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Case Report

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OHVIRA syndrome - A rare Müllerian abnormality

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Abstract

Obstructed Hemivagina and Ipsilateral Renal Agenesis (OHVIRA) syndrome is an uncommon congenital urogenital anomaly resulting from abnormal embryological development of the Müllerian and Wolffian ducts. This syndrome is characterised by a triad of a) Obstructed hemivagina, b) Ipsilateral renal agenesis, and c) Uterus didelphys. A 21-year-old female presented with heavy menstrual bleeding and dysmenorrhoea. Pelvic ultrasound revealed uterine anomalies, and magnetic resonance imaging (MRI) confirmed the diagnosis of OHVIRA syndrome, marking the first reported case at the RIPAS Hospital. A review of the literature highlights the importance of a high index of suspicion among clinicians—including paediatricians, gynaecologists, radiologists, and emergency medicine specialists—to facilitate early and accurate diagnosis. Timely diagnosis and intervention can lead to significant symptom relief, preserve reproductive function, and prevent major complications.

Keywords: Absent kidney, Heamatocolpos, Uterus didelphys

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INTRODUCTION

Congenital malformations of female genital tract are defined as deviations from normal anatomy resulting from embyological maldevelopmemt of the Müllerian or paramesonephric duct. They represent a rather benign condition with prevalence of 4-7%. Depending on the degree and type of anatomical distortion, they affect health and reproductive outcomes. Obstructed HemiVagina and Ipsilateral Renal Agenesis (OHVIRA)

syndrome is an extremely rare abnormality characterised by a triad of obstructed hemi vagina (OHV), ipsilateral renal anomaly (IRA**)** and uterus didelphys (**Figure 1**). It was first reported in 1922 and subsequently described in detail by Herlyn, Werner, and Wunderlich, after whom it was initially named the Herlyn–Werner–Wunderlich Syndrome.[1] Associated urological anomalies include ectopic insertion of ureter into the vagina in 50% of cases, ureterocoele and vesicoureteral reflux on the ipsilateral side, dysplastic kidney on the ipsilateral side, and there can be megaureter and vesicoureteric reflex on the contralateral side.[2]

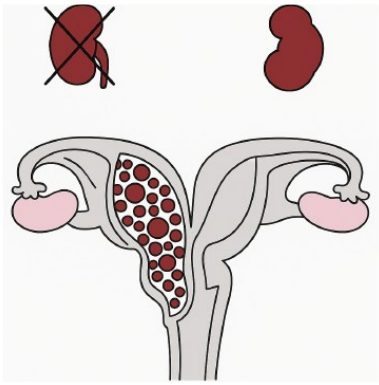

Figure 1: Graphical representation of the triad: double uterus (uterus didelphys), unilateral obstructed or blind hemivagina and ipsilateral renal agenesis (Illustration reproduced by R Ara).

CASE REPORT

A 21-year-old single female student was referred from the Emergency Department for admission due to heavy menstrual bleeding, accompanied by vomiting and epigastric pain persisting for four days. She had attained menarche at 12 years with regular menstrual cycles; however, her periods were frequently prolonged and painful, necessitating regular analgesia and frequent school absences. Her medical and surgical history was otherwise unremarkable, and her dietary intake was normal.

On examination, her abdomen was soft and non-tender, with no palpable masses. Genital inspection revealed an intact hymen without any mass protruding from the vagina. Pelvic ultrasound demonstrated a bicornuate uterus with two uterine horns; the left uterine horn had an endometrial thickness of 12.8 mm, while the right measured 18.3 mm. Additionally, a cystic area was noted at the cervical region. Her ovaries appeared normal, and no other pelvic pathology was observed. These findings raised suspicion for a Müllerian duct anomaly.

Upon admission, her serum haemoglobin level was 6.9 g/dL (normal range: 12.4–15.5 g/dL). She received two units of packed red blood cells, increasing her haemoglobin to 10 g/dL. A trial of tranexamic acid and mefenamic acid was administered.

Magnetic resonance imaging (MRI) of the abdomen and pelvis, and this confirmed the presence of a complete bicornuate uterus with a longitudinal vaginal septum. In addition, there was a non-enhancing structure with fluid-filled level which was seen in the vaginal canal measuring 4.2 x 4.0 x 2.2cm, suggestive of a haematocolpos. Both ovaries were normal. There was also absence of the right kidney likely associated with underlying the Mullerian duct anomaly. These findings

