



Case Report

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Obstructed Hemivagina with Ipsilateral Renal Anomaly (OHVIRA) Syndrome - A Rare Congenital Anomaly

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Abstract

Objective: We present a rare case of Obstructed Hemivagina and Ipsilateral Renal Anomaly (OHVIRA) Syndrome with a single septate uterus. This syndrome is shrouded in controversies concerning its nomenclature and development. We attempt to highlight the different viewpoints surrounding this unusual condition, which can be quite distressing for the affected young girls and confusing the treating doctors.

Design: Case report.

Materials and Methods: Case details and review of literature.

Results: A 14 year old girl was presented with recent onset severe dysmenorrhea and abdominal mass of twenty week sized uterus. She had already received treatment from different physicians and gynecologists, for her ailment, in her hometown. Imaging studies in our hospital revealed that this patient had absent right kidney, massive hematometocolpos on right side with normal endometrial echo seen on the left side. She was diagnosed to have OHVIRA syndrome. She underwent septostomy of the vaginal septum, drainage of hematometocolpos and hysterolaprosopy that showed single septate uterus with a broad fundus.

Conclusion: OHVIRA is a rare syndrome of mullerian&wolffian duct abnormality. A simple excision of the vaginal septum can relieve the patient of her symptoms. Given the rarity of the syndrome it is frequently misdiagnosed or diagnosed late. Delayed diagnosis leads to endometriosis endangering her already compromised fertility potential. Hence greater awareness and early diagnosis and timely surgery can prevent future complications.

Keywords

Mullerian duct anomaly; Obstructed hemivagina; Ohvira syndrome; Vaginal septum

Introduction

Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome with uterine anomaly is a rare disorder. Incidence of mullerian duct anomalies ranges from 0.8% to 4%. The true incidence of the OHVIRA syndrome is not precisely known, but according to the available literature it is estimated between 0.1-3.5 percent of all mullerian anomalies [1]. Patient is usually a young girl presenting

with dysmenorrhea. As the patient continues to have regular menstruation with normal flow, diagnosis is frequently delayed. Early diagnosis and excision of the vaginal septum will relieve the patient of her symptoms and also prevents subsequent development of endometriosis and infertility.

We present a case of OHVIRA syndrome and review the available literature regarding the controversies surrounding this anomaly.

Case Report

The patient, a 14 year old girl, had attained menarche one year back. She had regular menses with normal flow initially but, of late, had started developing progressively severe dysmenorrhea. She also complained of vomiting, constipation and difficulty in micturition, during menstruation and few days thereafter. In the last five months, different physicians had treated her symptomatically for vomiting and constipation, and gynecologists dispensed treatment for dysmenorrhea and urinary tract infection, in her hometown. The girl and her parents were scared of going through the torture of another menstrual cycle.

This young girl had well developed secondary sexual characters, breast - Tanner stage 4 with axillary and pubic hair - Tanner stage 4. Abdominal examination revealed a mass of twenty week sized uterus. The mass had varied consistency with cystic feel all over except on the superior aspect, where it was firm to feel. The external genitalia were normal; there was no visible bulge or bluish discoloration. On direct questioning the patient revealed that the mass increased in size during menses and slightly decreased thereafter. There was no history of similar complaints, anomalous children or ovarian cancers in her family.

Her hemoglobin was 8g %. Ultrasonography of abdomen and pelvis revealed absent right kidney and Uterus showed two cornual. Left cornu showed normal endometrial echo whereas right cornu was distended with echogenic contents within. Vagina was hugely distended with echogenic fluid suggestive of hematometocolpos (Figure 1). Communication with the left sided cornu could not be demonstrated; hence, mullerian duplication was suspected. MRI was advised which identified the left sided cornual communication with left sided cervix and a narrow communication with left side of vagina (Figures 2 and 3). Because of the huge right sided hematometocolpos the left sided communication of vagina and the cervix was seen as a hypo intense band. A provisional diagnosis of OHVIRA syndrome was made.

Examination under anesthesia showed a bulge in the right side of vagina occupying whole of upper vagina. Vaginal septostomy was done and 1.5 liters of dark chocolate colored blood was drained. Hysteroscopy confirmed septate uterus, with the septum extending from fundus of the uterus through the cervix till about five centimeters above the level of introitus (Figure 4). Laparoscopy done at the same sitting revealed a single uterus with a broad fundus (Figure 5). Right fallopian tube was dilated. Bilateral ovaries were normal. Superficial endometriotic spots were noted over the pelvic peritoneum. Vaginal septum was excised till the level of cervix. Post-operative recovery period was uneventful.

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On follow up the patient continues to have regular pain free menses, six months after the surgery.

