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A case report on OHVIRA syndrome managed in Goa medical college

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Abstract
Herlyn-Werner-Wunderlich syndrome is an unusual congenital anomaly of the female genitourinary system, which is described as uterine didelphys with Obstructed Hemi-vagina and Ipsilateral Renal Anomaly (OHIRA), also known as OHVIRA syndrome. Mullerian duct anomalies have an incidence of 2–3%. While OHVIRA constitutes 0.16–10% of these Mullerian duct anomalies. Symptoms usually present shortly after menarche when hematocolpos develops during menstruation resulting in dysmenorrhea and a pelvic mass. The pelvic mass is the collection of blood products within the obstructed hemivagina. The first study in the diagnostic work-up is usually ultrasonography, which typically demonstrates a pelvic fluid collection which can simulate other disease processes thus confounding the diagnosis. MRI findings of the pelvis reveal a didelphis uterus. Imaging of the abdomen reveals agenesis of the ipsilateral kidney. MRI is beneficial in characterizing the didelphys uterus and vaginal septum for pre-operative planning.

Keywords: Herlyn-werner-wunderlich syndrome, uterus didelphys, renal agenesis, hemivagina, OHVIRA

Introduction
Herlyn-Werner-Wunderlich syndrome is a rare congenital anomaly of the female genitourinary system. It is also known as OHVIRA Syndrome. It derives its name as it is characterised by:

- Didelphys uterus with biscolis
- Obstructed hemivagina or/and hematocolpos
- Ipsilateral renal anomaly

It is a type of Mullerian duct anomalies and accounts for just 0.16-10% of mullerian duct anomalies. Mullerian duct anomalies themselves are a rare entity with incidence in general population being 2-3%. Renal agenesis is the most common renal anomaly in OHVIRA syndrome, although other malformations like renal duplication and multicystic dysplastic kidney, have also been reported [3]. This condition is usually diagnosed at or after puberty with cases presenting as abdominal pain or dysmenorrhea. They defied the conventional theory of urogenital development till recently when Acien theory of development of entire vagina from mesonephric ducts was postulated. The most common diagnostic modalities used are ultrasonography and MRI. USG can diagnose the collection inside uterus or vagina. However, USG fails to identify the type of Mullerian anomaly while MRI has multiplaner and tissue characterization ability giving precise information. Treatment of this condition aims to relieve the pain by surgically excising the septum thus draining the hematocolpos and/or hematometra. This also helps prevent endometriosis as it will stop the retrograde menstrual seeding. Patients have been known to have normal sexual life and also reported to have carried pregnancies and delivering at term.

Case report
We present a case of a 14-year-old girl presenting to the outpatient department of Obstetrics and Gynaecology.
She had attained menarche at the age of 13 years-10 months back at the time of presenting to the facility. She used to have delayed 2 monthly cycle with menstrual flow lasting anywhere between 3 to 4 days, and no history suggestive of menorrhagia. Patient had complaints of having severe lower abdominal pain in the region of right iliac fossa since last 3 months which had gradually increased over time. There was no diurnal variation or variation with food. Patient did not give any history of trauma.

However, patient claimed that pain increased during the menses. There was no history of having fever or any vaginal discharge.

On general examination patient was conscious, no abnormal features, thinly built, afebrile appeared anxious, pulse of 96 beats/minute, blood pressure measured in the right arm in recumbent position was 100/60 mm of Hg. Respiratory system and Cardiovascular system examination were within normal limit. On per abdomen examination the abdominal contour was normal, no scars no sinuses. Abdomen soft to palpate with tenderness in the right iliac fossa with vague mass felt in lower abdomen on the right side felt to be arising from pelvis, corresponding to the size of 12 weeks gravid uterus. There was however no rigidity or guarding of the abdomen. Local examination revealed no abnormality.

Patient was sent for ultrasonography after an opinion regarding right iliac fossa with the surgeon.

USG revealed uterine didelphys with right cavity showing echogenic mass filling up the entire right cavity with absent right kidney. Patient was taken up for MRI which showed uterine didelphys with biscollis. The right sided uterine cavity was filled with blood measuring upto 8.1 cm in length and 7.2 cm in the width and compressing the cervical canal of the left side with right sided hematometra with hematocolpos. Both ovaries appeared normal with no free fluid in the pelvis. The right kidney was absent and the left kidney showed mild hypertrophy.

Patient and relatives were explained about the rare condition that the girl was having called OHVIRA syndrome and its complications and treatment options and were asked to follow up with routine preoperative evaluation.

Investigations

- Hb 11.1 gm%
- TC 11,300
- DC 72/22/4/1/1
- Platelet count 2.6 lakhs
- BU 21
- SC 0.9
- HIV -NR
- HBsAg -NR

