CASE REPORT

OHVIRA Syndrome with a Rare Presentation

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Abstract

Background: Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome is a rare complex Mullerian anomaly with an incidence of 1:20,000. The commonest presentation is pain in the abdomen and progressive dysmenorrhea. Other rare symptoms could include vaginal discharge, fever, infertility, or acute abdomen.

Case description: A 15-year girl presented with acute retention of urine which required catheterization. She had attained menarche 8 months back. On evaluation, the ultrasonography report revealed the presence of uterine didelphys with right side pelvic collection and absent kidney on the same side of the collection. On magnetic resonance imaging (MRI), a right obstructed hemivagina was clearly seen along with other findings suggesting the OHVIRA syndrome. Laparoscopically, a bulge was seen just below the right uterine horn which caused the retention of urine. Septal resection was performed vaginally, following which her symptoms subsided.

Clinical significance: Suspect OHVIRA syndrome in adolescent girls when there is a renal anomaly with Mullerian defect. The patient can also present with acute retention of urine because of hematocolpos. Early detection and treatment will help to prevent complications of endometriosis and adverse fertility outcomes.

Keywords: Acute urinary retention, OHVIRA syndrome, Renal agenesis, Uterine didelphys.

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Introduction

Girls in their adolescence do come with abdominal pain and menstrual pain in our day-to-day practice. In rare situations, where there is a missing kidney with uterine malformation and a vaginal collection, we should think of OHVIRA syndrome which is a complex genitourinary malformation.

Commonly, patients present with symptoms within a few years after the onset of menses. The common symptoms include cyclical pain in the abdomen, progressive dysmenorrhea, and sometimes palpable pelvic mass. They can present late as primary infertility or after infectious complications with fever, vomiting, and pelvic mass. It may be detected late in certain patients as they would be having normal menstrual flow from the unobstructed side. Sometimes, it is also diagnosed as early as in neonates where they present with muco or hydrocolpos or even prenatal period where the scan has shown pelvic collection in a female fetus. But urinary retention is a rare symptom in these patients. It may be because of hematocolpos pressing on the neck of the bladder and obstructing the urinary outflow.

Case Report

A 15-year-old girl came to our casualty with acute urinary retention for 1 day. She was not able to void urine though she had the urge to void. She also had complaints of colicky pain in the abdomen, which started with her present menstrual cycle, with a gradual increase in severity. Her last menstrual cycle was 15 days back. She went to a local hospital and was referred to our institution with Foley's catheter *in situ*.

She attained menarche 8 months back. Her prior cycles were on time and she bled for 3–4 days not associated with dysmenorrhea. General physical examination was unremarkable with stable vitals. She had normal secondary sexual characteristics (Tanner staging of breast, axilla and pubic hair were IV). On abdominal examination, there was no palpable mass or tenderness. Local examination revealed normal-looking external genitalia.

Abdomen and pelvic ultrasonography (USG) revealed right kidney could not be visualized (renal agenesis), a large well-defined thick-walled anechoic cystic lesion (12 x 6 x 7.2 cm) in the right adnexa? hematometrocolpos, and a possibility of uterus didelphys. Further evaluation by magnetic resonance imaging (MRI) was advised. The MRI of abdomen and pelvis showed two separate uterine horns (5.1 x 2.5 x 3.1 cm on the right side and 4 x 2.6 x 1.9 cm on the left side), two endometrial cavities, cervix, and vagina – suggestive of the uterus didelphys (type III), large dilated fluid-filled right hemivagina with communication with the right uterine horn suggestive of the obstructed vagina with hematometrocolpos, ipsilateral renal agenesis. Based on the imaging findings of unilateral renal agenesis, uterus didelphys, and unilateral obstructed hemivagina with resultant hematometrocolpos, the case was diagnosed as OHVIRA syndrome.

