A case of Herlyn-Werner- Wunderlic syndrome with recurrent lower abdominal pain

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Introduction

Congenital anomalies of the Mullerian duct system can result in various urogenital anomalies. Herlyn - Werner- Wunderlich (HWW) syndrome is a rare anomaly characterized by uterus didelphys with blind hemivagina and ipsilateral renal agenesis (1).

Mullerian duct anomalies have an incidence of 2–3%. While obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) also known as Herlyn Werner Wunderlich syndrome, constitutes 0.16–10% of these Mullerian duct anomalies (11).

This syndrome was described for the first time in 1922, and was suspected in a young woman with regular menstruation and gradually increasing pelvic pain and a pelvic mass formation, usually noticed after menarche (2).

This anomaly is generally observed in post-menarche adolescents and young women presenting as irregular menstrual cycle, dysmenorrhea, abdominal pain, and pelvic mass (3,4). It may also present with urgency, frequency and vaginal discharge (12).

It is really difficult to achieve an accurate diagnosis because menstruation is often regular and when the patient complains of cyclic dysmenorrhea, they are usually given anti-inflammatory drugs and oral-contraceptives, thus causing a delay in the diagnosis as they reduce or eliminate menstrual blood. Also it may be attributed to lack of understanding of this condition by radiologists, gynecologists, and pediatricians.This may lead to pelvic adhesions, endometriosis or infertility.

The experience with HWW syndrome was definitely limited, consisting of case reports. It was reported that laparoscopy is needed for accurate diagnosis and treatment (5).

For diagnosis, ultra sonography usually gives an accurate picture, by showing uterovaginal duplication, hematocolpos or hematometrocolpos along with the absence of ipsilateral kidney (4). However MRI has been considered as the imaging modality of choice by various authors (6).

Case history

A 15-year-old girl presented with chief complaint of lower abdominal pain during menses for last 6 months. Her menstrual history suggested no abnormality except dysmenorrhea, she denied any past medical or surgical history. One month ago she presented to the emergency room complaining of lower abdominal pain mainly on the right iliac fossa with history of anorexia, nausea and vomiting.

On examination secondary sexual characters were normal for age. Abdominal examination showed localized tenderness on the right iliac fossa; rebound tenderness was positive. She was suspected to have acute appendicitis versus ovarian torsion. Appendectomy was performed and the histopathology came back as normal appendix, the ovaries were inspected intra-operatively; they were reported as normal.

Two weeks later she has another attack of lower abdominal pain. Ultrasound examination revealed ovarian cyst (as reported by the radiologist) and absent right kidney. She was referred to the gynecologist for further assessment.

