Original Research Article

OHVIRA and OSVIRA - Two sides of the same coin

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Abstract

Background: OHVIRA and OSVIRA are rare developmental mullerian duct anomalies. OHVIRA is seen in young females and presents with lower abdominal pain and infertility. OSVIRA is male counterpart and presents in reproductive age with lower abdominal pain and dysuria. Both the anomalies are surgically correctable. USG is initial modality of choice and then confirmed with MRI or CT. The purpose of study was to recognize imaging features of these syndromes. **Materials and Methods**: This is retrospective study performed in our department at a tertiary health care institute on 7 patients (4 females and 3 males) from June 2017 to November 2019. All of them were evaluated with clinical history and initial USG abdomen(on SIEMENS X 300 USG MACHINE and Hitachi Aloka Arietta 70 USG Machines) .Subsequently MRI of abdomen and pelvis(on 3T Siemens Verio MRI scanner using body coil) / CT abdomen and pelvis(on SOMATOM DEFINITION AS+ CT SCANNER)was done .MR Sequences used were T1, T2 , DWI, SWI and gradient sequences in various planes and CT was done with plain and post contrast venous phases. **Results**: In our study, it is found that all the patients with OHVIRA syndrome presented with lower abdominal pain and dysmenorrheal and infertility in the married patient. All the patients showed uterine didelphys with obstructed hemivagina resulting in hematometra and hematocolpos and with ipsilateral renal agenesis.2/3rd of patients with ZINNER syndrome present with lower abdominal pain and dysuria .All the patients on imaging showed seminal vesicle cyst, dilated ejaculatory duct and ipsilateral renal agenesis. **Conclusion**: The knowledge about these syndromes amongst radiologist leads to their early diagnosis and creates a positive impact on life. Ultrasound is the initial modality which can suspect and diagnosis followed by MRI or CT for confirmation. Ipsilateral renal agenesis should prompt the suspicion of associated mullerian anomaly and vice versa.

Key Words: Mullerian, OHVIRA, OSVIRA, Zinner

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INTRODUCTION

Reproduction is one of the important characteristics of a living creatures; so also humans. It is an inevitable need for propagation of a specie. Infertility affects propagation of specie. In the human context its social and cultural impacts are profound. They cause huge emotional psychological and social burden on the person and family. There are many common causes of infertility which are frequently encountered and thereby familiar to radiologists. There are few uncommon which are either misdiagnosed or undiagnosed. This leads to significant delay in the management of the patient, thereby affecting the success rate of treatment. Knowledge on uterine anomalies is important as they have great impact on women's physical social and cultural quality of life. Prevalence of common uterine anomalies is reported to be 7% to 10% in the general female population.[1] Herlyn-Werner-Wunderlich (HWW) syndrome is one such rare and underdiagnosed developmental anomaly of female urogenital tract and includes finding of uterus didelphys and obstructed hemivagina and ipsilateral renal agenesis (OHVIRA).[2,3] Majority of the females suffering from

OHVIRA syndrome are diagnosed late due to the rare incidence of the anomaly and the non specific clinical presentation. Also, the menstrual flow from the unobstructed hemivagina gives the appearance of normal menstrual cycle. Consequently accurate diagnosis and surgical treatment gets delayed for several months or even years. OHVIRA could present with lower abdominal pain, severe dysmenorrhea, a pelvic or vaginal mass, abnormal vaginal discharge, intermenstrual bleeding[4,5], acute retention of urine, fever, vomiting[6], infertility and abdominal swelling or complication with pregnancy and labor [7] The counterpart of OHVIRA in males is OSVIRA which is more commonly known by the term Zinner syndrome. Zinner's syndrome is a rare developmental anomaly of mesonephric duct consisting of unilateral seminal vesicle cyst and ejaculatory duct obstruction and ipsilateral renal agenesis. It is due to developmental arrest in early embryogenesis affecting the caudal end of Mullerian duct [8]

AIMS AND OBJECTIVES

Based on our experience of 7 patients over the course of two and half years and these syndromes being so rare and yet important causes of infertility and morbidity to the patients, we aim to spread awareness about ZINNER and OHVIRA syndromes with their imaging findings.

MATERIALS AND METHODS

This study was performed in the Department of Radio-diagnosis in a tertiary health care institute in 7 patients referred to our department over a period of thirty months. 4 female and 3 male patients were included in the study based on the final diagnosis. The female patients were in the age range of 10-20 years and male patients were in the age range of 25-35 years in our study. These patients underwent Ultrasonography(USG) as the initial modality with 2-5 MHz curvilinear, 5 to 10 MHz linear probe and 4 to 9 MHz transvaginal probes of SIEMENS X 300 USG MACHINE and Hitachi Aloka Arietta 70 High End USG Machines. These patients then underwent Magnetic resonance imaging (MRI) of abdomen and pelvis on 3T Siemens Verio MRI scanner using body coil (using T1, T2 , DWI, SWI and gradient sequences in various planes) and Computed Tomography (CT) on SOMATOM DEFINITION AS+ 128 SLICE HIGH END CT SCANNER with plain and post contrast venous phases.

