Case Report

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Obstructed hemivagina with ipsilateral renal anomaly with ureteric remnant in a prepubertal female

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Abstract:

Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA), or Herlyn–Werner–Wunderlich syndrome, is a rare Müllerian duct anomaly that is characterized by a triad of uterus didelphys, unilateral obstructed vagina, and ipsilateral renal agenesis. Patients with this anomaly usually present after menarche with abdominal pain or a pelvic mass which worsens over some time. Herein, presented is a case of a 12-year-old nulligravid with cyclic pelvic pain and palpable pelvic mass after her menarche. A series of diagnostic tests were done which were compatible with the diagnosis of OHVIRA with an associated finding of ectopic ureteral insertion into the obstructed hemivagina and nonfunctional urinary bladder. The patient underwent diagnostic hysteroscopy, cystoscopy, and full resection of the longitudinal vaginal septum as well as drainage of hematocolpos, which are the main treatment of the patient with OHVIRA.

Keywords:

Ectopic ureter, ipsilateral renal agenesis, Müllerian duct anomaly, ureterocele, uterus didelphys

Introduction

Müllerian duct anomalies are a group of uncommon entities, which cause specific symptoms in adolescent females. Obstructed hemivagina and ipsilateral renal anomaly syndrome (OHVIRA), formerly known as the Herlyn–Werner–Wunderlich syndrome, is a very rare congenital anomaly of the urogenital tract characterized by the triad of uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis. Its estimated occurrence is between 0.1% and 3.8% of women worldwide. In the review of the *Philippine Journal of Obstetrics and Gynecology* from 2015 to 2019, two journal reports were published. Both presented a case series of four patients with OHVIRA.[1]

The exact cause, pathogenesis, and embryologic origin of OHVIRA are unclear.[1] Patients with this anomaly usually present after menarche with recurrent progressive pelvic pain, palpable mass, and rarely, primary infertility in later years.[1] The mainstay of diagnosing OHVIRA is radiologic findings.[1] Sonography and magnetic resonance imaging (MRI) are extremely useful in diagnosing and classifying Müllerian duct anomalies. Resection of as much of the obstructing vaginal septum as possible is the optimal surgery for patients.[1,2] Delay in its diagnosis and management is inevitable due to its rarity; however, the complications can be devastating which include endometriosis, ovarian endometrioma, upper genital tract infection, pelvic adhesions, ectopic pregnancy, recurrent pregnancy loss, and infertility.[1,3] Overall, timely surgical removal of the obstructing vaginal septum can provide rapid relief of symptoms, prevention of complications, and preservation of fertility in the majority of the cases.[1,2]

This article describes a case of OHVIRA with ectopic ureteral insertion to the

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obstructed hemivagina and nonfunctional urinary bladder who was diagnosed and treated early. This study aimed to increase awareness regarding Müllerian anomalies specifically uterine didelphys with a noncommunicating vaginal septum, to discuss the ideal diagnostic tools and management of OHVIRA, and to promote early diagnosis and treatment to avoid long-term complications.

