, consisting with haematocolpos. Origin: Maciel C, Department of Radiology, Hospital de São João, Porto, Portugal
Description: Transverse US image shows widely diverging uterine horns of a didelphys uterus. Origin: Maciel C, Department of Radiology, Hospital de São João, Porto, Portugal
Figure 3

Description: Oblique coronal T2 weighted image depicts widely divergent uterine horns (arrows) of a didelphys uterus. Notice mild dilatation of right uterine cavity. Full urinary bladder (B) is seen. Origin: Maciel C, Department of Radiology, Hospital de São João, Porto, Portugal
Description: Oblique coronal T2 weighted image depicts the two cervices (arrow) of a didelphys uterus.
Origin: Maciel C, Department of Radiology, Hospital de São João, Porto, Portugal
Description: Axial T2-W image shows two cervices (arrows). Notice the mild dilated right uterine cavity (curved arrow) in continuity with the right cervix, which in turn communicates with a dilated right hemivagina (asterisk). Origin: Maciel C, Department of Radiology, Hospital de São João, Porto, Portugal
Figure 4

Description: Coronal T2 weighted image shows right renal agenesis, ipsilateral to the obstructed hemivagina. Note the haematocolpos (asterisk). A solitary left kidney with mild compensatory hypertrophy is identified (arrow). Origin: Maciel C, Department of Radiology, Hospital de São João, Porto, Portugal.
Description: Axial T2 (A) and T1 (B) weighted images show marked dilated right hemivagina with low T2-signal-intensity and high T1-signal-intensity contents, due to blood products presence, corresponding to haematocolpos (asterisk). Origin: Maciel C, Department of Radiology, Hospital de São João, Porto, Portugal
Description: Sagittal T2 weighted image depicts the haematocolpos (asterisk). Origin: Maciel C, Department of Radiology, Hospital de São João, Porto, Portugal