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OHVIRA syndrome (Herlyn Werner Wunderlich syndrome): An unusual delayed presentation!

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Abstract
Uterine malformations results from the defective fusion of Mullerian ducts during the development of the female genital tract. The least common form of this malformation is OHVIRA syndrome, a very rare congenital anomaly of the urogenital tract involving both Müllerian ducts and Wolffian structures. It is characterized by a triad of uterine didelphys, ipsilateral renal agenesis and obstructed hemivagina. There is a wide variety of phenotypic presentations recognized as a spectrum of disease rather than a separate entity. It usually presents after menarche but may have delayed presentation depending upon the type.
Case report: A multiparous woman with previous two cesarean deliveries presented with complaints of pelvic pain, urinary retention and hematuria and an ultrasound report suggestive of bicornuate uterus with large hematocolpos /hematometra. On Examination under anesthesia –a huge cystic bulge on the anterior vaginal wall was seen. On MRI, bicornuate bicollis uterine anatomy was seen with right hematotrachelos. IVP was suggestive of absent right kidney. Laparotomy with the Right hemihysterectomy along with drainage of hematocolpos was done.

Keywords: Herlyn-Werner-Wunderlich syndrome, OHVIRA syndrome, uterine didelphys, obstructed hemivagina, ipsilateral renal agenesis, Right hemihysterectomy

Introduction
The Herlyn-Werner Wunderlich syndrome (uterine didelphys with hematocolpos and ipsilateral renal agenesis) was initially illustrated by Herlyn and Werner in 1971 [1]. Wunderlich in 1976 depicted the association of renal agenesis with a uterine didelphys and obstructed vagina [2].
Herlyn Werner Wunderlich syndrome is a rare congenital uterine anomaly that presents at early pubertal age or menarche, making it uncommon to suspect this syndrome at late reproductive age. A high index of suspicion is required for the diagnosis of this syndrome. When untreated, this may result in severe complications such as pyohematocolpos, pyosalpinx, or pelvic peritonitis, urinary obstruction, and long-term complications, such as endometriosis, pelvic adhesions, and increased risk of abortion or infertility. It is usually observed in post menarche adolescent girls and young women with dysmenorrhea, abdominal pain, irregular menses and pelvic mass [3]. Magnetic resonance imaging (MRI) is considered the noninvasive diagnostic modality of choice for Mullerian anomalies and associated renal abnormalities [4]. The surgical approach depends on the clinical complications, imaging findings, and associated complications.

Case report
Mrs. X, 31 yrs multiparous female, with previous two cesarean deliveries reported in OPD with chief complaints of acute urinary retention since one day, pain in lower abdomen and hematuria since two months. The patient also had an ultrasound report suggestive of a bicornuate uterus with large hematocolpos/hematometra. There was no history of fever, pelvic pain, dysmenorrhea, or dyspareunia. The patient attained her menarche at the age of 16 years and had regular menstruation cycle with no dysmenorrhea.

Investigation
The patient was catheterized to relieve urinary complaints. Her vitals were stable. Her abdomen was soft but tender without any guarding or rigidity. Examination under anesthesia was performed–a huge cystic bulge was seen on the anterior vaginal wall. Cervical os was seen posterior to bulge which was non-negotiable. On aspirating the bulge, 100cc of blood mixed organized collection was obtained. The collection showed no growth. On Cystoscopy, the bladder wall was normal.

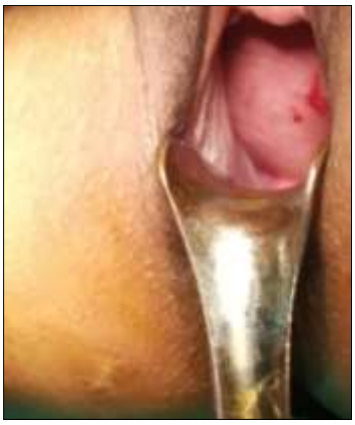

Fig 1: Cystic bulge is seen on the anterior vaginal wall on per speculum examination under anesthesia

On MRI, a large collection in the right cervix or hematotrachelos and two large horns with two separate Cervices and a single vagina was observed suggestive of bicornuate bicollis uterus. IVP showed normal functioning left-sided kidney and ureter with no contrast uptake on the right side even after 24 hours of injection of contrast suggestive of right renal agenesis.

