OHVIRA Syndrome, knowledge and review.

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Learning objectives

The Ohvira Syndrome is a rare congenital anomaly of urogenital tract that affects the Mullerian ducts and Wolff. It consists of the triad uterine didelphys associated with Obstructed hemivagina and Ipsilateral Renal Anomaly.

The objectives of this work is to present a review of this congenital anomaly of the urogenital tract and expose key radiological diagnosis through our experience in diagnosing the Ohvira Syndrome (O.S).

Background

The Herlyn-Werner-Wunderlich Syndrome (.H.W.W.S) is a rare congenital malformation of the Mullerian ducts described in 2006.

It consists of the triad Fig. 1 on page 5 : uterus didelphys, obstruction hemivagina and ipsilateral renal agenesis, whose early detection is important to prevent symptoms and possible future complications.

Among the ductal Mullerian anomalies (DMA),.H.W.WS Fig. 2 on page 6 is the least common, but better prognosis.

OHVIRA Syndrome (uterine didelphys associated with Obstructed hemivagina and Ipsilateral Renal Anomaly), broader term than the HWWS, to include other types of renal abnormalities (which will be associated with developmental abnormalities of the Müller's ducts: renal agenesis, double collector, duplication renal system and horse-shoe kidney).

Epidemiology:

Etiology and pathogenesis are unknown.

Frequency:

Until 2011, only about 200 were documented cases in the world literature.

It has a low prevalence and an undetermined rate ;

- Unilateral renal agenesis' incidence is 1/600 - 1/1200 and
Prevalence of genital anomalies associated with renal abnormalities in women is 25-89%.

- Prevalence of congenital uterine anomalies is approx. 6.7% of fertile people, 7.3% and 16.7% in infertile women with a history of recurrent abortion

**Embryology:**

There is an intimate relationship during embryogenesis between the urinary and reproductive systems that can explain the coexistence of abnormalities in the urinary tract and reproductive systems.

HWWS may represent a developmental abnormality both Müllerian (Paramesonephricus) as Wolffian (mesonephric).

The internal genital organs come from the paramesonephric ducts (Müller) and upper urinary tract derived from the mesonephric ducts (Wolff).

- In women, the Mullerian ducts, located laterally to the Wolffian ducts grow caudally and towards midline crossing them to contact, merge and form the uterovaginal canal from which the tubes are developed, the uterus and the 2/3 upper vagina. After completion of the merger of the Mullerian ducts, head towards the dorsal portion of the urogenital sinus, from which the lower 1/3 of the vagina is developed.

- Wolff ducts give rise to kidneys, and are inducers of proper fusion of the Mullerian ducts. By this, an alteration in the development of one of the flow ducts portion Wolff, origin can be associated with unilateral renal agenesis ipsilateral hemivagina imperforate.

- Since the vagina has a mixed mesodermal 2/3 higher (Muller) and 1/3 lower endoderm, vaginal abnormalities (hipoplasia / aplasia, duplication / septum) may occur in association with Müllerian anomalies or isolated respectively.

In the absence of the Wolffian duct, the duct of Müller moves laterally (in space where the Wolffian duct is absent) and may not be merged with contralateral duct, giving a uterus bicornis. The contralateral Müllerian duct gives rise to a vagina, while the displaced Müller form a blind pouch (obstructed or imperforate hemivagina). The introitus not be compromised, because its embryological origin comes from the urogenital sinus. The mesonephric remnant (Wolff) can stay in the vaginal wall and sometimes become cyst (cyst or Gartner duct).

**Classification:**

Ductal Mullerian Anomalies (DMA) are congenital malformations of the female genital tract as a result of the lack of development and / or absence of fusion of the Mullerian ducts or piping by default for lack of resorption of the uterine septum.
The absence of merger or formation of the distal segments of the Mullerian ducts can lead to 7 types of uterine abnormalities, grouped according to the specific type of embryological disturbance that originates, the most used of the American Society of Reproductive classification Medicine (ASRM) Fig. 2 on page 6.

The incidence of ductal Müllerian anomalies (DMA) is 0.5 to 5% being the most common type septate uterus and less frequent but better prognosis the uterus didelphys.

The HWWS sometimes it is diagnosed so late and erroneous, because of ignorance as cause of symptoms or disregard the embryological origin of the components of the genitourinary tract.

