Obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) syndrome—A “no-touch-approach”: A case report

Ahlam Abdullah AlGhamdi 1*

1Department of obstetrics and gynecology, Imam Abdulrahman Bin Faisal University, Dammam, SAUDI ARABIA


ABSTRACT

Introduction: Herlyn-Werner-Wunderlich (HWW)/obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) syndrome is a rare Müllerian duct anomaly (MDA). Usually presenting after menarche classically with dysmenorrhea. Here we present a challenging case, where the diagnosis was missed initially.

Case presentation: A 10-year-old girl presenting with dysmenorrhea and constipation, treated repeatedly with analgesics, laxatives and enema. A pelvic ultrasound and magnetic resonance imaging (MRI) confirmed the diagnosis of OHVIRA syndrome, which requires surgical intervention. We describe a “no-touch-technique” for resecting the septum. The patient had immediate relief of symptoms and a smooth post-operative recovery.

Conclusions: Early diagnosis and surgical management are crucial not only to relieve symptoms but to prevent long term complications that may affect the patients’ reproductive potential. Despite pelvic ultrasound being an affordable and a useful diagnostic tool, the diagnosis can be missed by inexperienced operators. Therefore, MRI is recommended to confirm and classify MDAs. The hysteroscopic “no-touch-technique” to resect the septum is an effective, safe and a convenient approach to treat the condition while preserving hymen integrity.

Keywords: case report, HWW, hysteroscopy, OHVIRA

INTRODUCTION

Herlyn-Werner-Wunderlich (HWW) syndrome is a rare variant of Müllerian duct anomalies (MDAs) consisting of uterine didelphys, obstructed hemivagina, and ipsilateral renal agenesis. The syndrome was initially described by Herlyn and Werner in 1971, and later Wunderlich in 1976 described association of right renal agenesis with a bicornuate uterus, a vagina and an isolated haematocervix [1]. Patients with HWW syndrome are usually asymptomatic until menarche, when they present with acute lower abdominal pain. It can also present with a pelvic mass and or acute retention of urine. Only a few hundred such cases have been reported in literature till date. Furthermore, the mainstay of management is surgical correction. We report a case with unusual presentation and describe a suitable treatment option in pediatric population. A written informed consent was obtained from the father of the patient for publication.

CASE PRESENTATION

A 10-year-old girl presented with her parents to the emergency department of King Fahad Hospital of the university, AlKhubar, Saudi Arabia, complaining of lower abdominal pain for three months. The history was obtained from the mother, who stated that her daughter was having cyclical pain for three months. It was colicky in nature, mainly in the lower abdomen and low back that radiates to the rectum causing constipation, lasting for 10 days each time. It has also interfered with her school attendance. She had no urinary symptoms, nausea, vomiting, or weight loss. The mother mentioned that her daughter had her menarche just six months earlier, and her cycles were regular, very minimal flow, for five days duration and is associated with secondary dysmenorrhea. The patient had frequent emergency room visits due to constipation, where she was given laxatives and enema and discharged home. She was born at term and had no family history of congenital disease. The patient had no medical illness, except for a solitary kidney that was diagnosed incidentally in early childhood. She never had any surgeries, or blood transfusions. Upon examination, her blood pressure, pulse rate and respiratory rate were all normal and she was afebrile. She was lying comfortably in bed, not in pain, and there was a lower abdominal distension. The patient was given analgesics for relief of symptoms. Her complete blood count was within normal limits with a hemoglobin level of 12.9 g/dl, hematocrit 37.8%, red blood cell counts 4.64×10^{12}/L, platelet count was 641×10^9/L, white blood cell count was 8.4×10^9/L, with normal differential count. The erythrocyte sedimentation rate was 23 mm/first hour. Routine urine and microscopic examinations showed no features of infection. Pelvic ultrasound revealed a round and elongated distended structure in the midline directly posterior to the bladder measuring 7.25×6 cm, with internal echoes and posterior enhancement and no internal vascularity. Findings were most
likely a hematometrocolpos, hence she was admitted to the gynecology department and a pelvic MRI was performed, confirming an MDA in the form of left hematometrocoplos and obstructing left hemi-vagina with Gartner duct cysts and absent left kidney, consistent with HWW syndrome (Figure 1 & Figure 2).

A schematic drawing of the abnormality is illustrated in Figure 3 for further explanation.

The parents were counseled regarding options of treatment, including surgical resection of the septum for symptomatic relief or removal of the left hemi-uterus and hemi-vagina. They opted for the first option at this stage, keeping in mind the possibility of re-closure of the hemi-vagina and the recollection of blood in the left hemi-uterus. The vaginal approach in a virgin girl is of concern in our region for cultural reasons. So, the plan was to use a vaginoscope “no touch” approach. The father consented for hysteroscopic resection of the vaginal septum and drainage of the hematometrocolpos for symptomatic relief. She received intravenous cefazolin and metronidazole pre-operatively and positioned in a lithotomy position and stirrups after induction of general anesthesia. The area was prepped and draped in a sterile fashion. The urinary bladder was catheterized, then a five mm, 12 degree rigid hysteroscope was introduced through the hymenal opening into the vagina using normal saline as fluid media. There were two vaginae, the hysteroscope was directed towards the right hemi-uterus and a small cervix was seen, then the right hemi-uterus was entered, and the cavity was visualized clearly, and the right tubal ostia was seen. Then the scope was directed towards the left vagina, where the septum was seen bulging and transected with the hysteroscopic scissors under ultrasound guidance. Large amount of dark brown thick secretions around two liters was drained. This was followed by irrigation of the left hemi-uterus with normal saline and suctioning till the cavity was seen clearly but the left tubal ostia was not appreciated. A 14 Fr foley’s catheter was placed into the left hemi-vagina with inflation of the balloon with 50 ml air to prevent adhesions of the incised vaginal septum. The patient had immediate relief of symptoms on her second day postoperatively and was