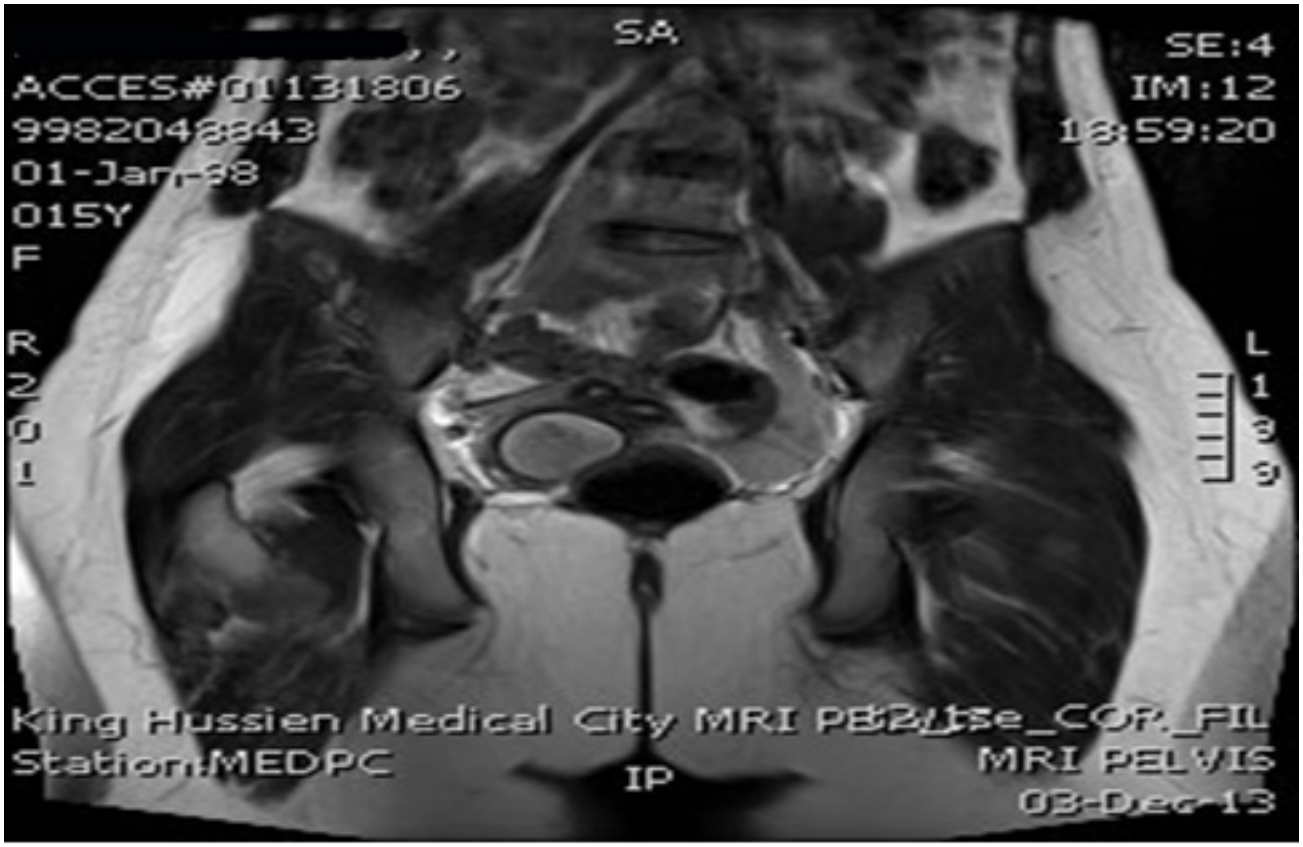
The patient was a virgin, thus vaginal examination was not possible.

Ultrasonography was done which revealed double uterus with hematocolpos and absent right kidney with marked dilatation of the lower part of the left ureter, while the left kidney was enlarged.

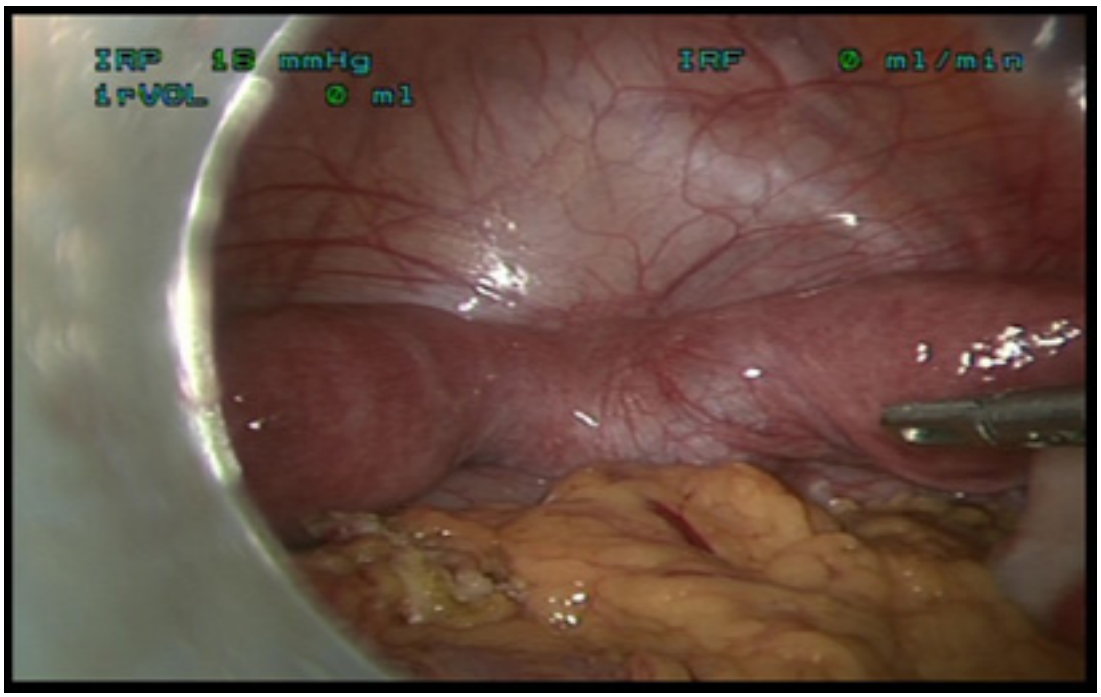
Congenital anomalies of the Mullerian duct system can result in various urogenital anomalies and Herlyn- Werner-Wunderlich syndrome is a rare anomaly characterized by uterus didelphys and blind hemivagina associated with ipsilateral renal agenesis.

