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Case Report

A rare urogenital malformation "Herlyn-Werner-Wunderlich syndrome" (Ohvira syndrome) discovered during an acute pelvic pain

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ABSTRACT

The Herlyn-Werner-Wunderlich syndrome (OHVIRA syndrome) is a rare urogenital malformation, characterized by a blind hemivagina, a didelphic uterus, and homolateral renal agenesis, it results from a defect in the development of the muller and Wolf ducts during the embryonic period, of unknown origin, the diagnosis is most often made at puberty during the first menstruation, sometimes late, particularly for non-obstructive forms: infertility or obstetrical complications, the diagnosis is based on pelvic ultrasound especially with 3D reconstructions and pelvic magnetic resonance imaging (MRI) which remains the standard gold for the detection of mullerian anomalies, laparoscopy can also play a double diagnostic and prognostic role in order to study the consequences of the blood reflection (hematosalpinx, inflammatory pelvis, endometriosis. In this work we report the case of an OHVIRA syndrome diagnosed late at the age of 36 years in a multiparous woman with acute pelvic pain simulating the picture of a torsion of the adnexa, we will discuss through this case the clinical, diagnostic and therapeutic aspects of this uterine malformation.

Keywords: OHVIRA syndrome, Obstructed hemivagina, Didelphic uterus, Hematocolpos, Renal agenesis, Vaginal septum

INTRODUCTION

The real incidence of uterine malformations is unknown, often recognized at puberty or later during menstrual or obstetrical manifestations, or within the framework of an infertility assessment, the didelphic uterus represents 5% of the malformations of the Müllerian ducts. Once diagnosed, it is necessary to look for a longitudinal vaginal septum which is associated with this type of malformation in 75%, Herlyn-Werner-Wunderlich (HWW) syndrome is a rare urogenital malformation characterized by a didelphic uterus, a blind hemivagina, a renal agenesis, the diagnosis is often made during the puberty period, sometimes late for the non-obstructive forms. The first line paraclinical examination in front of the suspicion of a uterine malformation is pelvic ultrasound especially with 3D reconstructions, pelvic magnetic resonance imaging (MRI) is the gold standard, laparoscopy has a double diagnostic and prognostic interest of this pathology, We report in this work the case of an OHVIRA syndrome diagnosed late in a multiparous woman with no particular obstetrical history, admitted to the emergency room for acute pelvic pain, we will discuss through this case the different clinical, paraclinical and therapeutic profiles of this pathology.

CASE REPORT

Mrs. YA, 36 years old, having medical history of a laparotomy 14 years ago, undocumented, married, gravida 3 para 3 (3 pregnancies carried to term with 3 live children), admitted to the emergency room for acute pelvic

pain, with right cruralgia, relieved by elevation of the legs. Clinical examination found a conscious patient, stable on both hemodynamic and respiratory levels, hyperalgesic 8/10 according to EVA, on speculum: a single cervical orifice of macroscopically normal aspect, healthy vaginal walls, on vaginal touch: mass at the level of the right anterolateral cul de sac with the presence of a separation furrow. The patient also reports the notion of an irregular cycle. Blood analysis showed negative beta human chorionic gonadotropin (bhCG) and C-reactive protein (CRP).

A pelvic endovaginal ultrasound was performed, visualization of two uterine calvities of which the right one is the site of a hematometria + a latero-uterine cystic mass finely echogenic (Figures 1 and 2).

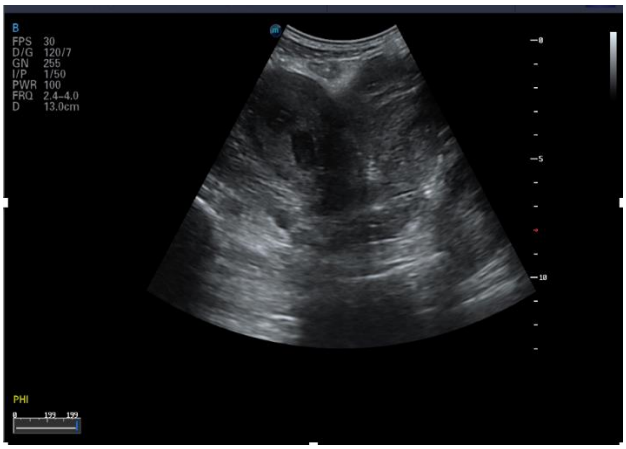

Figure 1: Ultrasound section showing a bicornuate uterus with hematometry in the right cavity.