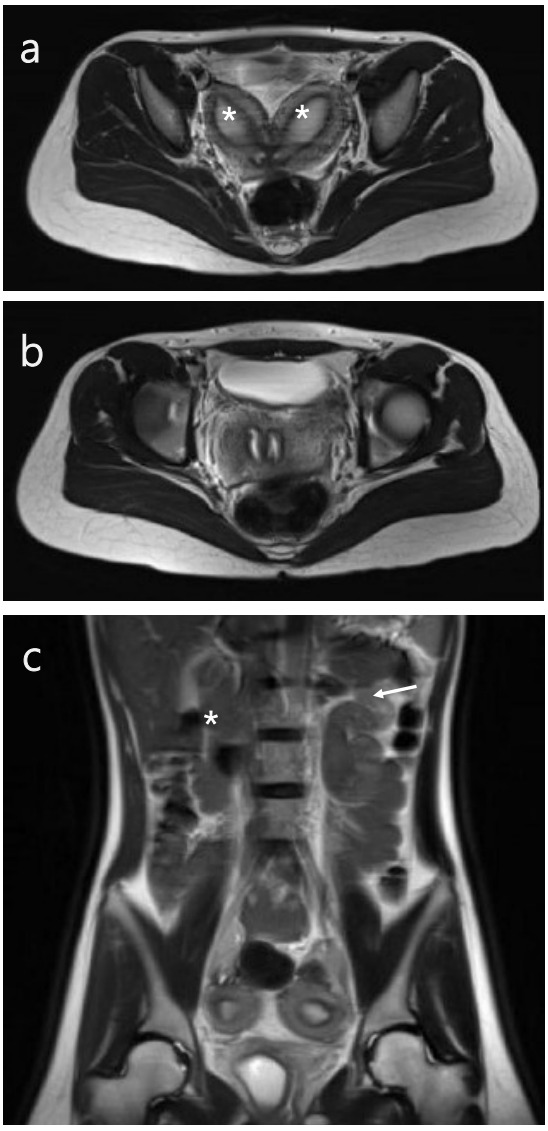

Figure 2: Graphical representation of the triad: a) double uterus (uterus didelphys *), b) unilateral obstructed or blind hemivagina and c) ipsilateral right renal agenesis (*) and a normal left kidney.

were in keeping with the OHVIRA syndrome. Based on the ESHRE/ESGE classification of female genital malformation the patients abnormality was graded as U3bC2V3 (**Table 1** – ***Refer to the Supplementary Text***).

The patient was seen in the Gynaecology clinic, counseled on her diagnosis, and started on tranexamic acid to control the bleeding. She was also advised on the need to excise the longitudinal vaginal septum and drain the haematocolpos.

Two months later, she underwent an examination under anaesthesia and hysteroscopy. During the procedure, it was noted that she had one cervix and one endometrial cavity on the left side, with a single ostium. On speculum examination, no obvious vaginal septum was observed, and there was no bulging or bluish discolouration of the vaginal mucosa on the right side where haematocolpos had been suspected. Slight hardening was felt in the right cul-de-sac compared to the left, but no definite mass was palpable. It was suspected that the septum was located higher up and the haematocolpos was deeper, making a vaginal approach unfeasible.

She was informed that a laparoscopic procedure, with or without laparotomy, would be necessary to excise the septum from above and release the haematocolpos. The laparoscopy would also allow a clearer view of her pelvic anatomy. This was explained to her postoperatively, but she declined further surgery at that time.

Six months later, she returned for follow-up, during which an ultrasound scan showed the haematocolpos remained stable in size at 4 cm, and she reported no pain. She was again advised that surgical intervention via laparoscopy or laparotomy would be required to release the haematocolpos and excise the septum; however, she remains reluctant to proceed with surgery. Currently, her haemoglobin levels are stable, and she uses tranexamic acid only during menstruation along with paracetamol for pain relief.

Additionally, she was referred to the nephrology service due to her renal agenesis. Her renal function is normal, and she was advised to avoid non-steroidal anti -inflammatory drugs, nephrotoxic medications, and herbal remedies. She will continue renal follow-up for monitoring.

Regarding her gynaecological condition, it was recommended that surgical intervention be reconsidered if her pain worsens or the size of the haematocolpos increases.

DISCUSSION

Normal development of the female reproductive tract is based on the proper formation, differentiation, and fusion of the Müllerian (paramesonephric) ducts. These ducts form as an invagination of the coelomic epithelium. In genetic female embryos (46, XX), the absence of Anti-Müllerian Hormone secretion allows these ducts to develop into the oviducts, uterus, and the upper two-thirds of the vagina. At around six weeks of gestation, the Müllerian ducts form and begin canalisation. Between seven and nine weeks, the caudal parts of the ducts fuse, followed by absorption of the midline septum and formation of a single uterine cavity between nine and thirteen weeks. The vagina forms through fusion of the cavity derived from the Müllerian ducts with the sinovaginal bulb, which progresses upward and fuses with the caudal Müllerian duct cavity to form the vaginal lumen. Failure or abnormal fusion of the Müllerian ducts can lead to anomalies such as a bicorporeal (bicornuate) uterus.[3]

