

Chronic Vaginal Discharge Syndrome Caused by Obstructed Hemivagina and Ipsilateral Renal Agenesis (OHVIRA) Syndrome: A Case Report

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Abstract

Background: Vaginal discharge syndrome is a common condition across the world with the main causes being infectious. Rare causes include obstructed hemivagina with ipsilateral renal agenesis (OHVIRA syndrome). **Case Presentation:** This case involves an 18-year-old woman with a 9-year history of vaginal discharge treated unsuccessfully by multiple physicians and different health facilities. After she presented to our hospital gynecology clinic with the same complaint she investigated with blood, urine, discharge wet-mount, pelvic ultrasound, and abdominopelvic computed tomography at different times. Preoperatively she was suspected to have OHVIRA syndrome which was successfully surgically treated with a full return of function. **Conclusion:** This case emphasizes the need to consider non-infectious causes commonly congenital mullerian anomaly in young women presented with long-time vaginal discharge symptoms soon after menarche and which are nonresponding to different multiple medical treatments. In peripheral health institutions like ours, it is best to consider computed tomography if available but better not to forget at least abdominopelvic ultrasound to look for congenital Mullerian anomalies.

Keywords

OHVIRA Syndrome, Vaginal Discharge Syndrome, Early Marriage

1. Introduction

Obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) is a very rare condition affecting between 0.1% - 3.8% of women and previously known as Herlyn-Werner-Wunderlich Syndrome, it was the first case described by Pur-

slow in 1922, who described as the anomalies of uterine didelphys, obstructed hemivagina, and ipsilateral renal agenesis [1]. The most common (50%) variant is left-sided obstructed hemivagina, isolated hematocolpos (55.9%), uterine didelphys (82.9%), and ipsilateral renal agenesis (92.2%) and the most common presentation consists of abdominal pain, dysmenorrhea (73%), and an abdominal mass secondary to hematocolpos (71%) [1]-[3]. The nonspecific presenting complaints and heterogeneity of the presenting symptoms can make the diagnosis a clinical challenge and few guidelines exist to aid in the best approach to clinical management [2] [4].

The syndrome, a rare Müllerian duct anomaly, presents a diagnostic challenge due to its complex nature and varied clinical manifestations, gives hard time for diagnosis. The advanced diagnostic approach like magnetic resonance imaging (MRI) and multidisciplinary approach is standard and is difficult to apply in resource limited setup. [5] [6]

OHVIRA syndrome, characterized by uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis, is typically managed through a minimally invasive approach, with a focus on surgical excision of the obstructed hemivaginal septum and hematometra drainage. This approach has been found to be effective, with a shift towards minimally invasive techniques due to improved imaging and preoperative diagnosis. Single-stage vaginoplasty is considered the gold standard treatment, with MRI findings potentially correlating with surgical outcome. However, vaginal stenosis is a potential postoperative complication. [4] [7] [8]

This is a case of obstructed hemivagina and ipsilateral renal agenesis from rural Ethiopia in a general hospital. Patient who presented with vagina discharge for a long duration of time visited a different facility for the same problem almost half of her life.

2. Case Presentation

An 18-year-old woman presented to our department with the main complaint of chronic vaginal discharge. She was married at the age of eight (though it is illegal for the country to marry any lady less than age 18) and her husband divorced her soon after her marriage. The discharge began soon after her divorce which coincided with menarche. The discharge was described as malodorous and copious at times. Over the next nine years, she visited five hospitals and received various oral and injected medications for which there were no records.

On presentation to Soddo Christian Hospital Gynecology referral clinic, both breast and pubic hair examinations were consistent with Tanner stage five. A minimal amount of malodorous discharge was noted on the speculum examination with a cervix that was slightly deviated to the left. There was no evidence of recto-vaginal or vesico-vaginal fistula, which was suspected due to her early age of marriage. Wet prep examination demonstrated red blood cells with no evidence of bacterial vaginosis or yeast vaginitis. For which she was initially treated

with cephalexin, doxycycline, and metronidazole and upon return in two weeks for follow up she was noted to have no improvement.

A transvaginal ultrasound scanning came up with the suspicion of a bicornuate uterus. The investigation was continued with the acquisition of a computed tomography scan (only advanced diagnostic modality for region) which demonstrated left renal agenesis, a duplicated ureter on the left, and a bicornuate uterus (**Figure 1** and **Figure 2**). From patient history and those two diagnostic findings, OHVIRA syndrome was the prime diagnosis and plans were made for examination under anesthesia as well as the appropriate surgical intervention.

During examination under anesthesia, except for visible discharge coming from the left side vagina very close to the cervical opening at around 4 o'clock, and slightly boggy to the left side vagina than right side, all other findings are consistent with clinical findings. Stab incision was made in the left lateral vaginal wall (the place where maximum boggy felt) (**Figure 3**). A large amount of bloody purulent material was noted which confirmed the presence of an obstructed hemivagina. The discharge drained completely then the wall of this medial hemivagina was then excised and repaired with a delayed absorbable suture. A communication with the cervix on the left side was noted and there was no appreciable cervix on the left. Patient transferred to the postanesthesia care



Figure 1. Computed tomography scan shows the absence of the left kidney and hypoechoic area to the left vagina (**Figure 1** is original).