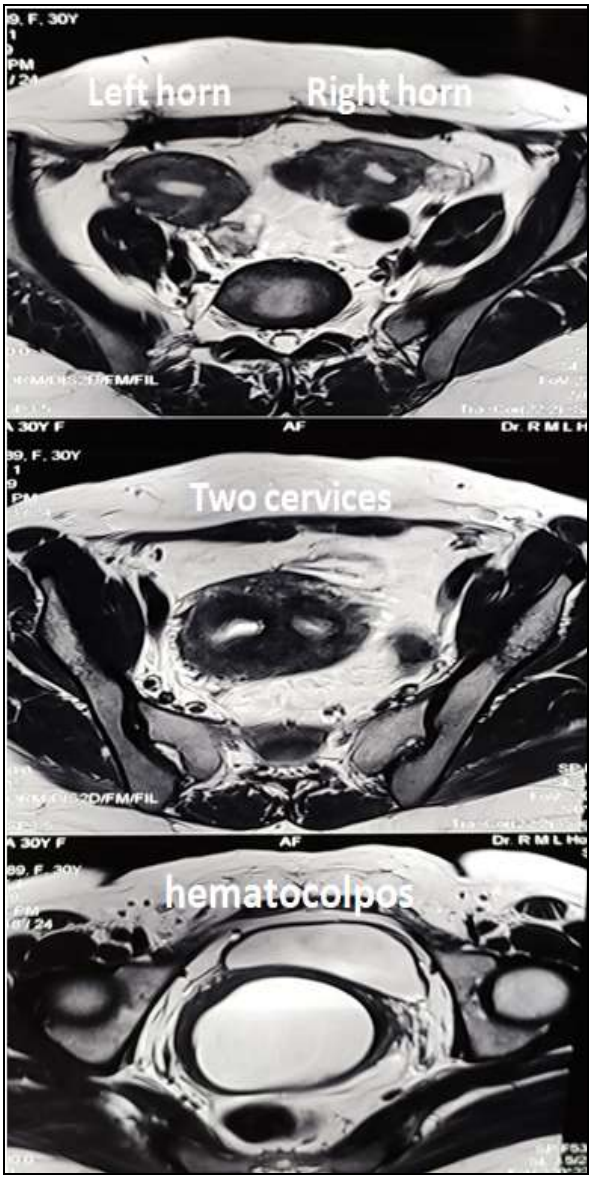

Fig 2: MRI showing two separate uterine horns (upper), two cervices (middle), hematocolpos (lower)

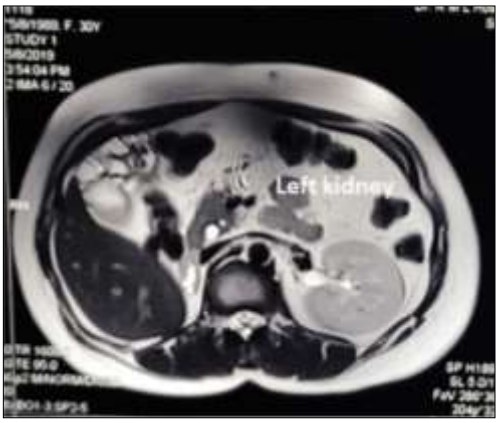

Fig 3: MRI image showing right renal agenesis

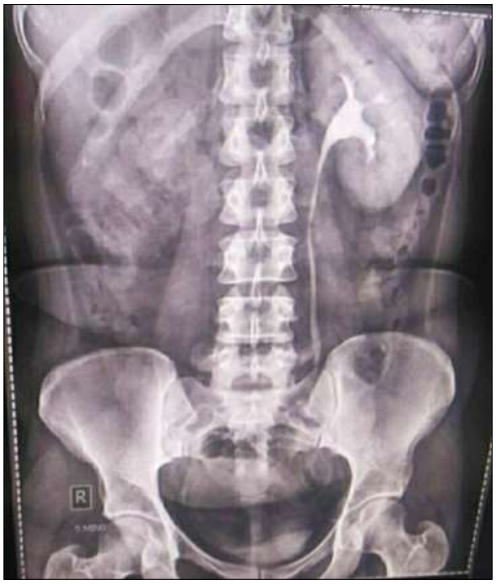

Fig 4: Intravenous pyelography showing solitary normal functioning left kidney

Treatment

The patient was planned for diagnostic laparoscopy. On laparoscopy, the bicornuate uterus was visualized with extensive adhesions. Surgery was then converted to laparotomy due to extensive adhesions between the omentum, anterior abdominal wall, and uterus. The abdominopelvic route was adopted to see the relation of the vaginal bulge with the uterus. A nick was given from the vaginal end on the bulge, from which frank ous was drained, suggestive of secondary infection of collected blood. Uterine cavity was opened, hegar's dilator introduced as a probe through the cavity. The probe was felt through the vaginal end indicating continuity of cavity with hematocolpos. As the patient's family was complete, there was no desire for future fertility, and to prevent secondary complications such as pus collection, endometriosis and recurrence, decision for hemihysterectomy was taken. The vaginal septum was excised to relieve the obstruction.

Follow up

In her subsequent visits, the patient remained asymptomatic. Ultrasound showed no collection. Vaginoscopy was done which showed two separate cervical os without any collection.