Examination under anesthesia and diagnostic laparoscopy was planned. Figure 1 shows the on per speculum examination

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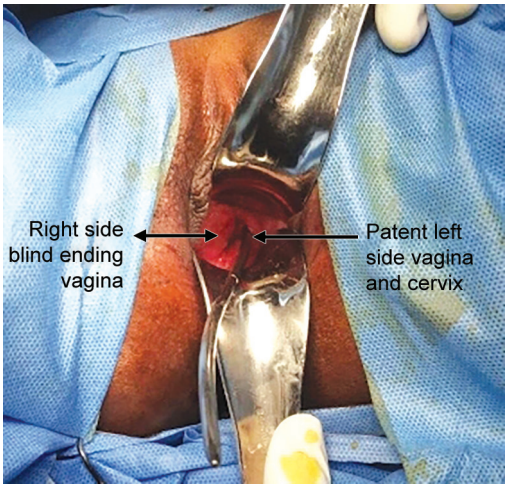

Fig. 1: Per speculum examination showing left cervix with Hegar's dilator *in situ* and right cervix is not visualized due to the thick septum

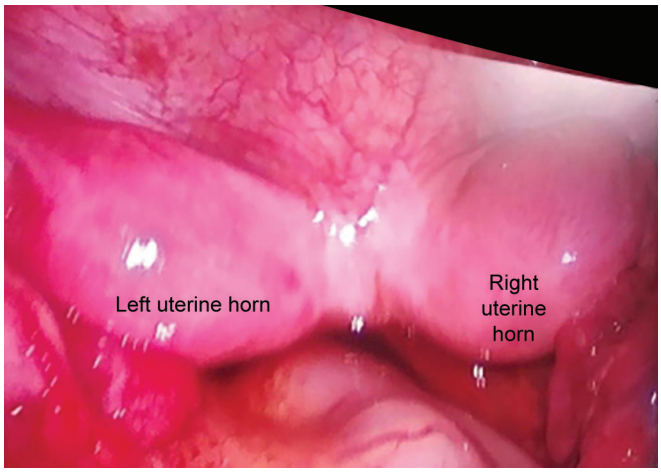

Fig. 2: Laparoscopic view of uterine horns and a bulge noted below the right horn which could have pressed on the bladder leading to retention of urine

of the left side cervix, which was visualized and confirmed by placing Hegar's cervical dilator, whereas the right-side cervix could not be visualized. A thick septum was noted. Diagnostic laparoscopy was done, which showed two uterine horns. Figure 2 shows that on the lower aspect of right uterine horn, a bulge was visualized (hematocolpos), which was probably pressing on the neck of the bladder and that was the reason for the retention of urine.

Vaginally septal resection was done and around 150 mL of chocolate color fluid (old collected blood) was drained. This was confirmed laparoscopically by a gradual reduction in the size of the bulge. Then right-side cervix was visualized, confirmed by placing Hegar's cervical dilator which could be passed till the fundus of the right uterine horn. The resected septal wall was sutured to anterior and posterior vaginal walls (like marsupialization). She was treated with hemivaginal septal resection and drainage of hematometrocolpos.

Following surgery, the catheter was removed and she voided urine normally, post-op period was uneventful. She came for follow-up after her next menses and she had no complaints.

Discussion

The first case of OHVIRA syndrome was reported in 1922. Renal agenesis and Ipsilateral blind hemivagina were described in 1971 by Herlyn and Werner. Wunderlich described the association of uterine malformation in 1976. Hence, it is also known as Herlyn-Werner-Wunderlich syndrome.

Obstructed hemivagina and ipsilateral renal anomaly syndrome occurs due to anomalous development of the Wolffian ducts. Wolffian duct gives rise to the ureteric bud which forms the kidney. Wolffian duct also acts as an inductor for the normal development of Mullerian duct. On the side, Wolffian duct is absent, the Mullerian duct gets displaced more laterally, and thus it fails to fuse with the contralateral Mullerian duct leading to uterine didelphys. The lateral displaced Mullerian duct (on the side of absent Wolffian duct) cannot come in contact with the urogenital sinus leading to obstructed hemivagina on the same side.

