MRI Diagnosis Of A Case Of Herlyn-Werner-Wunderlich Syndrom

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Citation

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Abstract

The Herlyn-Werner-Wunderlich (HWW) syndrome is a very rare congenital anomaly of the female urogenital tract that associates Mullerian duct anomalies (MDA) with mesonephric duct anomalies. This syndrome is characterized by the presence of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis. In literature HWW syndrome is also known as OHVIRA (obstructed hemivagina and ipsilateral renal anomaly) syndrom. Generally women affected by HWW syndrome are asymptomatic until puberty and have non-specific symptoms after menarche such as pelvic pain, dysmenorrhea, and palpable mass due to hematocolpos or hematometra. However, being an uncommon syndrome, HWW is often not considered a diagnostic possibility. Despite being a rare condition, the complications of this syndrome can be severe and women's fertility may be at risk. Therefore it is necessary to have a very high suspicion to be diagnosed and it is essential to combine all clinical and imaging findings. We report a case of a 15-years-old girl who presented with cycled pelvic pain, describing MRI findings, essential for the diagnosis. MRI is at present the best imaging tool to make diagnosis giving full information to surgery.

INTRODUCTION

The Herlyn-Werner-Wunderlich (HWW) syndrome is a very rare congenital anomaly of the female urogenital tract that associates Mullerian duct anomalies (MDA) with mesonephric duct anomalies. This syndrome is characterized by the presence of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis[1]. In the literature, HWW syndrome is also known as OHVIRA (obstructed hemivagina and ipsilateral renal anomaly) syndrom[2]. However, this definition is broader, because it includes other urinary tract anomalies such as double collecting system, renal duplication, and horseshoe kidney other than renal agenesis[3].

Generally women affected by HWW syndrome are asymptomatic until puberty and have non-specific symptoms after menarche such as pelvic pain, dysmenorrhea, and palpable mass due to hematocolpos or hematometra[1]. These most common findings arise generally from 2 to 12 months after menarche[4]. The diagnosis is also possible in neonates or in adults.[5]

Other common symptoms include intermenstrual bleeding, mucopurulent vaginal discharge and fever. Early diagnosis is a key factor to prevent complications[6], distinct in acute complications, such as pyohematocolpos, pyosalpinx, and pelviperitonitis, and long-term complications such as endometriosis, pelvic adhesions and increased risk of abortion or infertility[7]. However, being an uncommon syndrome, HWW is often not considered a diagnostic possibility [8]. Also, there can be syndrome variants whose knowledge is important for proper diagnosis and therapeutic management decisions [9]. Treatment is vaginal septectomy with drainage of hematocolpos[10,11]. After surgery around 80% of patients are able to conceive[12]. Despite being a rare condition, the complications of this syndrome can be severe and women's fertility may be at risk. Therefore, it is necessary to have a very high suspicion to be diagnosed and it is essential to combine all clinical and imaging findings.

We describe the case of a 15-years-old girl who presented with cycled pelvic pain.

CASE REPORT

A 15 years-old girl presented to the Gynecological Department of our Hospital with a history of worsening pelvic pain in conjunction with the menstrual cycle, which occurred every 28 days and lasted 3-4 days. She was not

sexually active and had no surgical or medical history, especially with regards to contraceptive pills and hormonal therapy. Her psycho-physical development was normal and within her family no congenital pathologies were reported.

The physical examination found normo-conformed external genitals and no palpable masses.

Because of the pelvic cyclic premenstrual pain, the clinical question arose as suspicion of endometriotic cyst suggesting a sonographic examination. A trans-abdominal ultrasound was performed. It showed an enlarged uterus with dysmorphic morphology and an inhomogeneous hematocolpos.

Due to the complexity of the case, pelvic RMI was performed with a 1.5 T magnet (Magnetom Aera, Siemens, Germany) and body phased-array coil. The examination was carried out by multiplanar sequences, DWI, T2 and T1 weighted, before and after intravenous administration of paramagnetic contrast medium (gadoteric acid, Dotagraf, Germany, 1.5 ml/kg).

