Case report

Acute abdomen: Unusual presentation of Herlyn-Werner-Wunderlich syndrome

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1. Introduction

Herlyn-Werner-Wunderlich (HWW) syndrome, also known as obstructed hemivagina and ipsilateral renal anomaly (OHVIRA), is a rare congenital malformation syndrome characterized by uterine didelphys, blind hemivagina and ipsilateral renal agenesis. The former name is due to its first report by Wilson in 1925, and the elaboration and description of the presentation described later by Herlyn and Werner in 1971 and Wunderlich in 1976 [1]. Here, we present a case of an 11-year-old female patient presenting to the emergency department with right lower abdominal pain mimicking appendicitis. She was later diagnosed as HWW syndrome. This case highlights the importance of knowledge on developmental anomalies in order to include it as part of the differential diagnosis. This case report has been reported in line with the SCARE 2020 criteria [2].

2. Case presentation

The patient is an 11-year-old Bahraini female presenting to the Accidents and Emergency with abdominal pain of 2 days duration. The abdominal pain is on the right lower abdomen. Its character is colicky with no radiation. The pain is aggravated by movement, with no relieving factors. It is associated with loss of appetite. There is no history of vomiting, fever, change in bowel habits or urinary symptoms. There was no similar previous episode nor any association with menstruation. Menarche was at 9 years old, with an irregular cycle, dysmenorrhea and oligomenorrhea. Patient was not sexually active nor on any medications. There was no relevant medical, surgical, drug, allergic, family or psychosocial history. On general examination, the patient appeared healthy, conscious, alert and not in distress with stable vitals. A focused physical examination only showed mild lower abdominal distension with rebound tenderness in the right iliac fossa.

Laboratory investigations, which included a complete blood count, electrolytes and urinalysis, were normal. The initial impression was acute appendicitis. However, further work-up was conducted via a pelvic ultrasound in order to rule out common gynecological conditions. Due to high suspicion of appendicitis, an abdominal ultrasound was conducted in order to rule out gynecological conditions such as a pelvic ultrasound. The ultrasound findings changed the approach, warranting a CT and MRI. The subsequent diagnosis was Herlyn-Werner-Wunderlich Syndrome. Intervention involved incision and dilation of the vaginal septum as it is the most effective and least traumatic method. On follow-up, the patient’s symptoms resolved.
done. Although appendicitis was at the top of the differentials, the ultrasound could not visualize the appendix with no intra-abdominal free fluid. Instead, the ultrasound showed a large well-defined oval cystic lesion (12 × 7 cm) communicating with the uterus, pushing the uterus superiorly and anteriorly. The cyst contains diffuse internal low-level echoes, suggesting hematocolpos or hydrocolpos. Abdominal ultrasound showed slightly enlarged left kidney due to compensatory hypertrophy with right renal agenesis (See Fig. 1). To visualize the renal anomaly, CT was conducted, which showed similar findings to the abdominal ultrasound with persistent mild, left hydronephrosis, hence confirming the ipsilateral renal anomaly (See Fig. 2). MRI was also conducted in order to visualize any uterine anomaly. It showed a large fluid-filled fusiform shaped structure (11.8 × 7.4 × 7.9 cm) in the pelvis extending to the lower abdomen in relation to the uterus. It illustrated evidence of two separate uterine bodies suggestive of didelphys or complete bicornuate uterus. The larger obstructed complex is fluid at the right side with a vertical septum measuring 3 cm in length and 0.5 cm in thickness. The left-sided uterine body is smaller in comparison with minimal fluid. The left uterus had its own separate cervix. Both ovaries, though displaced superolaterally, were normal. There were no abnormal adnexal masses or free fluid in the pelvis. MRI also confirmed the CT finding of right renal agenesis. MRI was hence suggestive of Mullerian duct anomaly with an obstructed bicornuate/didelphys uterus, with suspicion of a hemivaginal septum obstructing the right uterine body and hematometocolpos (See Fig. 3).

The imaging findings resulted in a deviation from the initial diagnosis and management of appendicitis to HWW syndrome. Preoperative preparations included NPO 6 h prior to the operation. The surgery was under general anesthesia in a lithotomy position. Operation done by a pediatric urology surgeon consultant and obstetrics and gynecologist consultant in the main tertiary hospital in the country. An intraoperative physical examination by vaginoscopy was conducted. The management of the patient was based on the most referenced surgical procedure in the literature, which is an incision and dilation of the vaginal septum. Prophylactic 3rd generation cephalosporin (ceftriaxone) 50 mg/kg given intravenously in order to prevent infection in the colpus. Following surgery, there were no adverse outcomes or medication requirements as the patient adhered and tolerated the provided intervention.

After discharge, post-intervention consideration was placed on follow-up in the outpatient clinic with repeated ultrasound every 6 months for two years. The patient was compliant to the ultrasound appointments. Prognosis for such patients is generally excellent, hence why the patient and her mother had a positive outlook and were satisfied with the outcome of the intervention.