The typical patient with OHVIRA syndrome presents after menarche with nonspecific symptoms, most commonly recurrent pelvic pain or dysmenorrhoea caused by progressive distension of the obstructed hemivagina. Other symptoms can include abdominal swelling, nausea, and vomiting during menstruation. In rare cases, symptoms may present shortly after birth as vaginal discharge. The median age of diagnosis is approximately 14 years, with a range from 12 to 29 years. Over time, blood collects in the obstructed hemivagina (haematocolpos) and can lead to haematometra (blood accumulation in the uterus) and haematosalpinx (blood in the fallopian tubes). This collected blood is susceptible to infection, which can result in pelvic adhesions.[4]

Diagnosis is usually made via ultrasound or MRI. Typical imaging findings include duplication of the uterus, cervix, and vagina (**Figure 2a**), unilateral haematocolpos or haematometra (**Figure 2b**), and congenital renal abnormalities on the same side as the obstruction, most commonly renal agenesis, but also occasionally multicystic dysplastic kidney or renal duplication (**Figure 2c**). Other associated findings can include Gartner's duct cysts and pelvic endometriosis.

MRI has advantages as it can more accurately differentiate haematometra with thin, stretched myometrium from haemorrhagic adnexal masses, and better visualize uterine contours, the shape of the uterine cavity, and cervical and vaginal abnormalities. This is crucial for classification of Müllerian duct anomalies and surgical planning. MRI also provides tissue characterisation of

septa and can distinguish the contents of obstructed cavities (fluid vs. blood), as well as detect coexisting renal or urethral abnormalities.[10]

Treatment focuses on relieving symptoms through medication and surgery and is best managed in a tertiary centre with multidisciplinary expertise. Surgical management typically involves removal of the obstructing vaginal septum. The classic approach is a two-stage vaginoplasty: the first stage involves drainage of haematometra, followed by a second surgery to resect the septum. Untreated cases can develop complications such as chronic pain, pelvic infections, pelvic adhesions, endometriosis, infertility, and menstrual irregularities.[4,5,8]

Early diagnosis and treatment can lead to complete symptom improvement, adequate reproductive prognosis, and help avoid major complications like endometriosis, pelvic adhesions, and infertility.[6] Successful pregnancies have been reported in women with OHVIRA syndrome; however, there is an increased risk of miscarriage, intrauterine growth restriction, spontaneous preterm labour, malpresentation, and foetal distress. Variations in uterine and cervical anatomy increase the likelihood of caesarean delivery and the need for cervical sutures. Consequently, pregnancies in these women are considered high-risk, partly due to altered renal function.[7]

Take Home Message

- ·OHVIRA syndrome is a rare congenital developmental abnormality of the Müllerian and Wolffian ducts, characterised by a triad of a) obstructed hemivagina, b) ipsilateral renal agenesis, and c) uterus didelphys.
- It manifests with recurrent pelvic pain and dysmenorrhoea caused by progressive distention of obstructed hemivagina (haematocolpos).
- Awareness, recognition and early diagnosis are important for symptoms resolution, improving reproductive outcomes and renal prognoses.

Abbreviations

OHVIRA	Obstructed Hemivagina and Ipsilateral Renal Agenesis
MRI	Magnetic resonance imaging

Declarations

Conflict of interests

The authors declare no conflict of interests.

Consent

Consent has been obtained from patient's parents for publication.

Acknowledgement

None.

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Table I: ESHRE/ESGE (European Society of Human Reproduction and Embryology/European Society for Gynaecological Endoscopy) consensus classification of female genital malformation. [1]

	Main class Uterine anomaly	Main sub-class	Co-existent sub-class Cervical/vaginal anomaly
U0	Normal uterus		*Cervix* C0: Normal cervix C1: Septate cervix C2: Double 'normal' cervix C3: Unilateral aplasia/dysplasia C4: Aplasia *Vagina* V0: Normal vagina V1: Longitudinal non-obstructing vaginal septum V2: Longitudinal obstructing vaginal septum V3: Transverse vaginal septum and/or imperforate hymen V4: Vagina aplasia
U1	Dysmorphic uterus	a. T-shaped b. Infantilis c. Others	
U2	Septate uterus	a. Partial b. Complete	
U3	Bicorporeal uterus	a. Partial b. Complete c. Bicorporeal septate	
U4	Hemi-uterus	a. Rudimentary horn with cavity (communicating or not) b. Rudimentary horn without cavity/aplasia (no horn)	
U5	Aplastic	a. Rudimentary horn with cavity (bi- or unilateral) b. Rudimentary horn without cavity (bi-or unilateral)/aplasia	
U6	Unclassified Malformations		

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