

Chronic pelvic pain caused by Herlyn-Werner-Wunderlich Syndrome: Case report and discussion

Fatma Zahra Affes¹, Hatem Frikha², Sana Menjli², Abir Karoui², Sami Mahjoub¹, Mohamed Badis Chanoufi², and Hassine Saber Abouda²

¹Centre de Maternité et de Neonatologie

²Affiliation not available

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Affes Fatma Zahra, Resident in obstetrics and gynaecology¹, Frikha Hatem, Resident in obstetrics and gynaecology¹, Menjli Sana, Physician assistant in obstetrics and gynaecology¹, Karoui Abir, Physician assistant in obstetrics and gynaecology¹, Mahjoub Sami, Professor in obstetrics and gynaecology¹, Chanoufi Mohamed Badis, Professor in obstetrics and gynaecology¹, Abouda Hassine Saber, Associate Professor in obstetrics and gynaecology¹

Author's affiliations:

¹Département of Gynaecology « C » Centre de maternité et de néonatalogie de Tunis

Name and contact details:

Affes Fatma Zahra

Email: fatmazahra310@gmail.com

Phone : +216 21444833

Address : Centre de maternité et de néonatalogie de Tunis 1007 La Rabta Tunis, Tunisia.

Introduction:

HWW Syndrome also known as OHVIRA (obstructed hemivagina and ipsilateral renal agenesis) is a rare genitourinary anomaly that occurs in females. It is classified as class III Mullerian dysgenesis.(1) This condition involves uterus didelphys and blind hemivagina associated with ipsilateral renal agenesis. It represents 5% of total Mullerian dysgenesis(2). The onset of symptoms usually begins after the menarche. Abdominal and pelvic pain are the most common symptoms. The diagnosis is difficult because of the infrequency of the syndrome and requires precise imaging to evaluate uterine cervical and vaginal morphology. Early detection and treatment of the syndrome can relieve the symptoms, improve the quality of life and prevent serious complications.(3) We present here a case of investigation and management this condition with pelvic pain in young female.

Presentation of case:

A 15 year-old female was referred to our university hospital with complaint of pain in the lower abdomen on the right side. She reported that she has the same chronic pain since her first menses at the age of twelve. She described her pain as mild in the first episodes then increasing gradually with each subsequent menstrual

cycle. Her menstrual cycles were irregular with moderate bleeding. She haven't had any sexual intercourse before.

Physical examination revealed a tender lump in the pelvic region on the right side of midline. Secondary sex growth was normal but the vaginal examination was not done.

Ultrasound examination of the pelvis was first performed exhibiting uterus didelphys. The right uterine cavity and cervical canal showed a collection suggestive of hematometra and hematocervix, the vagina was also dilated containing hypoechoic collection. The left uterine cavity and cervix were normal with homogenous endometrium. Both ovaries were normal.

We performed an ultrasound of the abdomen searching urinary tract anomaly, and it revealed the agenesis of the right kidney with mild compensatory hypertrophy of the left kidney.

MRI of the abdomen and the pelvis confirmed the findings of the ultrasound examination: The right kidney was absent in the right renal fossa with mild compensatory hypertrophy of the left kidney. Normal left uterus with its own corpus and cervix, the right uterus cavity was stretched and contained hypodense material and was connected to a cystic mass by its cervical canal obstructing the vagina.

A diagnosis of uterus didelphys with right hematometra, hematocervix and hematoocolpos with right renal agenesis suggestive of the Herlyn-Werner-Wanderlich syndrome was made.

Surgical intervention was planned. We first performed vaginoscopy under general anesthesia that showed a small vagina, normal cervix and uterus with one ostium. Septoplasty was prepared but was not possible because of the small size of the vagina, the septum was thick and profound. We decided to puncture the hematoocolpos to evacuate the blood and relieve the pain. The puncture was difficult because of the thickness of the blood clots so infusion of saline serum was first done then the puncture was easier, and 300 cc of blood was evacuated. The patient did well postoperatively and we decided then to inject medroxyprogesterone acetate every three months to prevent the accumulation of the blood in the right uterus and vagina. Further evaluation showed an asymptomatic patient, and ultrasound did not show any blood accumulation in the uterus or the blind vagina.