Figure 1. Pelvic MRI T2 sequence demonstrates duplication of uterus (*), cervix, & vagina (arrow) (reprinted with permission of the patient)

Figure 2. Pelvic MRI T2 sequence showing two cervices (arrow) & a left side unilateral septated lobulated hematocolpos (star) (reprinted with permission of the patient)

Figure 3. Schematic presentation of left OHVIRA (Source: Author’s own elaboration)
discharged from the hospital and prescribed oral ampicillin and metronidazole for one week and oral contraceptive pills. She was seen a week later in the clinic for catheter removal, where she was doing well.

**DISCUSSION AND CONCLUSIONS**

HWW syndrome is a triad of uterine didelphys, obstructed hemivagina, and ipsilateral renal agenesis. It was first reported by Purslow in 1922, as a maldevelopment of both Müllerian and Wolffian ducts [2]. The uterus, cervix, fallopian tubes, and the upper third of the vagina are derived from the Müllerian duct. While the kidneys originate as ureteric buds from the Wolffian ducts, and the lower third of the vagina is derived from the urogenital sinus [3]. The prevalence of MDAs has been reported to range between 1-5% in the general population and 13-25% in females with recurrent miscarriage [4]. The main cause of congenital MDA is defective fusion or septal resorption failure, which is influenced by polygenic, and familial factors [5]. Obstructed hemi-vagina and ipsilateral renal agenesis is explained by embryological arrest at eight weeks of pregnancy, simultaneously affecting both the Müllerian and Wolffian ducts [6]. It was studied 87 patients having double uterus with obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) over a period of 30 years [7]. It was found that classic HWW syndrome, was seen in 63 cases (72.4%), 10 out of 87 patients (11.5%) had a septate uterus whereas bicornuate uterus was seen in nine patients (10.3%) [7]. HWW has been further classified based on the morphology of the vagina [1].

The syndrome usually remains undiagnosed during early childhood [8]. Symptoms classically, dysmenorrhea arises a few months to a year of menarche. It can also present with a pelvic or vaginal mass, abnormal vaginal discharge, acute retention of urine, fever, vomiting [9], infertility, endometriosis, complicated pregnancy and labor [10]. The mean age of presentation is 15 years [11]. Our case presented six months after menarche at 10 years of age with dysmenorrhea and constipation and was treated with simple analgesics, laxatives and occasionally enema. The diagnosis was missed because she had normal menstrual flow, and no urinary retention, which are the usual presentations.

Diagnosis needs a high index of suspicion despite the rarity of the syndrome. Ultrasound is an initial, affordable and a simple tool but it’s operator dependent and diagnosis can be missed by inexperienced operators. Abdomino-pelvic MRI, however, is an excellent choice for evaluating, classifying MDA, and helps planning proper surgical treatment especially in pediatric patients [12].

Once the diagnosis of OHVIRA syndrome is made, surgical management is the cornerstone of treatment in order to relieve symptoms and to prevent long term complications related to retrograde menstrual flow including endometriosis, pelvic adhesions, and infertility [1].

Historically, surgeons advocated a two-stage procedure. Initial surgery to drain the hematocolpos and the second to resect the septum. However, single-stage vaginoplasty, is encouraged in the contemporary literature [6], the vaginal septum resection has been conventionally done using retractors, scissors, scalpels, and sutures [13]. Lately, minimal invasive approach has been adopted for better visualization, and less postoperative pain. Vaginal septotomy is usually done by hysteroscopic approach rather than laparoscopic technique [14]. Whereas laparoscopic approach is reserved for hemihysterectomy to treat patients with a didelphic uterus and a hypoplastic cervix [15, 16].

The technique we have adopted was described earlier [17]. The “no-touch” technique was the most suitable approach for our patient, to maintain hymenal integrity, which was of utmost importance to both the patient and her parents for cultural values related to sexuality, and religion. We believe this approach has several advantages including its safety, improved visualization, decreased post-operative pain since no vaginal retractors or speculum were used, and maintaining hymen integrity.

To our knowledge there are three cases with OHVIRA syndrome reported in the gulf region, two from Saudi Arabia [18, 19] and one from Bahrain [20]. One underwent a laparoscopic right hemivaginal drainage for hematocolpos and vaginal septotomy, whereas there were no details on the surgical intervention in the other two cases.

In conclusion, HWW/OHVIRA syndrome has variable clinical presentations. MRI is considered gold standard for diagnosis, classification of MDA, planning treatment strategies and diagnosing associated renal anomalies. Early diagnosis and surgical correction is needed to relieve symptoms and reduce the risk of long term complications. The “no-touch” technique vaginoscopy and hysteroscopic resection of the septum is a suitable treatment modality in pediatric and adolescence population.

**Funding**: No funding source is reported for this study.

**Acknowledgements**: The author would like to thank all the surgical team involved in the case.

**Ethical statement**: The author stated that no ethical approval is required; however, a consent was obtained from the patients’ father.

**Declaration of interest**: No conflict of interest is declared by the author.

**Data sharing statement**: Data supporting the findings and conclusions are available upon request from the author.

**REFERENCES**