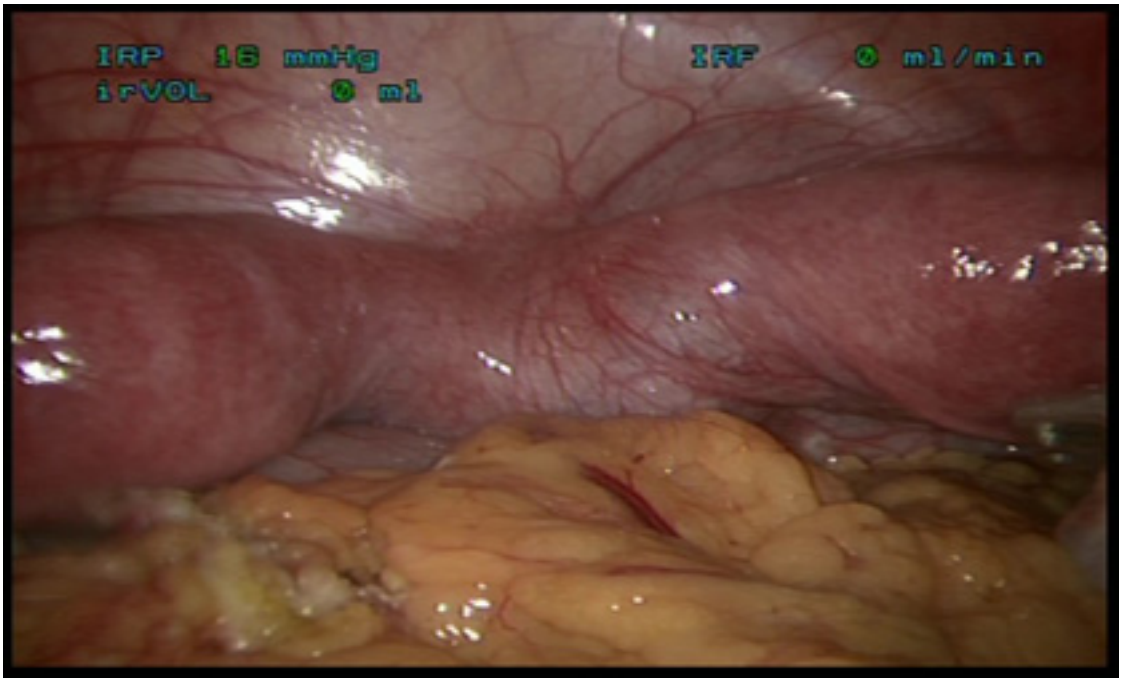
MRI imaging showed a utero-vaginal anomaly consisting of didelphys uterus and double vagina, one of which is obstructed and distended.,