OBSERVATIONS AND RESULTS

This was a retrospective study in 7 patients referred to the department of radiology in the tertiary care hospital , 4 of them being females and 3 being male.

Table 1: The female patients presented with following symptoms

Symptom	Frequency	Percentage
Pain in the lower abdomen	4	100
Dysmenorrhea	4	100
Infertility*	2	50

(* only two patients were married who presented with infertility)

Table 2: USG abdomen and pelvis and MRI abdomen of female patients reveals

Finding	Frequency	Percentage
Uterine didelphys	4	100
Obstructed hemivagina	4	100
Cervical canal stenosis	2	50
Hematometra	4	100
Hematocolpos	4	100
Ipsilateral renal agenesis	4	100

Table 3: The male patients presented with following symptoms

Symptom	frequency	Percentage
Lower abdominal pain	2	66.7
Dysuria	2	66.7
Non-specific complains*	1	33.3

*complains not related to abdomen or genito-urinary system such as acute febrile illness.

Table 4: USG abdomen and pelvis and MRI abdomen male patients reveals

Finding	Frequency	Percentage
Seminal vesicle cyst	3	100
Obstructed ejaculatory duct	3	100
Ipsilateral renal agenesis	3	100

RESULTS

It was found that all the patients with OHVIRA syndrome presented with lower abdominal pain and dysmenorrhea. In addition to the above symptoms , the married patient invariably presented with infertility.

All the patients invariably showed uterine didelphys with obstructed hemivagina resulting in hematometra and hematocolpos and with ipsilateral renal agenesis. 2 out of 3 patients with ZINNER syndrome present with lower abdominal pain and dysuria, while 1 patient presented with non-specific complains All the patients on imaging showed seminal vesicle cyst, dilated ejaculatory duct secondary to obstruction and ipsilateral renal agenesis.

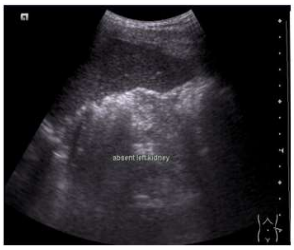
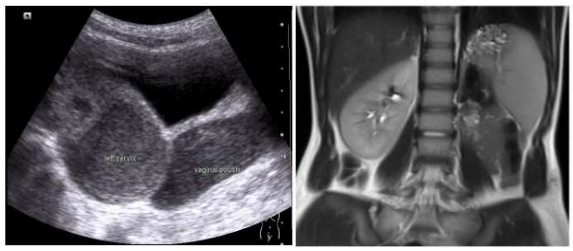


Figure 1



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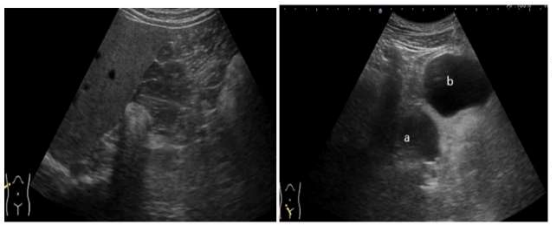

Figure 3

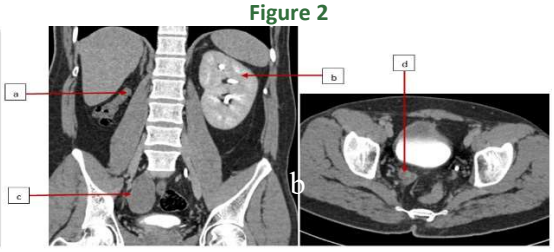

Figure 4

Figure 1: The two ultrasound images of 12 year old female presenting with lower abdominal pain. Image on left shows absent left kidney in left renal fossa. Image on the show left hemivagina, left hematocolpos and left uterine horn(hematometra). **Figure 2:** the MRI HASTE and T2 WI coronal images of the same female patient are shown here. The image on the left shows left renal agenesis. Image on the right shows uterine didelphys , normal right uterine horn (a) , left hematocolpos (b) , left hemivagina distended with blood products(c), right ovary(d) , left ovary(e). **Figure 3:** The two ultrasound images of 29 year old newly married male presenting with lower abdominal pain. Image on right shows absent right kidney in left renal fossa. Image on the right shows seminal vesicle cyst (a) and urinary bladder (b). **Figure 4:** CECT coronal and axial section of the same male patient is shown here. The image shows right renal agenesis(a) with compensatory hypertrophy of left kidney(b) and right seminal vesicle cyst(c) and tortuous right ejaculatory duct due to obstruction(d).