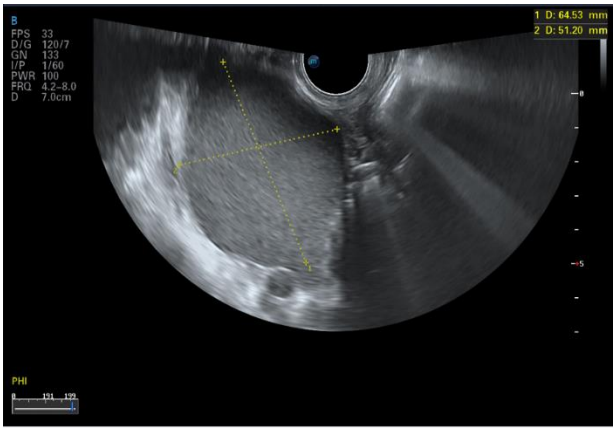

Figure 2: Finely echogenic right latero-uterine image (image was taken for an endometriotic or hematoma cyst).

As per computed tomography (CT) scan, uterine malformation of the didelphic uterus type, well-limited latero-uterine formation with regular contours measuring 72/55/52 mm most probably related to the right ovary, with an empty right renal pelvis (Figure 3).

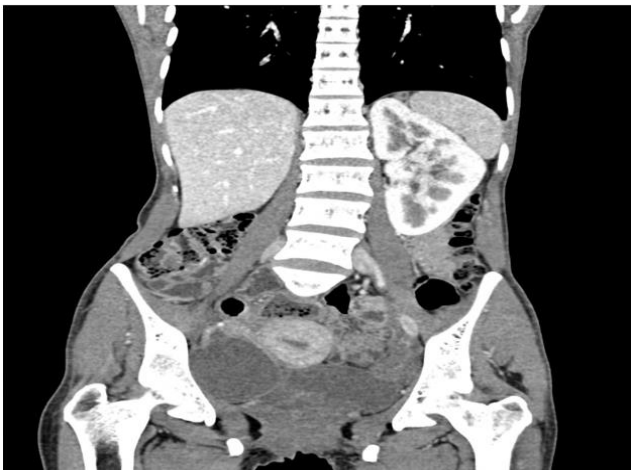

Figure 3: Axial scan section showing renal agenesis with two uterine structures and a right latero-uterine cystic mass.

The patient was made under analgesics that didn't calm the pain.

She underwent an exploratory laparoscopy which confirmed the presence of two uterine cavities with no visible adnexal mass. Combined with a vaginal touch, which confirmed the origin of the mass which was dependent on a right blind hemivagina, a low anterior detachment was carried out, then a puncture was performed with a cytopuncture needle bringing back more than 400 cc of chocolate brown liquid (Figures 4 and 5).

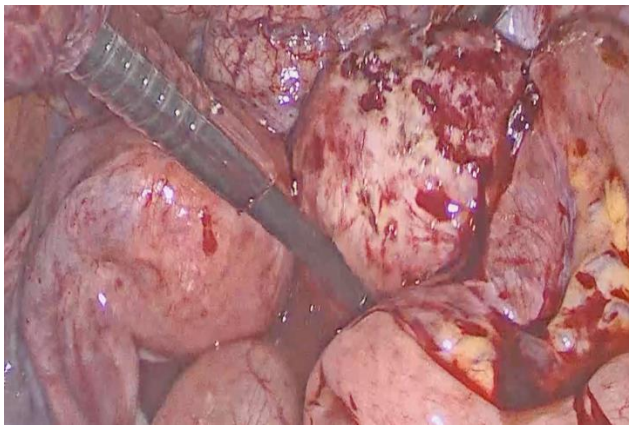

Figure 4: Laparoscopic view showing two uterine cavities.

