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A Rare Case of Herlyn-Werner-Wünderlich Syndrome with an Ectopic Ureter and A Communication Between a Hydrocolpos and The Bladder Neck in A Uterus Didelphys.

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Herlyn-Werner-Wünderlich síndrome,

1. Background

The HWW syndrome is clasified as a type III Müllerian disgenesia, that includes a uterus didelphys, a blind hemivagina and unilateral kidney agenesia [1]. Müller's ducts migrate to the midline and merge together to form the uterus, cervix and upper vagina at around the 8th week of pregnancy. Incomplete or absent fusion will produce two uteri. Kidney agenesia generally follows Müller duct malformation. The incidence of uterus didelphys related to HWW is approximately 1/2000 to 1/28.000 [2]. The purpose of this case description is to offer a better understanding of HWW syndrome pathophysiology, especially in adult patients presenting with longstanding common and unspecific symptoms.

2. Case Presentation

A 56-year-old patient with an infected hematometra is admitted to hospital. Her history includes right kidney agenesia and uterus didelphys diagnosed during a previous caesarean section, Gorlin's syndrome (multiple basal cell carcinoma), endometriosis and a breast cancer treated with a mastectomy and adjuvant chemotherapy and hormonotherapy. At admission the patient had a two-week complaint of pelvic pain and malaise accompanied by vertigo and a white coloring of her urine.

The patient also has a pelvic adhesive syndrome involving the anterior aspect of the uterus and the bladder diagnosed during a tubal ligation, and had presented hematuria from menarche to menopause. The patient had a history of urinary incontinence or vaginal discharge since her adolescence.

3. Investigations

Transvaginal ultrasound showed a unicollis bicornuate uterus. The left hemiuterus appeared normal with a uniform endometrium 6 mm thick and a right hemiuterus with a cystic contents 6-7 cm long, with a well-defined myometrium.

In an abdominal-pelvic magnetic resonance imaging (MRI) the uterus didelphys was also described with a single cervix visible. The left uterine horn was lateralized and in a slight retroversion without significant alterations. The right uterine horn was larger and in ante version. The images showed a 4 cm lesion suggestive of a myoma. The continuity of the right horn with the cervix could not be ascertained due to the existence of a 6x5 cm cystic lesion with three liquid-liquid levels that seems to connect with the endometrial lumen. This lesion is compatible with the original diagnosis of a non-communicating right hemivagina atresia.

The computerized tomography imaging results are similar, showing a right kidney agenesia and normal left urinary tract. A bicornuate uterus is described, the left horn connects with the cervix and the right horn has a cystic lesion in the isthmus-cervical area (Figure 1).

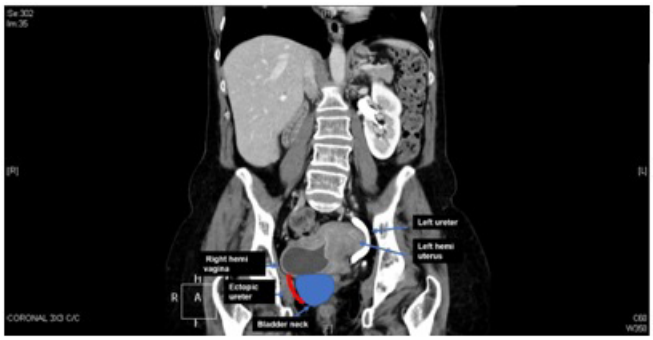

Figure 1: The computerized tomography

A diagnostic hysteroscopy was performed showing a single tubal ostium (probably the left one) or a rudimentary uterus. The was no septum or hemiuterus observed. This procedure also included a puncture of the right uterus under ultrasound guidance, obtaining 250 cc of purulent liquid. The biochemical analysis showed the presence of urea at a concentration of 545 mg% and a normal creatinine level (0,35mg%).

4. Treatment

A laparoscopic hysterectomy with bilateral adnexectomy was performed. During the procedure the right hemivaginal cavity was identified with a minimal connection to the left hemivagina which was closed. The pathological exam described a uterine duplication with inactive endometrium, a normal cervix and a right intramural myoma. There were also mesonephric remnants on the right vaginal wall.