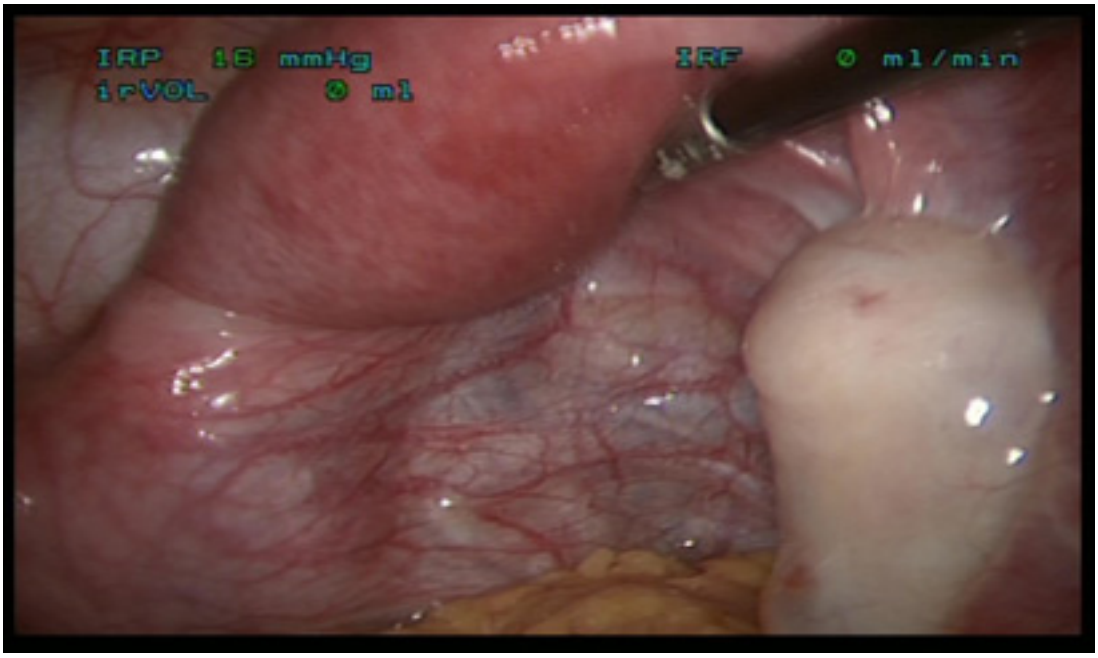
Figure 1: This MRI reveals double uterus cavities, double cervix and absent right kidney



A few months later, diagnostic laparoscopy was done for this patient which confirmed the diagnosis.







Discussion

This patient shows how it is difficult to diagnose and treat such uterine malformations. The presence of acute abdominal pain with symptoms like those of acute appendicitis without having proper ultrasound done by experienced radiologist and depending only on clinical picture lead to unnecessary surgical intervention.

Our patient's chief complaint was pelvis and lower abdominal pain with dysmenorrhea.

The symptoms usually begin after the menarche. Patients with this syndrome usually menstruate normally and may have no specific symptoms, except dysmenorreha. Thus 20% of these patients are diagnosed in their 20s and 10% are diagnosed beyond age 30 years (7).

Since the patient presented with an acute abdomen, appendicitis and ovarian torsion were a likely possibility following trans-abdominal ultrasound. The patient was taken to the theater, unnecessary laparotomy was performed.

Could more accurate diagnostic imaging have prevented the emergency intervention? Transvaginal ultrasound might have revealed the underlying disease in our patient which was hindered by her virginity.

The clinical manifestations and physical findings are very helpful to diagnose this syndrome. In addition, ultrasonography, computed tomography, MRI, and exploratory laparoscopy are used. MRI is the most effective method and helps to prevent unnecessary surgery (8).

Ultrasonography and MRI are widely and effectively used in the diagnosis of genitourinary anomalies, a 100% accuracy being reported for MRI because of its high accuracy and detailed elaboration of utero-vaginal anatomy (9, 10).

Transvaginal excision of the septum, large enough to allow a permanent drainage of the menstrual blood from the hemi-uterus is the appropriate mode of treatment as soon as the condition is diagnosed. The family of our patient did not accept doing transvaginal procedure as she is yet single.

Regarding their concern about her future fertility, they were reassured that women with uterus didelphys have a high likelihood of becoming pregnant (13), 80% are able to conceive (14).

At last, as a medical clue for the medical team if a young patient has renal anomaly look for associated vaginal and uterine anomaly this may help an early diagnosis

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