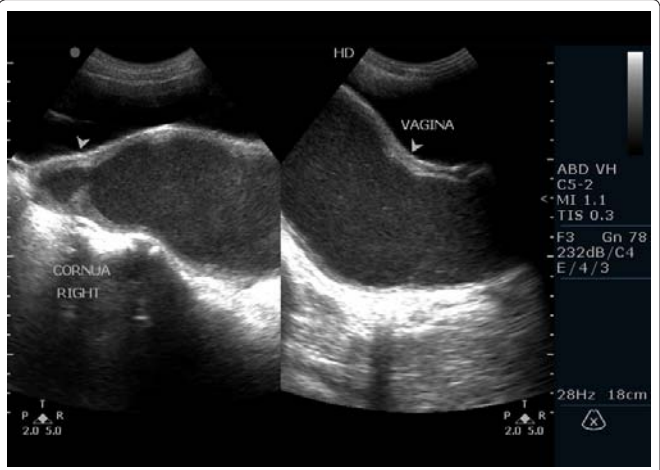


Figure 1: Ultrasoundimage of largehematometocolpos.

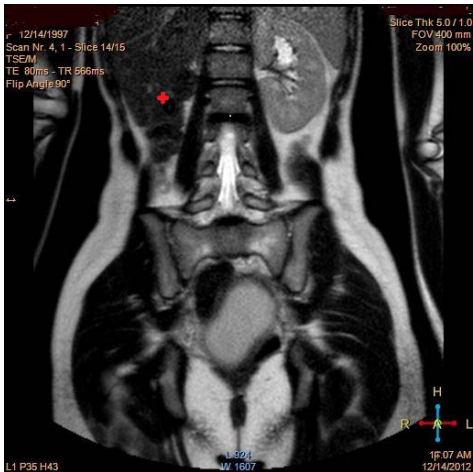


Figure 2: MRI image depicting absent right kidney and hematometocolpos.

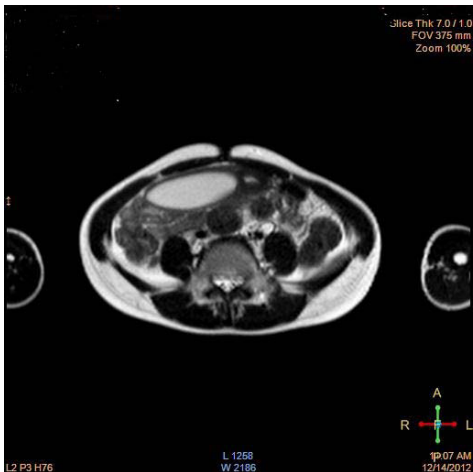


Figure 3: MRI image depicting left cornu with normal endometrium and right cornu showing large hematometocolpos.



Figure 4: Vaginal septum (after vaginal septostomy and drainage of hematometocolpos).



Figure 5: Single uterus with a large fundus.

Discussion

Origin of OHVIRA is shrouded in controversies. The origin of acronym OHVIRA and another similar condition called Herlyn-Werner-Wunderlich syndrome (HWW) syndrome has been laid to rest by Kimble [2]. Herlyn-Werner-Wunderlich syndrome (HWW) syndrome is a triad of obstructed hemi vagina, uterine didelphys and ipsilateral renal anomaly. The triad is named as HWW syndrome in Bulgarian article by Nalbanski and Gholum et al. used the term Herlyn -Werner-Wunderlich syndrome for the firsttime in English literature in 2006. Acronym OHVIRA is used to describe the triad of obstructed hemi vagina and ipsilateral renal anomaly and any other uterine anomaly other than uterine didelphys [2]. Both the syndromes are rare and are discussed as same entities in many case reports. The presentation and management of the patients with both the syndromes does not differ very much, so we are of the opinion that naming the entity with different names only adds to the prevailing confusion. We are of the opinion that this entity should be referred to as OHVIRA syndrome, andshould includeall forms of uterine anomalies.

The etiology of the syndrome is not known. There is a 12 fold increase in the prevalence of OHVIRA syndrome in the first degree relatives of the individuals with mullerian duct anomalies. Nicole Smizer has discussed a case of monozygotic twins with mirror imaged anomalies of OHVIRA [3]. Presentation in monozygotictwins with mirror imaged anomalies supports familial aggregation and might suggest a monogenetic phenotype of OHVIRA syndrome. This syndrome can be associated with other abnormalities like urogenital sinus, bladder exstrophy and other renal anomalies like dysplastic or duplicated kidney. The simultaneous insult to the paramesonephric system and metanephros could also suggest a multifactorial origin.