Case Report

A 12-year-old nulligravid presented with recurrent pelvic pain and palpable mass since menarche at the age of 11. Subsequent menses were regular, lasting for 7–10 days, soaking 1–2 pads per day, with associated dysmenorrhea and palpable hypogastric mass. Five months before admission, patient experienced her menarche which lasted for 13 days, consuming one lightly soaked pad per day, associated with crampy pelvic pain (10/10 in pain score) radiating to the right buttocks. There was no associated dysuria, changes in bowel habits, fever, nausea, vomiting, or body weakness. On the 10th day of the menstrual cycle, a consultation was done with a gynecologist. The patient was given celecoxib and medroxyprogesterone which offered relief and cessation of vaginal bleeding after 3 days. Interval history revealed the persistence of cyclic pelvic pain and hypogastric mass; however, no consultation was done. Two months before admission, cyclic pelvic pain was noted to be more severe prompting immediate consultation to another gynecologist and was advised admission for further evaluation and management. A pelvic sonography was done which revealed a normal-sized anteverted uterus with slightly prominent endometrium, no uterine or adnexal mass or cyst seen, and an opening from the urinary bladder to the cervix measuring 1 cm × 1 cm. A cystogram was done which revealed a distended urinary bladder, no filling defect identified, and no mass or fistula appreciated at the time of the study. Then, a renal ultrasound was done which revealed small size right kidney with calculus measuring 0.5 cm, small size left kidney with pelvocaliectasia, and urinary bladder fistula to the cervix measuring 1 cm × 1 cm. Transrectal and transabdominal ultrasound was done which showed two cervices, fused in the midline, both with distinct endocervical canals, wherein the right is dilated measuring 2.1 cm; there was a longitudinal, hypoechoic band measuring 0.4 cm which divided the vaginal canal into two up to the middle third of the vagina; there was a low-level echo fluid collection measuring 9.7 × 8.1 × 5.9 cm (volume: 242 mL) superior to the band; a connection between the contents of the cystic structure and the right hemivagina, and no connection was seen between the cystic structure and urinary bladder; the left cervix measured 2.2 cm × 1.8 cm × 1.6 cm; two corpora uteri were seen which were pushed anteriorly up to the level of the umbilicus; the right corpus measured 4.8 cm × 5.4 cm × 3.1 cm and the endometrium on the right was hyperechoic measuring 0.3 cm; dilating the endometrial cavity is a low-level echo fluid collection measuring 4.6 cm × 3.7 cm × 1.9 cm (volume: 16.9 mL) with intact subendometrial halo; the left corpus measured 3.7 cm × 3.4 cm × 2.3 cm and the endometrium was hyperechoic measuring 0.4 cm with intact subendometrial halo; the right and left ovaries were normal measuring 3.7 cm × 3.4 cm × 2.3 cm and 3.4 cm × 3.5 cm × 2.2 cm, respectively; there were no adnexal masses seen and no free fluid in the cul-de-sac. These ultrasonographic findings were suggestive of uterine didelphys, longitudinal vaginal septum with hematocolpos within the right hemivagina, thin endometrium right hematometra, normal ovaries, and incidental findings of right renal agenesis. Hence, a computerized tomography urogram with contrast was done which revealed right renal agenesis and uterine didelphys with fluid-filled structure seen coursing through the right side of the right uterus which indents inferior at the posteroinferior border of the urinary bladder. The left kidney, urinary bladder, and other visualized upper abdominal organs were unremarkable. These findings were suggestive of OHVIRA; hence, the patient was referred to a urogynecologist.

The patient has no previous surgical operation. She has no history of sexual contact.

On physical examination, the patient was conscious, coherent, and not in cardiopulmonary distress. Vital signs were stable. The abdomen was flat, with normoactive bowel sounds, tympanitic, soft, with noted direct hypogastric tenderness, and no palpable mass. The perineum was grossly normal with no lesion, discharges, and no palpable lymphadenopathy on the bilateral inguinal area. There was an anterior cystic mass palpated on digital rectal examination. The uterus was slightly enlarged and there were no palpable adnexal masses or tenderness.

An abdominopelvic MRI with contrast showed duplication of the uterine cavity and vagina. The right hemivagina was markedly distended with fluid signal intensity with a thin enhancing transverse vaginal septum noted at the level of the symphysis pubis, with leftward displacement and marked compression of the left hemivagina. A suspicious fistula was noted between the right and left hemicervices. A tortuous, dilated, blindly ending tubular structure was seen arising from the anterolateral aspect with noted bulbous expansion at its inferior aspect just posterolateral to the urinary bladder likely representing an ectopic right ureter with ureterocele [Figure 1]. Both ovaries appeared unremarkable with the right arising from the right hemiuterus and the left arising from the left hemiuterus. A 1 cm × 1.2 cm cyst was seen at the inferior and