The pelvic MRI demonstrated an uterus didelphys, urogenital anomaly consisting in two separate uteri with two separate cervices. The uterus didelphys was displaced cranially by a voluminous grossly rounded formation with clear limits, of 11x10x7 cm, characterized by high signal in T1-weighted sequences and moderately hyperintense compared to miometrium in T2-weighted imaging, as for the blood component; the mass communicated with the left sinus endometrial canal which also presented similar blood aspects in context and was compatible with hematocolpos, due to the absence of communication of the left uterus with the homolateral vagina, owing to the presence of a transverse septum.

The contralateral vagina was patent and laterally dislocated by hematocolpos. MRI showed an enlarged and globular right annex, with small follicular cysts and fluid-cystic formation, the latter with slender internal burials. Left annex had a regular appearance.

There was a certain amount of free liquid effusion in the Douglas pouch and between the intestinal loops. No endometriosic cysts were found. MRI examination was extended to the upper abdomen and displayed left kidney agenesis with compensatory hypertrophy of the right kidney.

In its specificity, the anomaly consisted of an uterus duplex bicornis septus (figures 1 and 2), a patent right hemivagina, a blind left hemivagina, a presence of hematocolpos (figures 3, 4a - 4b) and a left kidney agenesis with compensatory hypertrophy of the right kidney (figure 5).

The patient underwent surgery for vaginal septotomy and drainage of hematocolpos. Follow up is scheduled every six months.

Figure 1

Axial T2 weighted image displays uterus duplex bicornis septus.

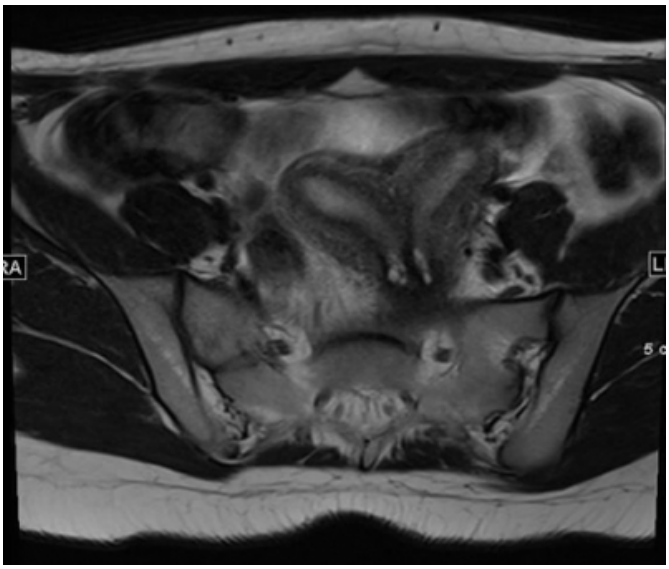


Figure 2

Axial T1 weighted image shows uterus duplex bicornis and blood content in the left uterine cavity.

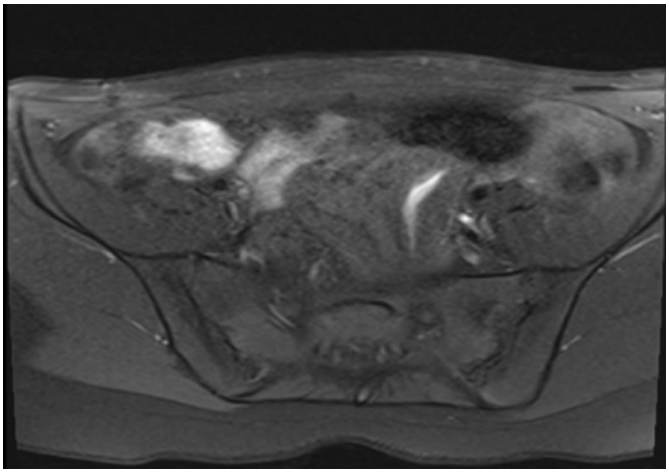


Figure 3

Sagittal T1 fat sat demonstrates blood signal in the uterine cavity connected with the hematocolpos.