Then an incision was made by the lower approach of the blind hemivagina and an excision of a right lateral vaginal collar was carried out, leaving an evacuation orifice. A drain was left in place which was attached to the lateral wall of the vagina and vulva with a number 1 vicryl absorbable thread. Patient made on antibiotics for 5 days. Clinically, the evolution was favorable with regression of the pain. An ultrasound was performed at day 8 showing a decrease in hematocolpos and hematometry (Figure 6).

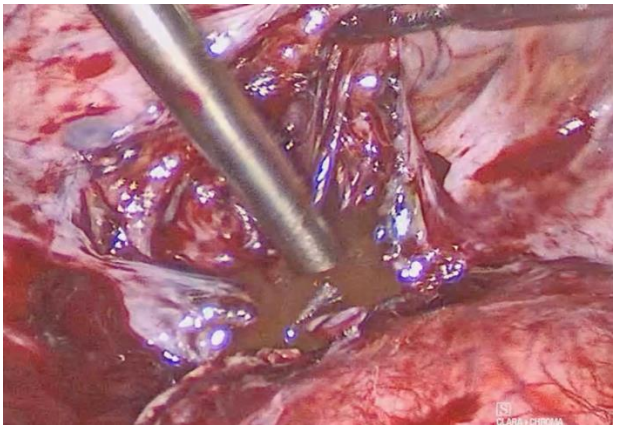

Figure 5: Puncture performed with a cytopuncture needle bringing back a chocolate brown liquid.

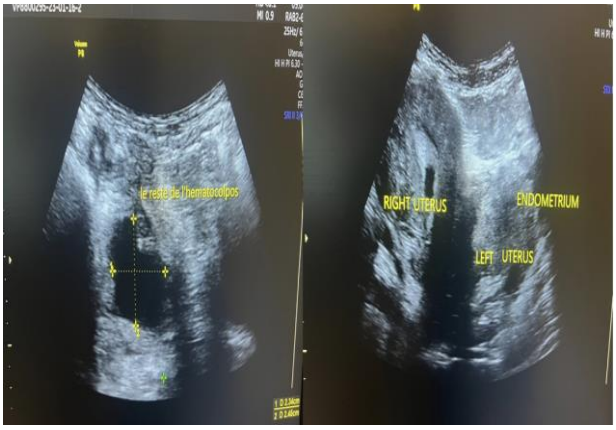

Figure 6: Ultrasound showing regression of hematocolpos and right hematometry.

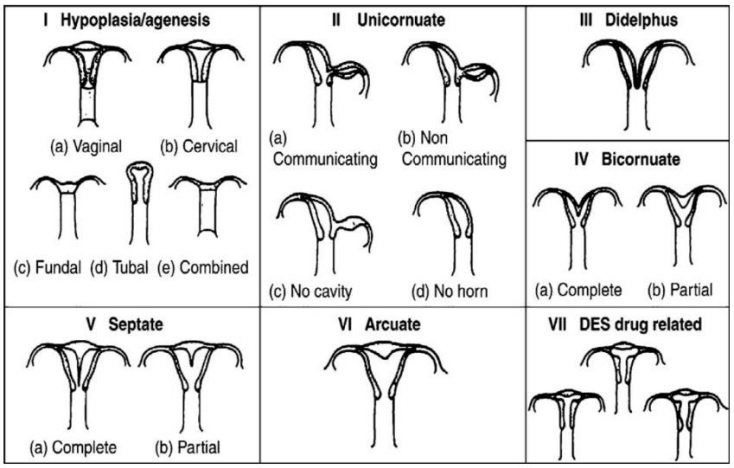

Figure 7: Classification of uterine malformations according to the American Fertility Society.