DISCUSSION

Both OHVIRA and ZINNER syndrome are mullerian duct developmental anomaly. OHVIRA is an obstructive mullerian anomaly caused by the lateral non-fusion of the mullerian ducts with asymmetrical obstruction. Renal agenesis most commonly occurs in association with uterine didelphys than with any other type of mullerian anomaly. The reported incidence of renal anomalies in this group is 20%.[9] OHVIRA is classically associated with uterus didelphys.[10,11] Obstructed unilateral vagina in patients with uterus didelphys is frequently associated with ipsilateral renal and ureter agenesis.

The classic triad of OHVIRA-

Unilateral low hemivaginal obstruction
Uterine didelphys
Ipsilateral renal agenesis

The OHVIRA syndrome is included in class III of the American Fertility Society classification (Table 1) of congenital uterine anomalies and often consists of uterine duplication, though septate uterus (Class V American Fertility Society classification) has been rarely associated.[12,13]

Table 1: American Fertility Society Classification of Female Genital Anomalies.[13]

Class	Name	Imaging Findings
Class I	Uterine hypoplasia or agenesis	Absent uterus or small uterus; lack of normal zonal anatomy
Class II	Unicornuate uterus	Single small banana-shaped uterus with a single fallopian tube; normal zonal anatomy; possible contralateral rudimentary horn
Class III	Uterus didelphys	Widely splayed, fully developed, uterine horns; fully developed unfused cervices; upper vagina may fuse and the septum may dissolve; vaginal septum is present in 75% cases
Class IV	Bicornuate uterus	Widely separated uterine horns; intercornual distance of >4 cm; concavity of the fundal contour or an external fundal cleft of >1 cm in depth; vaginal septum is present in 25% cases
Class V	Septate uterus	Convex, flat or minimally indented (1 cm); T2 hypointense (fibrous) or intermediate signal (muscular) septum
Class VI	Arcuate uterus	Normal variant; minimal indentation of the myometrium or fibrous component into the uterine cavity
Class VII	Diethylstilbestrol (DES)-related anomaly	Antenatal exposure to DES; T-shaped, hypoplastic uterus; narrow cavity; single cervix

Recently, HWW syndrome has been classified on the basis of the morphology of vagina (Table 2).[14]

Table 2: Classification of Herlyn-Werner-Wunderlich Syndrome Based on Vaginal Morphology.[14]

Classification	Subclass	Nomenclature	Findings	Clinical features
Class 1: Completely obstructed hemivagina	Class 1.1	Blind hemivagina	Completely obstructed hemivagina; uterus behind the septum is completely isolated from the contralateral uterus with no communication between the duplicated uterus and vagina	Hematocolpos; hematometra, hematosalpinx; hemoperitoneum; endometriosis; secondary pelvic adhesions; pyosalpinx; pyocolpos
	Class 1.2	Cervicovaginal atresia without communicating uteri	Completely obstructed hemivagina; cervix behind the septum is maldeveloped or atretic	Clinical features similar to the patients as in Class 1.1
Class 2: Incompletely obstructed hemivagina	Class 2.1	Partial reabsorption of the vaginal septum	Small communication exists between the 2 vaginas; uterus behind the septum is completely isolated from the contralateral uterus	Later age of onset, presents years after menarche; purulent or bloody vaginal discharge; can present with ascending genital infections
	Class 2.2	With communicating uteri	Small communication exists between the duplicated cervices	Menstrual blood drainage is still impeded

Class 1 consists of completely obstructed hemivagina with Class 1.1 including blind hemivagina and Class 1.2 including cervicovaginal atresia without communicating uteri. Our case possibly can represent variant of Class 1.2 with didelphus uterus and unilateral cervical atresia. Wolffian duct is located on both sides of the Müllerian duct and plays appears normal. important role in guiding fusion or resorption of wall between the Müllerian ducts. Ureteral bud arises from opening in urogenital sinus of Wolffian duct and any disruption in the development of these ducts can result in renal, uterine, and vaginal anomalies.