Discussion

Uterine anomalies have been reported in 0.1–2% of women and 4% in women with infertility. The uninhibited use of

hysterosalpingography, diagnostic hysteroscopy, and the routine practice of transvaginal ultrasound scanning (TVS) and three-dimensional transvaginal ultrasound (TDU) have led to an noticeable increase in the prevalence of uterine anomalies and, the figures cited previously could be even higher [5].

The uterine didelphys is associated in 15%–30% of cases with unilateral anomalies, i.e., obstructed hemivagina and ipsilateral renal agenesis [6, 7, 8]. Mesonephric duct (Wolffian duct) and para-mesonephric or Mullerian ducts are the two paired uro-genital structures which are derived from internal genital organs and the lower urinary tract. The exact etiology of OHVIRA syndrome is still unknown. The etiologic factors may include an insult during the developmental period or due to any polygenic, environmental or multifactorial inheritance [9]. A bicornuate uterus is due to an embryologic arrest occurring during the 8th week period of gestation, which eventually affects the mullerian and metanephric duct [10].

If one of the Wolffian ducts fails to develop, the ipsilateral kidney and ureter will not fuse in the midline. This mechanism of non-fusion can be partial or complete. Uterine didelphys is formed due to the non-fusion of both mullerian ducts. The duct on the affected side lacking the Wolffian duct displaces itself sideways and does not come into direct contact with the urogenital sinus in the centre resulting in the formation of a blind sac or obstructed vagina. The distal one third of the vagina derived from the urogenital sinus is usually not affected.

As a hemivagina is obstructed due to the septum, outflow of menstruation from that side is not possible, resulting in hematocolpos, rarely hematometra, and hematosalpinx. Furthermore, blood reflux into the abdominal cavity may result in endometriosis further complicated by pelvic adhesions, increased risk of abortion or infertility. Patients with OHVIRA syndrome usually present post menarche with dysmenorrhea, pelvic pain, or pelvic mass [11].

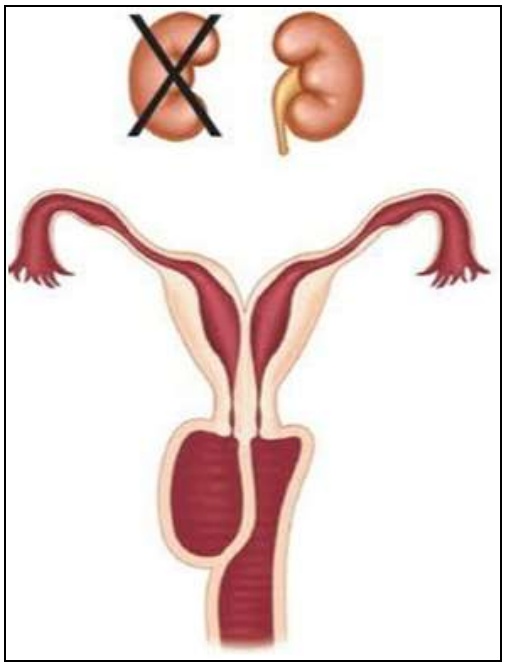

Fig 5: Pictorial presentation of OHVIRA or Herlyn-Werner-Wunderlich (HWW) syndrome showing uterine didelphys, blind hemivagina, and ipsilateral renal agenesis

Magnetic resonance imaging (MRI) is considered the noninvasive diagnostic modality of choice for the Mullerian anomalies with the advantages of no radiation exposure, multiplanar imaging with better soft-tissue resolution, better anatomic delineation of pelvic structures, and higher sensitivity for diagnosing complications such as endometriosis [2, 12].

Delayed presentation of OHVIRA syndrome is a very rare entity. Literature is very limited for these presentations. In a retrospective case series study by A. Kriplani *et al*. in 2019, a 41 yrs female with severe dysmenorrhea and previous cesarean delivery was intraoperatively diagnosed to have uterine didelphys with hematocolpos with obstructed hemivagina following which the patient underwent total laparoscopic hysterectomy with excision of vaginal septum [13].

The treatment of choice in these cases is surgical excision of the vaginal septum to relieve the obstruction and prevent the development of further complications.

Conclusion

The most common presentation of OHVIRA syndrome is pubertal or early onset dysmenorrhea that or may not be accompanied by other complication as mentioned earlier. The unusual delayed presentation of OHVIRA syndrome is one of the diverse presentation. Here, major concern was to relieve obstruction and prevent recurrence by hemi-hysterectomy rather than preserving the uterus which would have been the preferred option in most cases as they present early in life.

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Conflict of Interest

The authors have no conflict of interest to declare that is relevant to the content of this article.

Informed Consent

The patient's informed consent for publication of this report was obtained.

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