ESHRE/ESGE classification
Female genital tract anomalies

	Uterine anomaly		Cervical/vaginal anomaly	
	Main class	*Sub-class*	*Co-existent class*	
U0	Normal uterus		C0	*Normal cervix*
U1	Dysmorphic uterus	**a.** T-shaped **b.** Infantilis **c.** Others	C1	*Septate cervix*
U2	Septate uterus	**a.** Partial **b.** Complete	C2	*Double 'normal' cervix*
U3	Bicorporeal uterus	**a.** Partial **b.** Complete **c.** Bicorporeal septate	C3	*Unilateral cervical aplasia*
U4	Hemi-uterus	**a.** With rudimentary cavity (communicating or not horn) **b.** Without rudimentary cavity (horn without cavity/no horn)	C4	*Cervical aplasia*
U5	Aplastic	**a.** With rudimentary cavity (bi- or unilateral horn) **b.** Without rudimentary cavity (bi- or unilateral uterine remnants/aplasia)	V0	*Normal vagina*
U6	Unclassified malformations		V1	*Longitudinal non-obstructing vaginal septum*
			V2	*Longitudinal obstructing vaginal septum*
			V3	*Transverse vaginal septum and/or imperforate hymen*
			V4	*Vaginal aplasia*
U			*C*	*V*

Associated anomalies of non-Müllerian origin:

Figure 8: 2013 ESHRE/ESGE classification of uterine malformations.

DISCUSSION

Uterine malformations result from a defect in the development, division or regression of the Müllerian ducts during the embryonic period. Depending on the mechanism and the level of damage, there are several types of anomalies summarized according to different classifications.

The first is the classification of the American Fertility Society (currently American Reproductive Society) which is a modification of the original work of Buttram and Gibbon in 1979 which is based essentially on the anatomy of the female reproductive system, it is a simple and clear system, which has been successfully adopted as the main system of classification for almost two decades (Figure 7).[1,2] The disadvantage of this system is that some types of malformations are not included such as: didelphic uterus with obstructed vaginal septum (our case), bicornuate uterus with cervical or vaginal aplasia.[4-6]

Another classification based on embryology was created by Acien et al and helps to better understand the origin of disorders of the different elements of the urogenital tract but this system is not very simple or user-friendly.[4]

Hence, the development of the new European Society of Human Reproduction and Embryology (ESHRE)/Society for Gynaecological Endoscopy (ESGE) anatomical classification of 2013.[3] The most recent one which has mainly clinical and anatomical orientation thus more practical, uterine anatomy is the basis of the new system, cervical and vaginal anomalies are grouped into independent coexisting subclasses. It seems that the new system meets the needs and expectations of a large group of experts in this field. Clinicians can use Figure 2 for an easy and accurate description of the anomalies to classify or draw the pattern of the malformation (Figure 8).[3]

The real incidence of Müllerian duct anomalies is unknown, often recognized at puberty or later during menstrual or obstetrical manifestations, or in the context of an infertility assessment, the normal development of the EGDs may also be the cause of a delay in diagnosis. Some authors have demonstrated an equally high incidence in fertile women ranging from 1/200 to 1/600, the didelphic uterus represents 5% of Müllerian duct malformations.[10,11]

Ohvira syndrome (Herlyn-Werner-Wunderlich syndrome) is a rare urogenital malformation, characterized in its classical form by: a didelphic uterus, a blind obstructed hemivagina and associated ipsilateral renal agenesis. It results from an embryological arrest of the "paramesonephric" (the duct that gives rise to the OGI) and mesonephric (the duct that gives rise to the kidneys and ureters) at 8 weeks of gestation. The malformations of the urinary tract appear at the same time when Wolf's (mesonephric) and Müller's (paramesonephric) ducts are topographically close; renal agenesis, reported in several series is almost constant.[9] The exact etiology of this malformation is unknown, however, genetic, environmental and endocrine factors may be incriminated.[8] According to the different classifications, this anomaly is classified as type III according to the American Fertility Society and U3bC2V2 according to the new ESHRE classification, there are other variants of this pathology: the blind hemivagina with renal agenesis can be associated with a septate or total bicornuate uterus instead of a didelphic uterus (U2b according to ESHRE and Iva or Va according to AFSM).[7] According to the retrospective study of Zhu et al, the OHVIRA syndrome is classified into two types according to the permeability of the blind hemivagina.[12]

Class 1 complete obstruction of the hemivagina (our case): in this case the clinical manifestation is generally early, as soon as the first menstruations start, pelvic pain, abdominal impaction by hematocolpos which may be complicated by peritonitis, or urinary retention due to the mass effect of the hematocolpos (our patient has a median laparotomy scar most probably related to a previous peritonitis).[12]

Class 2 of late discovery most often due to the fact that the second uterus is drained by a semipermeable hemivagina (class 2.1) or by a communication between the two cervixes (class 2.2) the clinical picture is often that of a recurrent genital infection or an upper genital infection.