Clinical presentation:

Patients may present with lower abdominal pain, dysmenorrhoea, infertility, paravaginal mass, excessive foul smelling mucopurulent discharge and intermenstrual bleeding depending on the existence of uterine or vaginal communications.[15]

In patients with palpable abdominal, pelvic or vaginal mass (mucocolpos or pyocolpos), mullerian duct anomalies must be excluded.[16,17] The diagnosis of this rare mullerian anomaly,also known as Herlyn-Werner-Wunderlich Syndrome (HWWS) should be suspected in women who have ipsilateral renal agenesis with a pelvic mass.[18]

Diagnosis:

The role of imaging is to help detect, diagnose and distinguish surgically correctable forms of mullerian malformations from inoperable forms.[19] Abdominal or transvaginal ultrasound is cheap and noninvasive, whereas magnetic resonance imaging (MRI) is highly sensitive. The vaginal septum is difficult to visualize on ultrasound and is best shown on MRI.[20]

Complications:

Uterus didelphys has been associated with higher rate of infertility, spontaneous abortion, intrauterine growth retardation, preterm labor and postpartum bleed.[21] The obstruction of outflow tract may result in retrograde flow of the menstrual products and endometriosis. The superadded infection leads to pyometra and pyocolpos.

Treatment:

The treatment included vaginoplasty with or without metroplasty. Early diagnosis and treatment leads to restoration of fertility and prevention of complications and thus , positive impact on patients's life.

OSVIRA

Zinner's syndrome, first described in 1914 by Zinner, is a rare congenital malformation of the seminal vesicles and ipsilateral upper urinary tract.[22,23] An insult occurring before the 7th gestation week causes maldevelopment of the distal part of the mesonephric duct producing atresia of both; ejaculatory duct and the ureteric bud [25]. The close embryologic relationship between the genital and urinary tracts explains the developmental aberrations leading to this anomaly.

The Mesonephric or Wolffian duct is a paired structure. It forms the male reproductive system as well as the ureteric bud,. The orifice of the distal mesonephric duct and the ureteric bud separates between 6th-8th gestational week, and the ureteric orifice migrates toward the metanephric blastema, whereas the distal part of mesonephric duct forms the hemitrigone ,the bladder neck, the urethra up to the external sphincter, the seminal vesicle, vas deferens, ejaculatory ducts, epididymis, paradidymis, and appendix of the epididymis . This is under the influence of testosterone and anti-Mullerian hormone[26]. During the 4th to 6th week of gestation, the metanephric blastema secretes growth factors, which induce the growth of the ureteric bud toward it. The ureteric bud too secretes growth factors and proliferates, fusing with the metanephric blastema ,thus , inducing the blastema to become the primitive kidney on undergoing mesenchymal to epithelial transition[27]. Disturbance in any of these events of induction during the period of embryogenesis like mutation of metanephric blastema, or disruption of retinoic acid signaling, causes

inhibition of ureteric bud growth with failure of fusion of the ureteric bud with the metanephric blastema and renal agenesis or renal hypoplasia. Simultaneous failure of the ureteric bud to separate from the lower part of mesonephric duct leads to atresia of ejaculatory ducts and obstruction of the seminal vesicles with accumulation of secretions resulting in cystic dilatation[28].

The classic triad of OSVIRA-

Ipsilateral ejaculatory duct obstruction
Seminal vesicle cysts,
Ipsilateral renal agenesis

Presentation: The patients are usually diagnosed in the 2nd-3rd decade of life and present with symptoms of dysuria (37%), frequency (33%), perineal pain (29%), and epididymitis (27%) [24] or at times found incidentally on imaging for other causes.

Diagnosis: Diagnosis is suspected in patients with above symptoms and the triad is seen on abdominal USG and then confirmed with CT or MRI.

Complications

Ejaculatory duct obstruction leads to the characteristic symptoms of dysuria and causes infertility in this rare developmental anomaly of the male genitor-urinary system.

CONCLUSIONS

Early diagnosis of OHVIRA and OSVIRA syndromes with appropriate surgical intervention leads to restoration of fertility and decreases the long term morbidity. The rarity and unusual presentation of these anomalies may contribute to their diagnostic delay The key to early diagnosis and treatment is a high clinical suspicion on the enumerated complaints in the patient followed by an ultrasonography. Ipsilateral agenesis of kidney should prompt the suspicion of associated mullerian anomaly and vice versa. USG remains the first modality for imaging. It allows diagnosis in nearly all these cases when strict protocols with high level of suspicion are followed. High resolution USG improves the detailed structural evaluation. Absent kidney is a first marker to further evaluation. Trans-rectal USG helps in better visualization of the ejaculatory duct and the seminal vesicles. Usually the findings of USG are well documented and confirmed by MRI or CT. MRI allows easy and multi-planar imaging to delineate pelvic anatomy and the anatomical relations of various structures in all the planes that improves surgical understanding of the abnormality and allows various measurements to be taken. MRI images are more clinician friendly as USG images are not well understood by clinicians. The knowledge about these syndromes amongst radiologist leads to their early diagnosis and creates a positive impact on life.

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