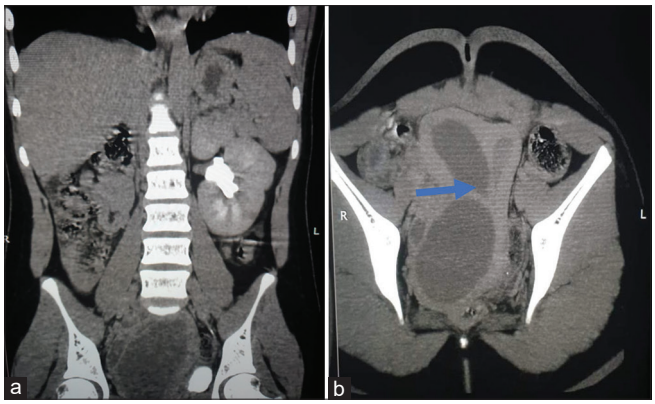

Figure 1: Computed tomography scan of the urinary tract. (a) Right renal agenesis with normal left kidney. Didelphic configuration of the uterus, with fluid seen in the endometrial cavity on both sides. The arrow on (b) shows the nonresorbed uterovaginal septum

anteriorly to the vagina. The right kidney was absent, and the left kidney was enlarged measuring about 5.0 cm × 6.4 cm × 11.6 cm, with mild left hydronephrosis detected, and the left ureter was nondilated. These MRI findings were compatible with OHVIRA [Figure 2].

The initial assessment was nulligravid, Herlyn–Werner–Wunderlich syndrome, uterine didelphys, double vagina with noncommunicating longitudinal vaginal septum, obstructed right hemivagina with right hematocolpos, and right renal agenesis.

The patient underwent resection of the noncommunicating longitudinal vaginal septum, drainage of hematocolpos, diagnostic hysteroscopy, and cystoscopy on the 9th day of her menstruation.

During vaginoscopy, there was note of an intact pinkish hymen measuring about 10 mm in the anteroposterior diameter and the vaginal canal measured 3 cm from the inferior border of the obstructed right hemivagina to the hymen. The left hemivagina measured 8 cm from the hymen, compressed by the longitudinal vaginal septum, pinkish vaginal canal with rugae, and left cervix seen superior to the longitudinal vaginal septum. During the resection of the longitudinal vaginal septum, 500 ml of blood was evacuated from the right hemivagina and the edges of the resected septum were sutured to the remaining upper and lower vaginal mucosa using interlocking sutures. The right cervix was seen after resection of the longitudinal vaginal septum. During hysteroscopy, both right and left hemiuteri showed slightly thickened endometrium with tubal ostium. Upon withdrawal of the hysteroscope from the right hemiuterus, there was an opening in the vagina just lateral to the right cervix. Upon entry, there was a bladder-like mucosa seen. There was no ureteral opening inside the bladder-like mucosa pouch and no connection to the functional urinary bladder. On cystoscopy, the

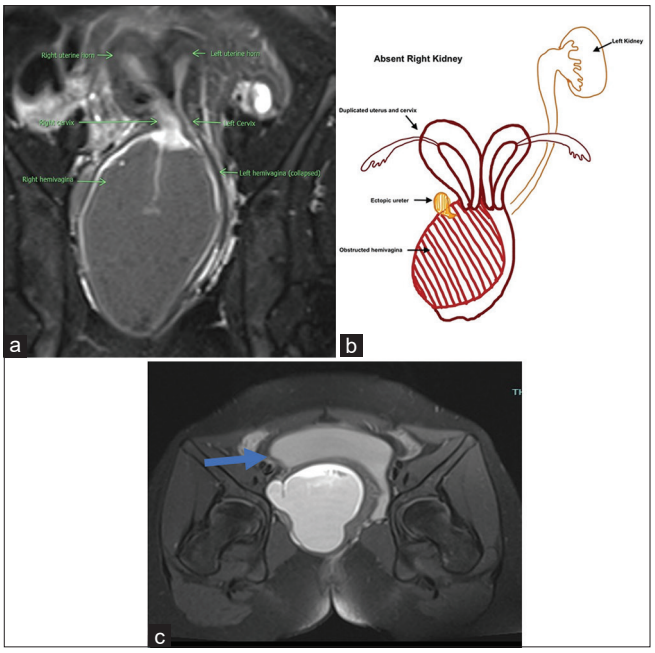

Figure 2: MRI of the uterus. (a and b) (Line diagram showing the anatomy) Duplicated uterus, cervix, and vagina, hematocolpos in the right hemivagina with a tortuous, dilated, blindly ending tubular structure arising at its anterolateral aspect with noted of bulbous expansion at its inferior aspect just posterolateral to the urinary bladder likely representing an ectopic right ureter with ureterocele (c) and absent right kidney. MRI: Magnetic resonance imaging

functioning bladder was visualized with a steady jet of fluid seen on a single ureteral opening on the left. There was no right ureteral opening seen. The histopathologic report revealed fibromuscular tissue with focal stratified squamous and mucinous epithelia consistent with vaginal septum [Figure 3].