**Clinic:**

Asymptomatic until menarche because of the absence of specific symptoms.

Symptoms depend primarily on the vaginal obstruction.

1. Clinical presentation after menarche.

2. Cyclical pelvic pain, recurrent and progressive, secondary to hematocolpos (due to the accumulation of haematic content on blind hemivagina), can also be haematometra, haematosalpinx even haemoperitoneum.

3. Pelvic mass with or without clinical described.

4. Problems associated:

   Early pelvic endometriosis (due to reflux of blood in the abdominal cavity), infertility, menstrual disorders and obstetric complications such as infection collections (pyocolpos, pyometra or pyosalpinx), urinary retention, etc.

**Diagnosis:**

**A) From suspicion:**

1. A fetus or newborn:

   - Diagnosed of multicystic dysplastic kidney or absence of kidney would be wise to seek Muller duct obstruction (look for genital abnormalities).

   With presence of cystic mass further bladder and absence of kidney associated consider didelphic uterus, imperforate vagina and hidrocolpos.
- Uterine anomalies are best detected in the neonatal period where the uterus is still under maternal hormonal stimulation (prominent echogenic endometrium and myometrium) and worse in childhood, where the uterus size and shape do not allow diagnosis, to menarche.

2. In adolescents may be suspected by clinical but usually is nonspecific. Consider that in patients with Müllerian duct anomalies (MDA) be binding assess the urinary tract, because the association of renal tract anomalies (more frecuent Right kidneyl agenesis, although the cause is unknown) with MDA, more than 30 % of cases are due to a common embryonic origin.

**B) Radiological manifestations:**

Suspecting MDA, first test is ultrasounds and it will should be followed by MRI which is the test of choice for diagnosis.

Ultrasound to diagnose: utero-vaginal duplication, haematocolpos, haematometra Fig. 4 on page 7 Fig. 10 on page 13 (fluid collections with echoes), suspected uterine anomalies (didelphic or bicorn bicolis), absence of ipsilaterial kidney, multicystic dysplasia, and in rare cases hydronephrosis secondary Fig. 3 on page 7 Fig. 4 on page 7 Fig. 10 on page 13

The acquisition of multiplanar MR images can assess uterine morphology, obstruction or otherwise of each channel and the nature of the content (pT1-FS). It is more sensitive to evaluate uterine contour, uterine cavity's shape, septum's characteristics, and associations such as endometriosis, pelvic inflammation and adhesions.

Laparoscopy can be used as ransom when diagnosis is not fully clear with RM.

**Differential Diagnosis:**

Differential diagnosis is made with other abnormalities of the uterine development as uterus bicornis, unicorn uterus with rudimentary horn contralateral non-communicating, imperforate hymen and hypoplasia or agenesis of the cervix.

**Treatment:**

The treatment is surgical resection of the septum and draining obstructed vagina, allowing relieve acute symptoms, resolving obstruction to prevent endometriosis and fertility preservation

**Images for this section:**
Fig. 1

- **I Hypoplasia/agenesis**
  - (a) Vaginal
  - (b) Cervical
  - (c) Fundal
  - (d) Tubal
  - (e) Combined

- **II Unicornuate**
  - (a) Communicating
  - (b) Non-communicating

- **III Didelphys**
  - (a) Complete
  - (b) Partial

- **IV Bicornuate**
  - (a) Complete
  - (b) Partial

- **V Septate**
  - (a) Complete
  - (b) Partial

- **VI Arcuate**

- **VII DES drug related**

*Fig. 1—Classification system of müllerian duct anomalies used by the American Society for Reproductive Medicine* [3] (Reprinted with permission). DES=diethylstilbestrol.
Fig. 2