In the postoperative period there was urine output through the vagina and a contrast computerized tomography was ordered under the suspicion of a vesicovaginal fistula. It described a complex genitourinary malformation in the pelvis with a urethral diverticulum that connected to the right hemivagina, which in turn was joined to the left hemivagina.

A high pressure retrograde cystography showed an abnormal connection with the right hemivagina through the bladder's base (Figure 2).

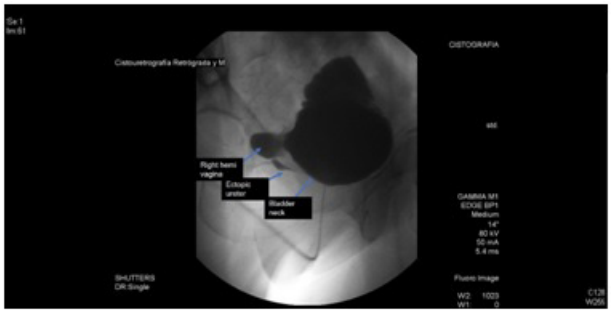

Figure 2: High pressure retrograde cystography

5. Outcome and Follow-Up

A cystotomy was performed revealing an abnormal ureter implantation in the bladder neck which communicated with the right hemivagina. This case was resolved with a resection and occlusion of the ectopic ostium of the ureter.

6. Discussion

Müllerian abnormalities have a wide variety of presentations at different points in life. They are present in up to 7% of women of childbearing age and have kidney malformations associated in 40% of cases [3].

HWW syndrome is a combination of a class I A vaginal abnormality and a class III double uterus, accompanied by a kidney agenesis. It is a rare syndrome with a complex diagnosis [5].

Cases of uterus didelphys with an obstructed hemivagina and kidney agenesia can present alongside other urinary tract malformations of the same side (OHVIRA) [6, 7], an embryological alteration occurring in the 8th week of gestation [8].

There are few cases published that have findings similar to ours, the first dating to 1995, published by Shibata et al [9].

The association of kidney agenesia and ectopical ureter is not uncommon. However, cases of an ectopic ureter that present with kidney agenesia and an abnormal communication between the bladder neck and an atresic vagina are extremely rare [10, 11].

It is possible that the origin of this malformation is related to the persistence of Gartner's duct vestiges, which could join the ectopic ureter and the vagina [13]. HWW syndrome is generally discovered in puberty with unspecific symptoms such as a progressive pelvic pain, dysmenorrhea, or a palpable pelvic mass due to hematocolpos or hematometra withheld by the obstructed vagina [2]. Our patient had both episodical pelvic pain and what appeared to be urinary incontinence, common in this type of patients [12].

Case presentation was particular in that there was hematuria from a young age during the menstruation and urinary incontinence in menopause, alongside occasional abdominal pain and urinary tract infections.

Ultrasound imaging is probably the first step towards a differential diagnosis [14], and whilst we did not use 3D ultrasound imaging it may provide highly dependable, objective and measurable information about uterine cervix, cavity and outer limits of the uterine body [15]. 2D ultrasound is also useful in diagnosis of ectopic ureter implantation, especially in an imperforate hemivagina [11].

MRI has also proved to be an excellent tool for the study of genitourinary alterations, and is even superior to ultrasound at identifying uterine contour, cavity shape and size, and tissue characteristics [15-17]. It has been shown to have a higher diagnostic sensitivity as compared to ultrasound of intravenous urography. In our case, MRI readily showed malformation characteristics however the communication with the bladder was not identified, and the high pressure cystography was necessary.

7. Learning Points/Take Home Messages

It is reasonable to conclude that a comprehensive gynecological examination, transvaginal ultrasound (especially 3D) and MRI should be first line of management when symptoms suggest a genital tract malformation. Invasive tests such as hysterscopies or laparoscopies should be reserved for surgical planning [2].

A urinary tract examination is also highly recommended in this patient population.

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