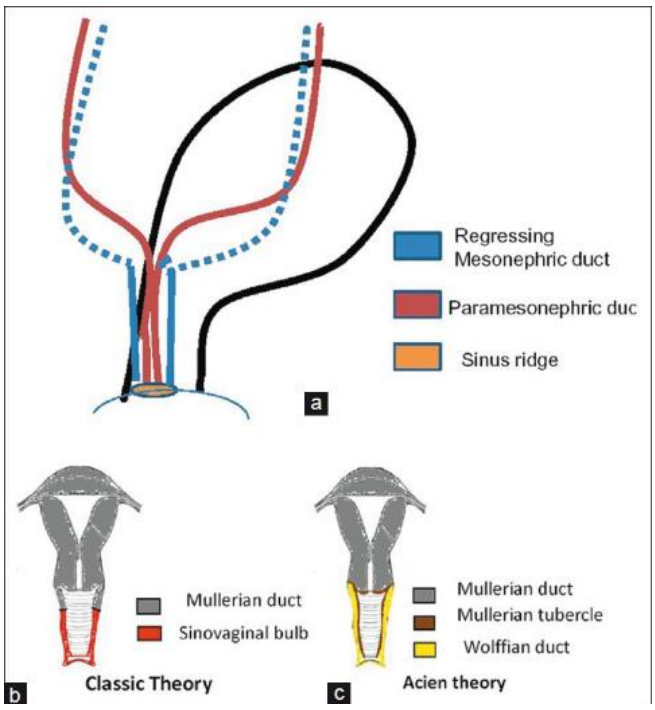
After getting a preoperative evaluation patient was admitted for the surgical intervention in view of her condition. Patient was taken in operating theatre and under spinal anaesthesia a cruciate incision was given on the vaginal septum on the right side to drain the collected blood. Then a pigtail catheter was passed for the drainage of the collected blood from the right sided uterine cavity. A part of collected blood was sent for culture and sensitivity. The catheter was left in place attached to a bag for drainage of the fluid. It was kept in situ for 7 days after the bag showed no further fluid getting collected. The catheter was then removed under strict aseptic precaution and then the ultrasonography repeated to see for the size. The right sided uterus had come back to the normal size. Patient felt symptomatically better and was advised to follow up for further management of the same. Patient is now planned for surgical removal of the septum in the second sitting.

Discussion

This syndrome of obstructed hemivagina and ipsilateral renal anomaly was first reported in 1922 and is known as Herlyn-Werner-Wunderlich syndrome or, more recently, by the acronym OHVIRA (Obstructed Hemivagina with Ipsilateral Renal Anomaly). The reported incidence in various case series is 0.1-3.8%.[4] Most commonly there is uterus didelphys although a few cases of single septate uterus have also been reported. Most common renal anomaly is renal agenesis. There have been reports mentioning cases with renal duplication and multicystic dysplastic kidney [3, 5].

Most commonly patient with this rare condition presents after menarche with nonspecific symptoms of recurrent pelvic pain or dysmenorrhea from progressive distension of the obstructed hemivagina. Mean median age of diagnosis of this condition is 14 years. Early and accurate diagnosis is important for prompt therapeutic intervention which can relieve symptoms, prevent further complications related to chronic obstructed hematocolpos (Such as endometriosis and pelvic adhesions), and also maintain reproductive capacity. This condition is difficult to diagnose and often remain undiagnosed in some cases even till mid-20s to early 30s. This delay in diagnosis can be attributed to the fact that being a rare entity there is lack of knowledge of the disease, delay in patient presenting to the OPD as there may be regular menstruations as the vaginal obstruction is incomplete and slow extension of hematocolpos.

By the end of the 20th century, it was thought that the embryology of female urogenital system was completely understood. While the uterocervical development was believed to occur from paired paramesonephric ducts; sinovaginal bulb from urogenital sinus was thought to develop into lower vagina; the kidneys and ureters were believed to arise from wolffian pronephros, mesonephros, metanephros. Aberration in vertical or horizontal fusion or arrest of paramesonephric ducts during course of development was explained various uterine anomalies. However, some complex uterine anomalies such as OHVIRA syndrome (Obstructed Hemi Vagina with Ipsilateral Renal Agenesis) could not be explained by the conventional theory of urogenital development and especially its coexistence with renal anomaly. This led to postulation of a new theory on development of vagina by Acien. Acien postulated that vagina develops from mesonephric ducts [6].



This image [7] shows embryological development of female reproductive system. (Fig a)

Fig b shows classic theory where uterus, cervix, fallopian tubes

along with upper 1/3rd of vagina develop from Mullerian ducts while the lower 2/3rd of the vagina develop from sinovaginal bulb.

Fig C shows Acien's theory of development of vagina which states that uterus, cervix and fallopian tubes develop from Mullerian ducts while vagina develops from Mesonephric (Wolffian duct).

Acien theory states that as mentioned in the classic theory the development of uterus with fallopian tubes and cervix occur from paramesonephric or Mullerian ducts, the entire vagina develops from mesonephric or Wolffian ducts. This precisely explains its coexistence with renal anomalies. Acien has also postulated that only the vaginal lining is of Mullerian origin as it develops from Mullerian tubercle.

Although ultrasonography is the most common imaging modality used in gynecology and it detects the presence of uterine didelphys but it can be difficult to differentiate a hematometra with thin, stretched myometrium from hemorrhagic adnexal masses. MRI with its multi planar capability, superior tissue characterization, and large field of view is the modality of choice in this condition. MR imaging can accurately depict the

- Uterine contour and the shape of uterine cavity,
- Associated cervical and vaginal anomalies,
- Provides tissue characterization of the septa,
- Characteristics of the contents of obstructed blood, and
- Detect any coexisting renal and/or urethral abnormalities.

Primary management of the OHVIRA syndrome is requires surgical intervention like of excision of vaginal septum to relieve obstruction. Historically, surgeons advocated a two-stage procedure, with the initial surgery to reduce the hematocolpos and the second to re-sect the excess septum after a period of wound-healing and vaginal remodelling.

However new trend is to do both in the single stage vaginoplasty with complete resection of the septum which not only relieves symptoms but also prevents long term complications like endometriosis and even improves sexual functions and conception chances.

Conclusion

Ohvira syndrome is a rare congenital urogenital anomaly which may present with wide range of clinical presentation. It is often difficult to diagnose without correct modality. MRI plays a major role in diagnosing the same. Though rare and difficult to diagnose has simple surgical management giving good results.

Appropriate surgery is a single stage procedure to either excise or completely divide the obstructing septum. Early correct diagnosis is required to relieve the symptoms and prevent complications, caused by retrograde menstruation which may result in endometriosis and, also, preserve sexual and conception abilities.

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