Fig. 3: Intravenous urography: Absence of left kidney and ureter. Right hydronephrosis grade IV. Hypodense pelvic mass protruding caudally on the bladder
Fig. 4: Abdominal US: Right hydronephrosis grade IV with dilatation of the excretory system (up to 18 mm proximally). Echogenic Collection in lesser pelvis was consistent in hematocolpos
Fig. 5: MRI: Ax T2w: Uterus didelphys with presence of two cavities, the right one with hyperintense contents and the left one hypointense relative to hematometra. Cor T1w: Communication between left hemiuterus and blind hemivagina
Fig. 6: MRI: Cor T1w: Communication between left hemiuterus and blind hemivagina
Fig. 7: T2 weighted sagital MR images showing: Uterus with blood content (hematometra) (yellow) and normal uterine content (blue) Enormeous cavity, 170 mm diameter CC, hypointense with content that corresponds to the imperforate vagina (green) regarding hematometracolpos and mass producing effect on adjacent structures as the bladder. (red)
Fig. 8: T2 weighted sagital MR images showing: Uterine cavity (yellow) in communication with imperforate hemivagina (green), with hipointense material related to subacute hematic content (hematometracolpos) and a small cavity (blue) which corresponds to hemiuterus with hyperintense content that is bound to normal hemivagina.
**Fig. 9:** T1 and T2 weighted axial MR images showing Normal right hemivagina (yellow) Hiperintense collection in T1w and hipointense in T2w in imperforate left hemivagina regarding hematic content (red).
Fig. 10: Upper images: Abdominal US: Right kidney not visualized (agenesis, hypoplasia, atrophy?) and left kidney increased in size (12.5 mm), probably compensating with good cortico medullary differentiation and showing no dilatation of the excretory system. Below images: Abdominal US: A midline level pelvis, an heterogeneous echogenicity collection with solid and cystic areas without evidence of vascularization with Doppler ultrasound, which appears to be intrauterine collection. Double uterus was suspected with hematometra so MRI study was done.
Fig. 11: T1 and T2 weighted axial MR images showing a vaginal fluid collection in relation to evolved hematocolpo (Pink).
**Fig. 12:** Left image: T2 weighted coronal MR image showing 2 hemivaginas divided by septum (Red) hypointense Right image: T2 weighted sagital MR images showing an uterus didelphys retroverted (Yellow) with pathways uterine cavities filled liquid material
Findings and procedure details

- Findings

1. **Vertical Septo vaginal** that allows obstruction, Fig. 12 on page by alteration in the lateral fusion of Müllerian ducts downstream, causing division of the vaginal cavity giving rise to two hemivaginas, one of which communicates with the outside and the other not, which condition the appearance of haematocolpos.

Septum is not observed by US, but haematocolpos, is identify as a liquid heterogeneous image with marked posterior acoustic enhancement.

RM septum appears in 75% of patients and, generally better observed in coronal or axial plane as linear fine structure with low intensity signal in different sequences. The pT2 sequences allow testing the low signal intensity of the septum with hyperintensity of the vaginal mucosa and inner secretions. Haematometra Fig. 5 on page Fig. 7 on page Fig. 8 on page haematocolpos Fig. 6 on page Fig. 7 on page Fig. 8 on page Fig. 9 on page Fig. 11 on page, are better characterized by MRI, and their appearance will depend on the bleeding time. Blood can be seen subacute (1-4 wk) hyperintense on both sequences.

2. **Didelphic Uterus**: Fig. 5 on page Fig. 8 on page Fig. 12 on page

This is the result of a lack of fusion of both ducts of Müller, resulting in two completely separate cavities, without communication with each other. Thus, the uterus is presented with two separate and divergent horns, associated with two uterine cervix.

May be suspected by US, but diagnosis by MRI is more accurate.

Differentiate uterus bicornis bicollis (with some degree of sustained communication between the two horns) and didelphys uterus, it may not be easy, even by MRI; in bicollis will be a big cleavage of the fundus and the didelphic, distinct two uterus, also in 75% of the uterus didelphys there is a longitudinal vaginal septum which is present in 25% of bicornes. Identify unilateral renal absence by MRI will help the diagnosis.

3. **Agenesia renal**: Renal congenital anomaly most frequently associated with Müllerian malformations. Typically ipsilateral to the vaginal abnormality (prevalence in right side) with possible presence of contralateral renal compensatory hypertrophy.
Suspected by US, Diagnosed by CT or MRI or even intravenous urography. Fig. 3 on page

4. Findings at ovarian level:

- Endometriosis cysts: hyperintense in pT1-FS, with "shading sign" in pT2 and variable signal due to chronic bleeding with accumulation of high concentrations of protein and iron in endometriomas (endometriotic lesion hyperintense in pT1 and hypo / hyperintense on pT2).

- Possible functional cysts without clinical relevance hyperintense on pT2 and hypo / isoointense in pT1.

Conclusion

Recognition and early surgical treatment of this rare entity is important to prevent symptoms, and complications, and preserve fertility.

Personal information

References

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