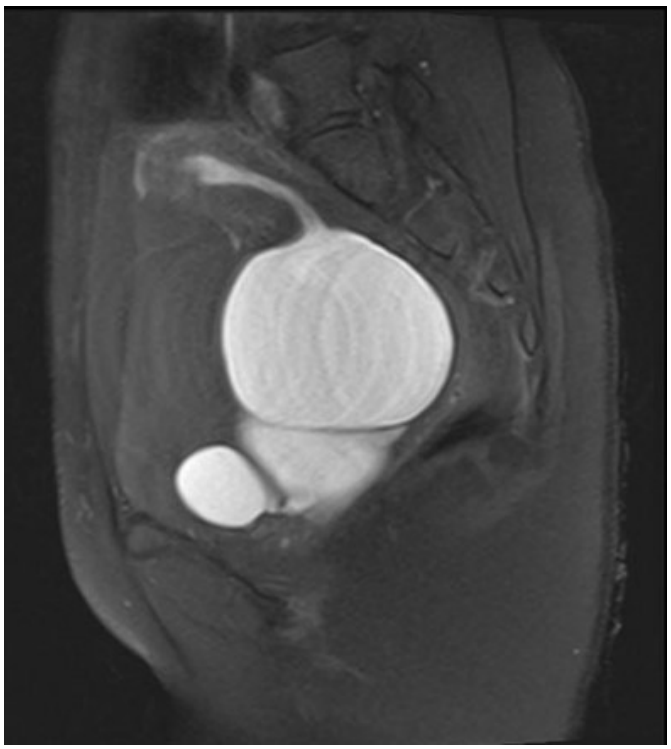


Figure 4

Axial T2 weighted (4a) and Axial T1 fat sat (4b) images exhibit the hematocolpos. The bladder is dislocated anteriorly and small amount of free fluid is placed posterior to hematocolpos.

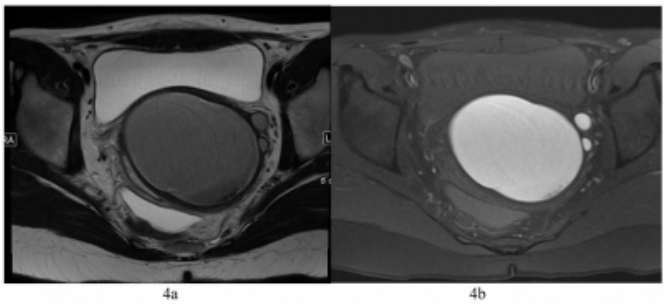
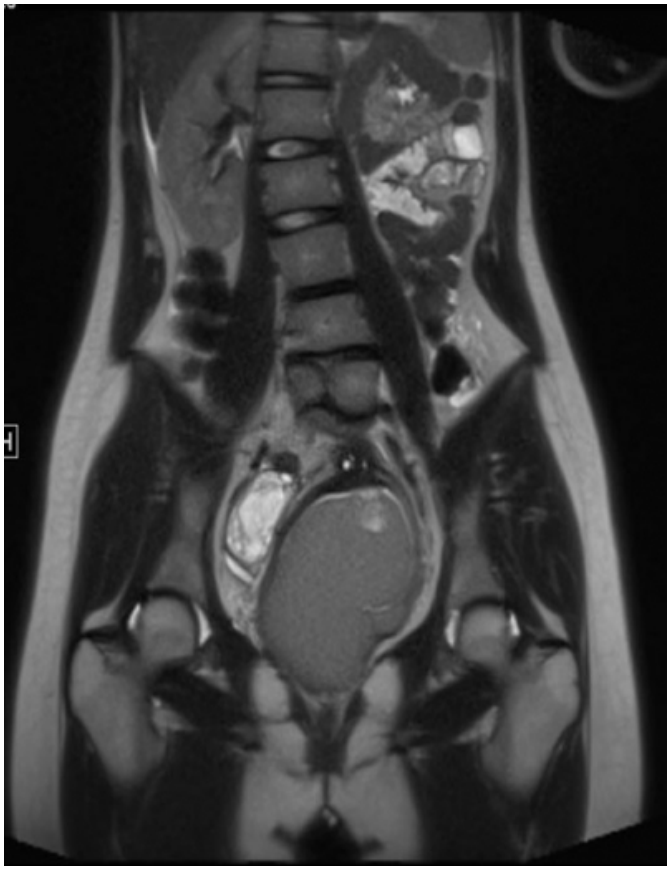


Figure 5

Coronal T2 weighted image displays hematocolpos, follicoles in the right ovary and left renal agenesia. Collateraly L5 soma has “butterfly morphology”.