Lan Zhu et al. suggested that the syndrome can be classified into two types (classes 1 and 2) depending on whether the obstruction is complete or partial. It is again subdivided into 1.1, 1.2, 2.1, or 2.2. Our case belongs to class 1.1, wherein the affected hemivagina is completely obstructed, and the uterus behind the septum is totally isolated from the contralateral one. This condition has a good prognosis after septal resection. In class 1.2, the hemivagina is completely obstructed and the cervix behind the septum is maldeveloped/atretic, and these cases need hemihysterectomy. Whereas in class 2.1, a small communication is noted between the two hemivaginas, which results in the late detection of the cases. In class 2.2, there is a small communication between the duplicated cervices, and these cases usually present with spotting per vagina.[1]

The diagnosis of OHVIRA syndrome is mostly done after menarche because patients usually present with cyclic, increasing lower abdominal pain, and progressive dysmenorrhea due to the formation of hematocolpos resulting from long-standing retained menstrual blood in the obstructed hemivagina. But many a times, it may go undiagnosed because the menstrual flow from the normal side could be misleading. Dysmenorrhea, of course, is a common symptom in this age group and commonly gets treated by analgesics without thorough evaluation.

Rarely, it can present as acute abdominal pain, abnormal vaginal discharge, infertility. Initial clinical diagnosis is incorrect in the majority of cases, because of its rare incidence and misleading presenting signs and symptoms. But there are no case reports of acute retention of urine in OHVIRA syndrome cases. Funda et al. showed that no patients presented with acute urine retention in a set of 32 patients with OHVIRA syndrome. All of them either presented with pain in the abdomen and dysmenorrhea; one with primary infertility and one with acute pain in the abdomen.[2] Another study of 13 patients also did not demonstrate acute urine retention as a symptom.[3] A case series of 8 patients too showed that only abdominal pain and dysmenorrhea have the common presenting symptoms.[4] Sharma R et al. reported that if vaginal discharge is persistent and resistant to treatment, then it could show as a rare manifestation of OHVIRA syndrome class 2.1.[5] This case highlights that the retention of urine could also be a symptom in OHVIRA syndrome as the pelvic collection (hematocolpos) can press on the neck of the bladder.

There are few case reports wherein this condition has been diagnosed in the premenarchal period as well, as they can have pyometra and symptoms related to it.[6]

Right-sided abnormalities are more common (60–70%) compared with the left sided abnormalities for reasons unknown. Even in our case, it is on the right side. In girls with unilateral renal anomaly and a Mullerian anomaly, kindly look to rule out OHVIRA syndrome. Other than renal agenesis, renal dysplasia, renal cysts, double or ectopic ureter can be associated. Some other associations reported include high bifurcation of aorta, duplication of inferior venacava (IVC), intestinal malrotation, and ovarian malposition.

Ultrasonography and MRI are effectively used in the diagnosis of genitourinary anomalies. A 100% accuracy has been reported for MRI. It helps in the evaluation of uterine defect, presence of septa in vagina, atretic cervix, and complications like endometriosis. In our case, MRI was helpful to confirm the diagnosis, as in USG, hemivagina was not detected. These findings were subsequently confirmed by the examination under anesthesia and diagnostic laparoscopy.

Single-stage vaginal resection/vaginoplasty is considered the gold standard. Hysteroscopic septal resection may be preferred in young girls to maintain virginity. Hemihysterectomy is an alternate option for atretic cervix, proximal vaginal septum, or if infectious complications have occurred. So, depending on the type of malformation, either resection of the septum or half the uterus may be necessary. If more time for the decision regarding surgery is required, continuous oral contraceptive pills can be started to avoid the formation of a hematometrocolpos. The septum could be oblique or transverse and the thickness of the septum may vary. Postoperatively, patients should be followed-up and closely monitored for possible closure of the septum.

Conclusion

The emphasis is on different ways of presentations of anatomical malformations of Mullerian and urinary systems so that it can be detected early and can be timely corrected. OHVIRA syndrome patients can also present with acute retention of urine, as the hematocolpos might press on the bladder neck and obstruct the urinary outflow.

High level of suspicion can result in early diagnosis and prompt the surgical intervention which can prevent complications in adolescent girls and provide them with good future fertility.

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