One month after discharge, the patient had her menstrual period soaking 2–3 pads per day with no associated hypogastric pain or pelvic mass. After her menstrual period, a digital rectal examination revealed good sphincteric tone, smooth rectal wall, empty rectal vault, unenlarged uterus, and no palpable adnexal mass or tenderness. Six months after discharge, the patient claimed that she has been menstruating monthly, soaking four pads per day, lasting for 5 days, with no associated hypogastric pain or palpable mass.

Discussion

Müllerian duct anomalies affect the development of the female genitourinary system.[4] Uterus didelphys and blind hemivagina associated with ipsilateral renal agenesis are collectively known as OHVIRA, a rare variant of Müllerian duct anomalies.[2,4] The incidence of uterus didelphys related to OHVIRA is approximately 1 in 2000–1 in 28,000, and it is accompanied by unilateral renal agenesis in 43%.[1] In our institution, this is the first confirmed case of OHVIRA with an ectopic ureteral insertion to the

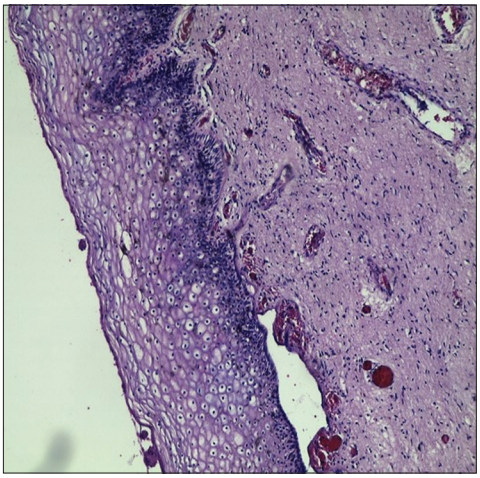

Figure 3: Histopathologic imaging of the resected septum. A fibromuscular tissue with focal stratified squamous and mucinous epithelial consistent with vaginal septum

obstructed right hemivagina and a nonfunctional urinary bladder.

According to the American Society for Reproductive Endocrinology, OHVIRA represents class III uterine anomaly, which is a consistent finding in our patient.[2,5] Mesonephric and paramesonephric ducts are two paired urogenital structures from which internal genital organs and lower urinary tract are derived.[6] A didelphic uterus results due to embryologic arrest during the 8th week of gestation. This ultimately affects the Müllerian and metanephric duct.[3,7] The characteristic features of our patient make us think about the exact embryogenesis of this abnormality. The presence of the ectopic ureter with insertion into the obstructed right hemivagina suggests that the mesonephric ducts distal to the origin of the ureteric bud get absorbed in the paramesonephric ducts instead of the probable bladder. This may be the primary mechanism which causes the pathology. The abnormal renal ascent and position can be explained by the abnormal ureteric bud and abnormally fused mesonephric duct causing obstruction to the normal ascent.[8,9]

A study by Jiali Tong *et al*. reported that there were significant differences between patients with complete obstruction and incomplete obstruction. The primary gynecologic complaint is dysmenorrhea, and some presented with intermittent mucopurulent discharge and irregular menstruation.[2] In the presence of menstrual outflow obstruction, hematometra, hematosalpinx, and endometriosis were the main reasons for cyclic pelvic pain and irregular menstruation.[1,2] A vaginal septum is present in 75% of the cases of uterus didelphys. Patients with this anomaly are usually asymptomatic until menarche. They present with recurrent progressive pelvic pain and/or a mass and rarely, in later years, with primary infertility.[1] It is reported that most patients are diagnosed from 2 months to 1 year after menarche.[4] Physical examination typically reveals a unilateral pelvic mass, more commonly on the right than the left, with a ratio of 2:1.[4] The retention of menstrual blood in the obstructed hemivagina leads to the formation of hematocolpos which is clinically detected as a pelvic mass. Our patient presented with cyclic pelvic pain and palpable pelvic mass since menarche, worsening over the past months despite no other associated symptoms, with physical findings of grossly normal perineum and slightly enlarged uterus, and different imaging studies compatible with OHVIRA.