Discussion:

Herlyn Werner Wanderlich syndrome is a rare congenital defect in the development of genitourinary tract. It is characterized by uterus didelphys with obstructed hemivagina due to lateral non-fusion of mullerian duct with asymmetric obstruction. It is associated to renal agenesis ipsilateral to the obstructed vagina.(4)

This syndrome was described for the first time in 1971 by Herlyn and Werner. Then in 1976 renal aplasia with bicornuate uterus and simple vagina in the presence of an isolated hematocervix was described by Wunderlich.(5)

This kind of anomaly was classified to two types: type I a completely obstructed vagina and type II an incompletely obstructed vagina.(3)

The clinical presentation depends on the type of obstruction, it usually consists on severe dysmenorrhea starting after the menarche. The pain typically increases due to the volume of the hematometrocolpos when the vagina is completely obstructed however incomplete obstruction may be the origin of intermittent and mild symptoms. (6)

Besides hematometra, complete obstruction causes hematosalpinx as well as bleeding in the periadnexal and peritoneal space. Endometriosis may also occur as a consequence of the blood reflux in the peritoneal cavity and progress causing pelvic adhesions and infertility.(3)

Few cases of acute abdominal pain, abnormal vaginal discharge or acute retention of urine has been reported.(7) Rarely, spontaneous perforation of the obstructed vagina may occur leading to pyocolpos and pelvic inflammatory disease and also septic shock.(8)

The diagnosis of the Mullarian duct anomalies can be done using HSG, US, 3D US and MRI.(9) Conventional ultrasonography can detect the hematocolpos and help to identify the type of the Mullarian duct anomalies because it displays the features of the external uterine contour.(10) MRI is the gold standard for diagnosis with higher sensitivity in detecting the uterine morphology and the continuity of the vagina, thickness and localization of the vaginal septum.(7) However, the agreement between the 3D US and MRI has been also shown in several studies in the evaluation of this type of anomalies.(11)

HSG was used before the development of MRI and US, its ability is limited because it is unable to display the external contour of the uterus. (9)

Laparoscopy is not necessary nowadays for the diagnosis thanks to the advanced imaging technologies and it is reserved only if the diagnosis isn't clear after imaging. (12)

Currently, the main treatment of the OHVIRA syndrome is full resection of the vaginal septum in order to achieve the continuity of vagina.(13) Vaginoplasty can be performed in single stage where the resection of the vaginal septum is complete or in two stages: draining the hematocolpos first then resection of the septum.(8) Infection, anatomic distortion and incomplete previous resection were the main indication of two stages vaginoplasty.(14) Laparoscopy doesn't seem to be required for the surgical management in most cases.(14)

However, Fauchin and al reported a case of hemiuterus with absent cervix where the vaginoplasty was not possible, the treatment was a laparoscopic hemi-hysterectomy with salpingectomy.(15) Rarely, hemi hysterectomy may be also considered in case of recurrent stenosis.(7)

Hysteroscopic resection of the septum under transabdominal guidance has also been reported in order to preserve the integrity of the hymen.(16)

When postponement of surgery is needed for young adolescents, amenorrhea can be maintained by the gonadotrophin-releasing hormone analogues.(13)

Conclusion:

OHVIRA syndrome is the most common obstructive Mullarian duct anomaly diagnosed after menarche in young adolescent. It is responsible of cyclic pain. It is usually treated with vaginoplasty and drainage of the hematocolpos. In case of recurrent stenosis or difficult septum resection, unilateral hysterectomy is the treatment of choice.

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Contribution to authorship:

Dr Affes Fatma Zahra: Initial diagnostics and treatment of the patient and case report manuscript.

Dr Frikha Hatem: Initial diagnostics and treatment of the patient and case report manuscript.

Dr Menjli Sana: Initial diagnostics and treatment of the patient.

Dr Karoui Abir: Initial diagnostics and treatment of the patient.

Dr Chanoufi Mohamed Badis: Initial diagnostics and treatment of the patient.