3. Discussion

Mullerian development involves elongation, fusion, canalization and septal resorption. Failure in the fusion stage that proceeds elongation causes uterine didelphys (double uterus). Incomplete septal resorption at 20th week gestation leads to the blind hemivagina. Failure of normal development also affects the renal system as both genital and urinary systems originate from the same mesoderm fold, thus causing renal agenesis as the most common renal anomaly [3].

HWW syndrome is variable in onset and presentation. This is due to the different classifications and subtypes, such as the completely obstructed hemivagina and incompletely obstructed hemivagina [4]. Presentations can be in the form of dysmenorrhea, lower abdominal and pelvic pain, menstrual irregularities, and/or urinary manifestations [5–8]. In the presented case, there was only abdominal pain reported on the right lower abdomen, menstrual irregularities and dysmenorrhea.

The age of presentation is mostly following menarche but could still present at a later or earlier onset [8,9]. Although the uterus is still under the stimulation of maternal hormones in the neonatal period, detection has rarely been reported in newborns and infants due to asymptomatic presentation. It should therefore always have a high index of suspicion to prevent complications through early intervention. The presented case occurred at 11 years old, two years following menarche.

Ultrasound is the first employed modality to diagnose HWW. This is due to it being non-invasive, fast, grossly evaluating uterine anomalies and fluid collections in the vagina or uterus. MRI is a well-established tool in the diagnosis of uterine anomalies due to its ability to highlight the uterine morphology, cervixes, fluid collections or vaginal obstruction. It also precisely describes the anomaly and guides the operative management [10]. Those modalities are the ones commonly used in suspected HWW syndrome. There are cases where different modalities were additionally used, such as an IVP, hysterosalpingography, diagnostic laparoscopy and/or CT [6,8].

The most optimal management to relieve the obstruction is resection of the vaginal septum by incision and dilation, ideally on the 1st day of menstruation for proper septum identification [11]. The presented case

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**Fig. 1.** Ultrasound of the abdomen showing well-defined oval cystic lesion (12 × 7 cm) communicating with the uterus.

**Fig. 2.** CT of the abdomen showing enlarged left kidney and mild, left hydronephrosis with right renal agenesis.
was managed accordingly as it is the most effective, least traumatic and mostly referenced in the literature. The literature also reports other methods of managing HWW syndrome, such as hysteroscopic septum resection, laparoscopy/laparotomy or hemihysterectomy [5,8].

Preservation of fertility is a major concern in HWW syndrome, which is achieved by early management. There are multiple factors contributing to the delay of diagnosis. For instance, dysmenorrhea is usually managed by non-steroidal anti-inflammatory drugs and oral contraceptives, leading to menstrual suppression or reduction of pain without identifying the underlying cause [3]. Also, a patient with incomplete hemivaginal obstruction would not complain of symptoms related to outflow obstruction since menstruation is normal. Delayed diagnosis may lead to complications like endometriosis due to the retrograde blood flow, pelvic adhesions and infectious complications, including pyohematocolpos, pyosalpinx and pelvic abscess [12].

Essential lessons learned from such case presentation range from differential diagnosis to investigations and the possibility of preventing long-term complications. The differential diagnosis is affected by multiple factors. In the pediatric population, there are some common conditions like adenomesentritis or appendicitis. Gynecological conditions might also have a similar presentation to HWW syndrome. Thus, a major limitation is the rarity of the syndrome and its occurrence in a relatively small population in the country, with overlap between various disciplines. Radiology plays an essential role in the approach to acute abdomen. Ultrasound is the preferable initial investigation to identify the cause of acute abdomen in a female patient since it helps exclude gynecological abnormalities and other surgical causes of acute abdomen [13].

4. Conclusion

Herlyn-Werner-Wunderlich syndrome is a rare congenital anomaly. Patients with this syndrome present after menarche with complaints like dysmenorrhea, lower abdominal pain and/or urinary symptoms. Diagnosis is missed or delayed until the acute symptoms or complications arise. It is important for gynecologists, pediatricians and radiologists to consider it in the differentials in a female patient presenting with acute abdomen in order to detect it early on.

Informed consent

Written informed consent was obtained for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Sources of funding

There were no sources of funding used for this case report.

Ethical committee approval

Ethical approval was obtained as per the international guidelines.

Research registration

None declared.

Guarantor

Aysha Saleh; Maryam Alrashid; Saeed Al Hindi.

Additional information

Special thanks to the multidisciplinary team of pediatricians, obstetric and gynecologists’, and radiologists.

Provenance and peer review

Not commissioned, externally peer-reviewed.

CRediT authorship contribution statement

Aysha Saleh, Maryam Alrashid: writing – original draft, writing – review and editing, visualization, managing the literature searches.

Saeed Al Hindi: conceptualization, surgical therapy for this patient, supervision. All authors read and approved the final manuscript.

Declaration of competing interest

Authors declare that there is no conflict of interest.

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Fig. 3. MRI of the abdomen showing evidence of complete bicornuate uterus.
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