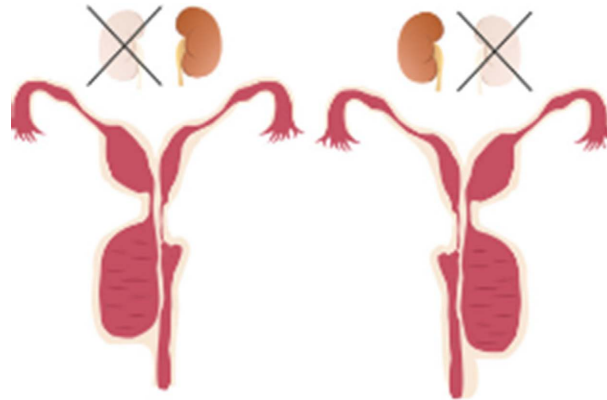


Figure 2. Summary of obstructed Rt and Left hemivaginal and ipsilateral renal agenesis. To get uterine anomaly, commonly bicornuate uterus. Our case also had uterine anomaly, likely bicornuate uterus which we suspected during TVS examination because there was indentation which looked more than 90 degree. This finding supports the embryogenesis that gives lower 1/3 vagina is from different embryonic origin (urogenital sinus) than upper 2/3 and rest of female genital tract from mullerian duct [9].

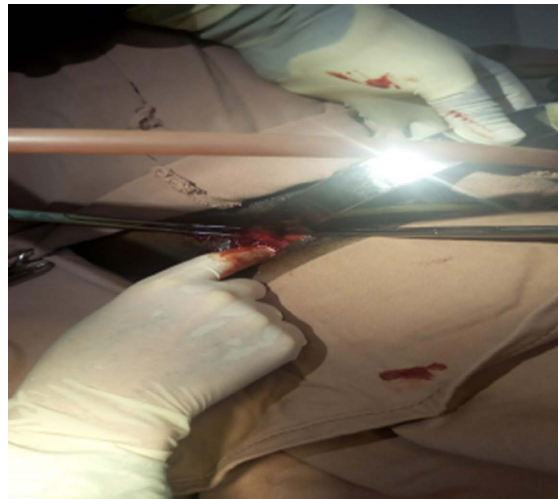


Figure 3. The surgeon excised the left lateral wall of the vaginal septum and examined the extent of obstructed hemivagina before repair (Figure 3 is original).

unit with stable vital signs then the maternity floor after 4 hrs of stay in recovery.

Postoperatively she did well and was discharged home on postoperative day number one. She did not return for her initial follow-up at six weeks because of financial and social issues. However, she did return five months postoperatively and was doing well without any complaints. Her vaginal discharge had resolved and she was having regular menses without any other additional gynecologic complaints. Examination demonstrated a normal-appearing vaginal wall, appropriate length and opening with a single cervix in the midline.

3. Discussion

Even Though not a common presentation, one case report showed the same problem patient presented with acute abdomen and dysmenorrhea managed

with hemivaginal septum resection and anastomosis [10] and another case reported acute urinary retention as a presentation for the patient who had OHVIRA syndrome diagnosed later and managed with vaginal septum resection [11]. So from this, we realize that patients with the mentioned syndrome can present either acute or chronic complaints as in our case, considering obstructive uterovaginal anomalies warranted.

One retrospective case study on 27 cases of congenital mullerian anomaly, of which 26 were managed vaginal reconstruction with additional laparoscopy for clarification of primary diagnosis and followed for more than 12 yrs for evaluation of surgical outcomes stating one case ended up with repeat vaginoplasty, two cases with infection, four cases with anatomical distortion, another two cases with restenosis and eight cases with vaginal septum adenosis [4]. But in our case, we managed with opening the obstructed hemivagina completely and sutured the edge with a vaginal approach and the patient did well. We did not do laparoscopy for confirmation because we agreed that it doesn't add value for patients but rather adds cost since we have had other investigations before that showed Mullerian and renal anomalies.

Because of the nature of disease and variety of clinical presentation, timely diagnosis is challenging in many countries especially in resource limited. Pelvic ultrasound and examination under anesthesia shall be considered as minimal diagnostic modality in resource limited setup, but in better set computed tomography is very helpful though the magnetic resonance imaging is diagnostic.

Long time follow up after surgery for patients who are managed for OHVIRA syndrome is very important before five years of age if applicable for success of surgery, and specific complications for the surgery like vaginal stenosis. But because of the diagnosis challenge and diagnostic material shortage, the diagnosis takes a long time like in our case. Despite the age of surgery the important point is to get regular follow up after surgery is recommended. In resource limits setup like our place we recommend at least one time follow up in 4 - 6 wks of surgery for those who have financial and other constraints to make regular follow up.

4. Conclusion

OHVIRA is a challenging diagnosis to make, especially in resource-challenged countries. The complaints can be non-specific and challenging to properly diagnose. In cases where symptoms coincide with menarche, it is wise to consider the diagnosis of OHVIRA and perform the necessary testing. Magnetic resonance imaging is diagnostic, but if it is not available, computed tomography scan and/or abdominopelvic ultrasound can be used. We were able to address her surgical needs vaginally without the need for laparoscopy or laparotomy, which is ideal and recommended as a minimally invasive surgery approach.

Consent

Our institution has no formal consent for case reports but for ethical issues I

consented to patient according to the Ethiopian law which amended any person age 18 and above can represent himself with his signature on issues regarding himself or for his family.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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