Acien has recently challenged the accepted classical embryological theory of development of female urogenital system. Acien has proposed that vagina is completely of mesonephric (wolffian) origin; hence, OHVIRA is not a Mullerian duct anomaly but a

mesonephric anomaly. OHVIRA syndrome is included in the second group i.e 2.1 of his proposed Embryological-clinical classification for female genital-urinary malformations [4]. Bajaj et al. have summarized the embryological development as related to OHVIRA syndrome. An early failure of metanephric diverticulum to develop from mesonephric duct results in agenesis of ureteric bud, leading to ipsilateral agenesis of ureter and kidney. Mesonephros is responsible for development and positioning of paired paramesonephric ducts in close proximity. Due to failed positioning of paired paramesonephric duct, the two hemiuteri and hemi cervixes fail to unite resulting in müllerian anomalies associated with OHVIRA syndrome [5].

OHVIRA syndrome is rare and lack of awareness of this syndrome frequently leads to delayed diagnosis or misdiagnosis. The classical presentation is that of a young girl presenting with severe dysmenorrhea, few months to one year after attaining menarche, as in our patient. Usually they are treated symptomatically or as endometriosis until they develop an abdominal mass and pressure symptoms. Retrospective study of cases has shown that the mean age of presentation is about 15 years [6]. Pelvic pain is the most common presenting symptom (90%) followed by an abdominal mass (40%) and pressure symptoms. Patients can also present at a later age with foul smelling vaginal discharge due to pyocolpos [7].

Ultrasonography is helpful in diagnosis of these cases, but MRI is usually conclusive. The standard management of these patients is excision of the vaginal septum and drainage of hematometrocolpos. Cetinkya et al. have discussed about the use of hysteroscope to excise the vaginal septum and preserving the hymenal integrity [8]. Laparoscopy may be done in the same sitting to clearly identify the uterine anomaly. Endometriosis is a frequent finding during laparoscopy.

After excision of the vaginal septum, precautions must be taken to keep the outflow tract patent. The patient may use vaginal mould at nighttime. Cooper et al. used a tracheobronchial stent to keep the outflow tract patent [9].

Fertility of patients with OHVIRA syndrome is not substantially compromised. Septate uterus alone is not an indication for surgery. However surgery is indicated if there is infertility or adverse obstetric outcomes. Hysteroscopic metroplasty is the choice of surgery for the uterine defects.

Conclusion

OHVIRA is a rare syndrome of Müllerian & Wolffian duct abnormality. A simple excision of the vaginal septum can relieve the patient of her symptoms. Given the rarity of this syndrome it is frequently misdiagnosed or diagnosed late. Delayed diagnosis leads to endometriosis endangering her already compromised fertility potential. Hence greater awareness and early diagnosis and timely surgery can prevent future complications.

References

1. Resetskova N, Christianson M, Kolp L (2012) Uterine didelphys with obstructed hemivagina and ipsilateral renal agenesis with hydronephrosis. *Fertil Steril* 97: s30-s31.
2. Kimble RMN, Kimble RM (2009) The obstructed hemivagina, ipsilateral renal anomaly, uterus didelphys triad. *Aust N Z J Obstet Gynaecol* 49: 4-7.
3. Smizer N, Appelbaum H (2012) Obstructed Hemivagina and Ipsilateral Renal Anomaly in Monozygotic Twins. *J Pediatr Adolesc Gynecol* 25: e27-e48.
4. Acien P, Acien MI (2011) The history of female genital tract malformation

classifications and proposal of an updated system. *Hum Reprod Update* 17: 693-705.

5. Bajaj SK, Misra R, Thukral BB, Gupta R (2012) OHVIRA. Uterus didelphys, blind hemivagina and ipsilateral renal agenesis: Advantage MRI. *J Hum Reprod Sci* 5: 67-70.
6. Christianson MS, Yates MM, Woo I, Khafagy A, Garcia J.E, et al. (2012) Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA): diagnostic features and management of a frequently misdiagnosed syndrome. *Fertil Steril* 98: S222.
7. Dhar H, Yasser AR, Illham H (2011) Uterus didelphys with obstructed hemivagina, ipsilateral renal agenesis and right pyocolpos: A case report. *Oman Med J* 26: 447-450.
8. Cetinkya SE, Kahraman K, Sonmezer M, Atabekoglu C (2011) Hysteroscopic management of vaginal septum in a virginal patient with uterus didelphys and obstructed hemivagina. *Fertil Steril* 96: e16-e18
9. Cooper AR, Merritt DF (2010) Novel use of a tracheobronchial stent in a patient with uterine didelphys and obstructed hemivagina. *Fertil Steril* 93: 900-903.

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