DISCUSSION

Herlyn-Werner-Wunderlich is a rare congenital syndrome described for the first time in 1922 and characterized by uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis.[13] The incidence of this uncommon anomaly is very small and in the literature the syndrome often appears as a single case report or as a small series[14].

Clinical features are pelvic and abdominal pain, as happened in our case. The syndrome is usually diagnosed following menarche. Due to its rarity, there can be a delay in diagnosis which increases the risk of complications such as endometriosis and infertility[5]. In all patients with Mullerian anomalies, it is compulsory to evaluate the urinary tract because of the embryological relationship between renal and mullerian anomalies. At the same time female neonates with renal agenesis should be checked for genital malformations [15].

Zhu L. et al., suggested gynecologists world-wide to rely on a new classification of this syndrome because the clinical

characteristics differ significantly between the completely and incompletely obstructed vaginal septum. The new classification, displayed in figure 6, is based on the presence of a complete hemivaginal obstruction with or without cervical aplasia (class 1) and incompletely obstructed hemivagina communicating either between the vaginas or uteri (class 2). The mean age at onset of symptoms (years) was much younger in patients with a complete hemivagina obstruction (12.86 ± 1.84) versus incomplete obstruction (21.68 ± 7.43) [14,16].

Pelvic endometriosis is more common in young patients with obstructive genital abnormalities, in particular one-fifth of patients with HWW are susceptible to this condition. [16] According to the study of Tong et al. based on population of 70 patients, the lateral obstruction of the hemivagina and the renal agenesis is more frequent in the right side (60%). [12] In our case it was present in the left side.

Ultrasound imaging, computed tomography, and most importantly, magnetic resonance imaging (MRI) provide essential information for diagnosis. In particular, T2 sequences give the anatomo-morphological informations indispensable for the correct diagnosis, avoiding surgical interventions/laparoscopy, which were necessary in the past to diagnose this rare anomaly [17].

The standard treatment of HWW syndrome is surgical.

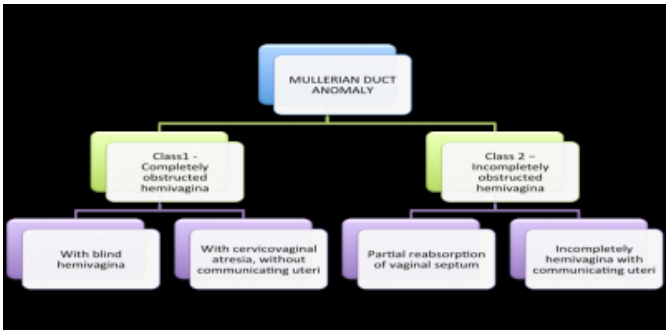
Despite the Mullerian duct anomaly, fertility in patients with HWW syndrome is preserved; after vaginal septal surgery around 80% of patients are able to conceive. In women who conceived before undergoing treatment, pregnancy occurred in the uterus ipsilateral to the hemivagina septum in 36,5% of cases [12]. A membranous duct has built between the uterus and the vagina, the patient is followed up with six-monthly checks to evaluate the patency of the duct.

In the future, with the consent of the patient, a resection of the dividing septum between the two uteri will be performed to reconstruct a single cavity and allow a safer pregnancy.

It is desirable for the patient to become pregnant in her 20's.

Figure 6

Classification of HWW syndrome by Zhu L. et al.[16]



CONCLUSIONS

The HWW syndrome is characterized by the triad of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis. It causes abdominal pain in young females. Early diagnosis and treatment are essential to relieve symptoms, preserve fertility and prevent acute and long-term complications.

Full attention from pediatricians and gynecologists is required to make a differential prognosis in female patients suffering from lower abdominal pain and, after menarche, dysmenorrhea.

MRI is at present the best imaging tool to provide essential information for diagnosis and surgery.

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