The diagnosis is rarely made in the infantile period, cases of muco or hydrocolpos have been reported.[13] For some women the discovery was late in the course of a clear cell carcinoma of the vagina or a cervical ADK on the obstructed side.[14] In case of presence of the above described symptoms or in case of doubt, the diagnostic investigation includes: a detailed questioning in order to determine the character of the pain, its chronology, a physical examination preferably under sedation which is recommended by the American Academy of Pediatrics.[15,16] For complementary examinations, ultrasound is the first-line examination that has a sensitivity of 93% and a specificity of 100% for the detection of uterine malformations, mainly with 3D reconstructions (in our case, the ultrasound was performed in an emergency context and confirmed the presence of a bicornuate uterus with a hematoma on the right side, but given the size and structure of the hematoma, it was taken to be a latero-uterine mass, which was confirmed by the CT scan performed as an emergency examination).[7] We can deduce that the urgent context of the patient made the diagnosis more difficult. Pelvic MRI is the gold standard for the study of the female genital tract, and is the reference examination for confirming the diagnosis; its accuracy is 100% for the detection of abnormalities of the mullerian duct.[17] Hysterosalpingography is only of interest for the detection of a communication between the 2 uterine cervixes (class 2.2) or when the diagnosis is made late, a fistula may form, and only HSG can reveal it.[19] Laparoscopy can help or confirm the diagnosis (in our case the diagnosis was made in the operating theatre), in

addition to its diagnostic interest, laparoscopy also allows the consequences of the blood reflux to be assessed (prognosis on female fertility), which mainly comes down to tubo-ovarian adhesions, hydro or even pyosalpinx, and late peritoneal endometriosis.[19] The therapeutic strategy for this syndrome consists of emptying the hematocolpos and ensuring continuous drainage of the uterine cavity by performing a wide resection of the vaginal septum in order to avoid fibrosis and secondary stenosis, only class 1. 2 (cervico-vaginal atresia with complete obstruction) according to Zhu et al requires hysterectomy of the undrained uterus because the obstacle is high up, non-contact vaginoscopy is a curative technique where the introduction of the hysteroscope can be done without using instruments that can cause hymenal damage.[17] In the case of postoperative pain, using an L-shaped hysteroscopic electrode, the vaginal septum is incised and then a Foley catheter inflated with 50 to 80 ml of SS is placed in the obstructed hemivagina, this technique allows good surgical results to be obtained.[21] Fertility is preserved in the majority of cases, but there are risks of miscarriage, ectopic pregnancies, threats of premature delivery due to a reduction in the lumen of the uterine cavity, and other obstetrical complications, in particular dystocic presentations, which are the cause of increased rates of caesarean section and intraoperative diagnosis of this malformation.[20] This is an exceptional case, taking into consideration the history of the patient, who is a multiparous woman without any notion of abortions or dystoic delivery, the late age of the diagnosis which can be explained by the formation of a fistula between the 2 cavities, hence the delay in the constitution of the hematocolpos.

CONCLUSION

Herlyn-Werner-Wunderlich syndrome (OHVIRA syndrome) is a rare urogenital malformation of unknown origin. Diagnosis is often early at the first menstrual period, sometimes the diagnosis is late for other semi or non-obstructive variants. The treatment must be early to avoid the physical complications and the psychic impact of this pathology, it is based on a continuous drainage of the uterus concerned by the obstruction. The fertility profile in the majority of cases is preserved apart from obstetrical complications.

With the fetal morphological echographic evolution, it seems necessary to think of this syndrome during a unilateral renal agenesis in a female fetus.

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