

Case Report : Herlyn Werner Wunderlich Syndrome

¹Sarah Safdar, ¹Ahmed Elgaml and ¹Sridevi Sankaran

¹ Frimley Health NHS Foundation Trust

Introduction

Herlyn-Werner-Wunderlich Syndrome (HWW) is a rare congenital anomaly that occurs in females. It is characterized by a triad of uterus didelphys, blind hemivagina, and ipsilateral renal agenesis. The syndrome results from the failure of the müllerian ducts to fuse during fetal development. It often presents with abdominal pain, dysmenorrhea, and a palpable mass. The incidence rate is truly unclear but some studies have reported incidence of mullerian anomalies to be in the range of 0.1% to 3% [1,2].

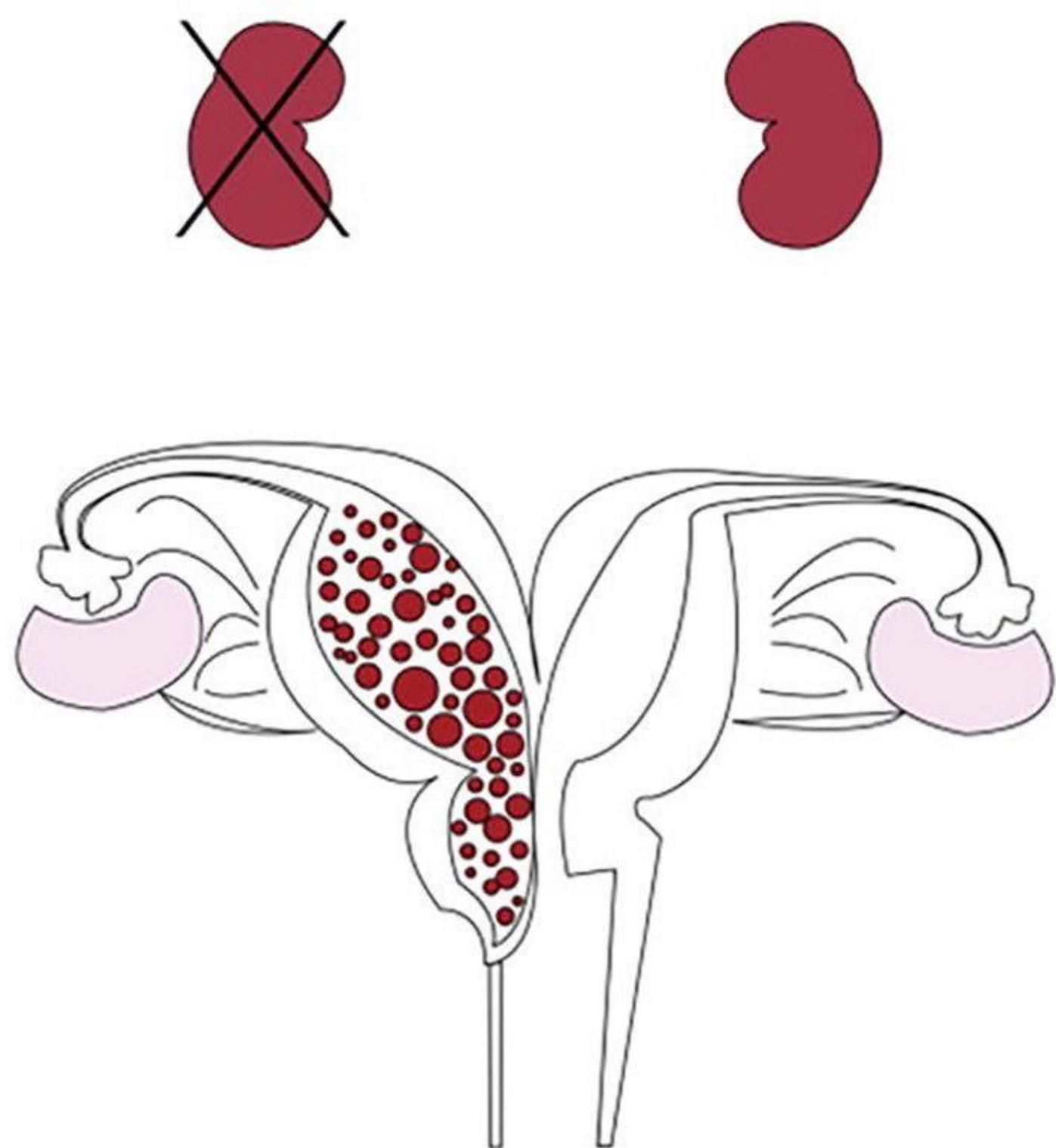


Fig 1. Pictorial representation of HWW [3]

CASE REPORT

Primary Complaint:

- Lower abdominal pain, primarily in the right iliac fossa, for six days

History of Patient Illness:

- Pain has been worsening overtime and patient was unable to stand due to pain
- Reported Fever of 38.1C
- Menarche 11 months prior and last menstrual period (LMP) was six days prior to presentation

Physical examination:

- Mild lower abdominal tenderness

Diagnostic evaluation:

- Laboratory results showed elevated white blood cell count (WCC) of 18.2 and CRP of 10
- Ultrasound revealed a gynaecological structural abnormality, leading to suspicion of appendicitis
- MRI revealed congenital left renal agenesis with septate uterus/cervix/hemivagina and obstruction of the left-sided tract low in the cervix or at a blind ending hemivaginal remnant, resulting in haematocervix, haematometra, left haematosalpinx, and moderate volume haemoperitoneum, consistent with a diagnosis of HWW syndrome

Management and outcome

- Patient was referred to a tertiary care hospital (Paediatric Surgical team)
- Commenced on progesterone for symptomatic relief
- Definitive treatment would be surgical.

Discussion:

Herlyn-Werner-Wunderlich Syndrome is a rare condition that presents with nonspecific symptoms such as lower abdominal pain, dysmenorrhea, and a palpable mass. The diagnosis is often delayed due to its rarity and lack of awareness among physicians. Imaging studies, such as ultrasound and MRI, play a crucial role in the diagnosis of HWW syndrome. Symptoms of appendicitis may overlap with HWW syndrome therefore, history taking, examination and radiological investigation should correlate before making a definitive diagnosis.

Treatment options include conservative management with hormonal therapy or surgical intervention. Surgical intervention is the definitive treatment for this syndrome.

Conclusion:

MRI appears to be the investigation of choice to diagnose this rare congenital condition. This type of rare case should be managed by a multidisciplinary team including paediatric surgery to provide prompt treatment and help reduce and prevent later complications. It is important to document and preserve the manifestations of this rare congenital syndrome.

References:

- 1-Zurawin RK, Dietrich JE, Heard MJ, et al. Didelphic uterus and obstructed hemivagina with renal agenesis: case report and review of the literature. J Pediatr Adolesc Gynecol 2004;17:137–41
- 2-Smith NA, Laufer MR. Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome: management and follow-up. Fertil Steril 2007;87:918–22
- 3.Jha S, Abdi S. Diverse Presentations of OHVIRA Syndrome: A Case Series. Clinical Pediatrics. 2021;60(1):6-8. doi:10.1177/0009922820953582