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Details of Ethics Approval

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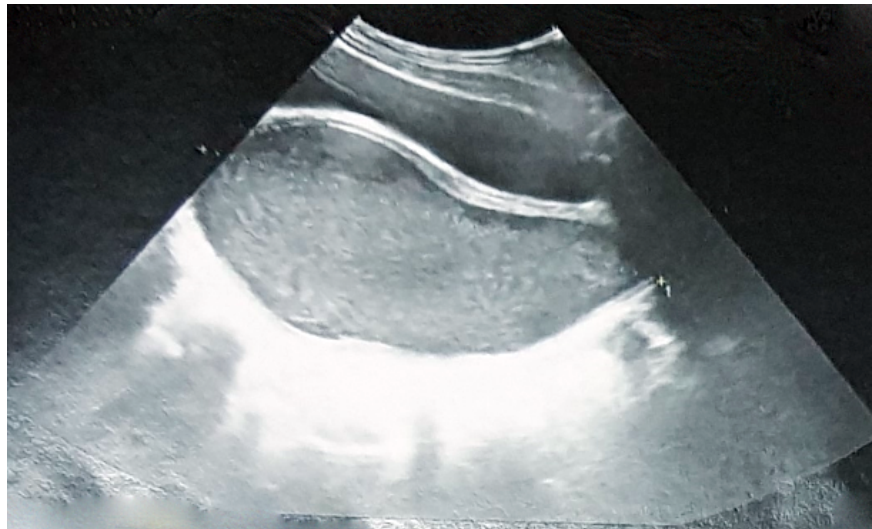
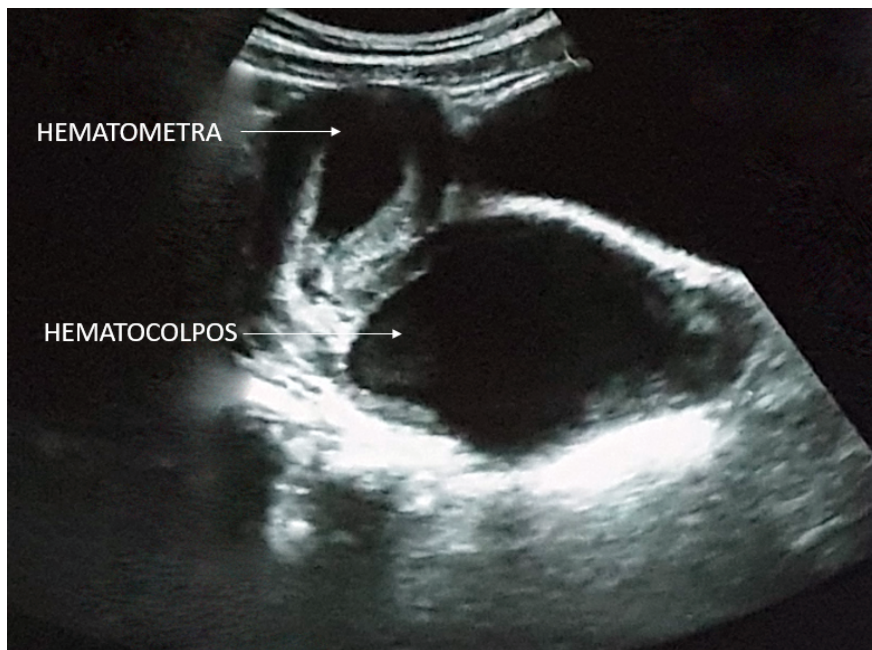
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Consent for publication:

A consent was signed by the parents of the patient.

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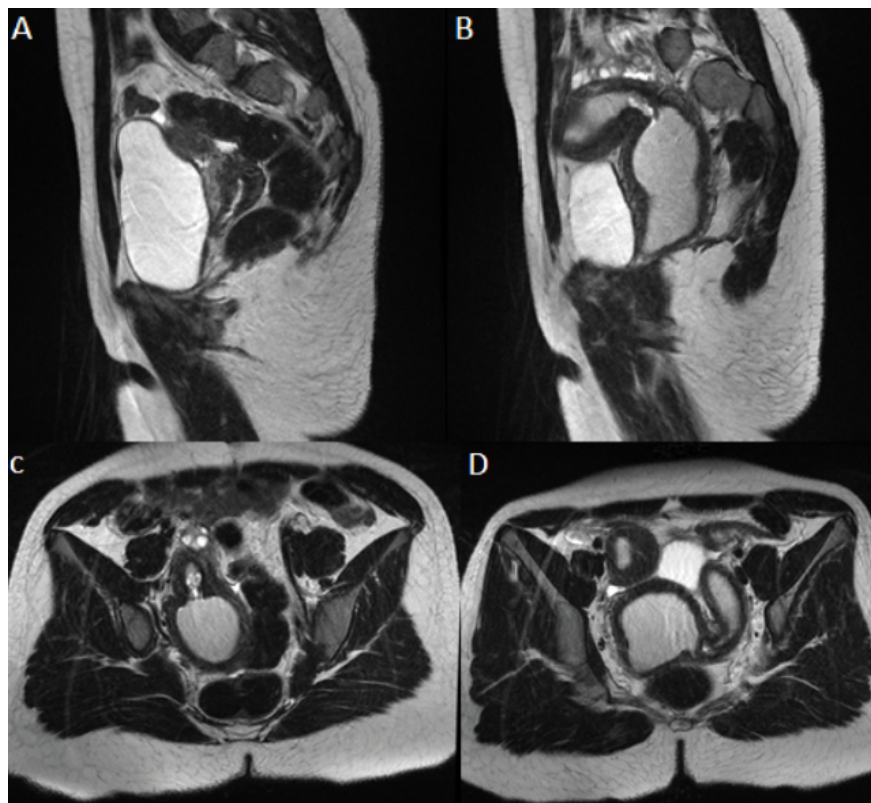


Figure caption list:

Figure1: Pelvic ultrasound image showing hematocolpos (echogenic contents).

Figure 2: A transabdominal ultrasound image showing the right dilated uterus with hematometra and the hematocolpos.

Figure 3: **A**)Sagittal magnetic resonance image (MRI) showing the normal uterine left cavity.**B**)Sagittal image of the same patient showing the dilated right uterine cavity and the hematocolpos.**C**)Coronal MRI showing the right cervix and the hematocolpos **D**)Coronal image showing the right uterine horn and the normal left uterine cavity with hematometra.