Sonography and MRI are extremely useful in diagnosing and classifying Müllerian duct anomalies and are considered the mainstay in diagnosing OHVIRA.[1] The ultrasonography has an accuracy of 85%–92% which shows features of this condition which include uterus didelphys, bicornuate uterus, with or without uterine effusion, and ipsilateral renal agenesis with compensatory hypertrophy of the contralateral kidney.[1] MRI is the most useful tool in classifying uterine anomalies, in which discrete images are required, and helps to prevent unnecessary surgery.[1] Even in such cases, given the high accuracy of MRI (96%–100%), laparoscopic examinations of hysterosalpingography are reserved for very limited cases.[1] Hysteroscopy can detect intrauterine adhesions and communication between the duplicated endometrial cavities. In our patient, different imaging tests were done showing uterine didelphys, noncommunicating longitudinal vaginal septum, obstructed right hemivagina with hematocolpos, and right renal agenesis, suggestive of OHVIRA with an incidental finding of an ectopic right ureter with ureterocoele on MRI. Because of the rarity of an ectopic ureter associated with uterine didelphys with obstructed hemivagina, the diagnosis of an ectopic ureter can be easily overlooked. In our case, the ectopic ureter and uterine didelphys were seen on MRI and were confirmed on diagnostic hysteroscopy and cystoscopy. Embryologically, the reproductive system develops in close relationship with urinary system. Therefore, renal function and renal ultrasound should be routinely followed due to other urologic abnormalities.[4]

Currently, the most effective treatment is surgery. Surgical management aims to provide symptomatic relief, prevent complications, and maximize the fertility of a patient.[1] For patients with symptomatic obstructed hemivagina, resection of as much of the obstructing vaginal septum as possible is the optimal surgery for patients and drainage of the hematocolpos.[1,2] Most patients can recover completely after resection of the vaginal septum. The best time for surgery is approximately at the time of menstruation as a large distended hematocolpos is easy to visualize and palpate, which aids in resection.[2] After being diagnosed with

renal agenesis or renal malformation by imaging studies, laparoscopic or transabdominal resection of the atretic uterus is suggested.[2] After surgery, the next step is to suppress menstruation temporarily to prevent re-accumulation of menstrual blood.[3] However, in our patient, she was allowed to menstruate and was evaluated right after menstrual bleeding ceased with noted unremarkable findings. The ectopic ureter was managed conservatively and was not excised since there were no urinary symptoms such as urinary incontinence and urine leakage experienced by the patient. Various studies have reported that the communication of an ectopic ureter with obstructed hemivagina leads to renal distention and prevents involution. However, they also stated that there occurs self-involution of the ureteric remnants once the obstructed hemivagina is drained.[9]

Summary

OHVIRA should be suspected in cases with cyclic pelvic pain and also in neonatal cases with any renal malformations. Understanding its broad spectrum of phenotypic presentation may aid in screening out the over-looked population. The diagnosis of this syndrome may be difficult due to its rarity, and mostly in a clinical setting, pelvic pain and mass are the only presenting signs and symptoms of OHVIRA which are not pathognomonic of this syndrome, resulting in misdiagnosis. Therefore, detailed history, physical examination, and high suspicion are necessary. Diagnostics are valuable in confirming the features of this syndrome. MRI is the method of choice for diagnosing OHVIRA. Surgical intervention should be the first line of therapy with full resection of the vaginal septum and drainage of hematocolpos.

Conclusion

Early diagnosis and timely intervention are essential for preventing potential complications. Patients also need to be searched for urologic and gynecologic problems and regularly monitored for renal function.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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