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A rare case report of urogenital anomaly in a teenage girl: Herlyn–Werner–Wunderlich syndrome/OHVIRA syndrome (Obstructed Hemivagina and Ipsilateral Renal Anomaly)

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Abstract

Background Herlyn–Werner–Wunderlich syndrome also known as OHVIRA syndrome is a rare complex congenital developmental anomaly characterized by the triad of uterine didelphys, obstructed hemivagina causing hematometrocolpos and ipsilateral renal anomaly.

Case presentation Here we report a case of a 14-year-old girl who presented with acute onset lower abdominal pain. Ultrasound and magnetic resonance imaging showed uterus didelphys, hematometrocolpos, obstructed hemivagina and right renal agenesis. Patient underwent hematocolpos drainage.

Conclusions OHVIRA syndrome is an uncommon congenital anomaly. Imaging plays a major role in diagnosis. Surgery is the treatment of choice to resect the septum and relieve the obstruction. An early correct diagnosis is the goal to relieve the symptoms and prevent complications, caused by retrograde menstruation which may result in endometriosis and, also, preserve sexual and conception abilities.

Keywords Herlyn–Werner–Wunderlich syndrome, OHVIRA—obstructed hemivagina, ipsilateral renal agenesis, Uterine didelphys

Background

OHVIRA syndrome is characterized by uterine didelphys, obstructed hemivagina causing hematometrocolpos and ipsilateral renal anomaly. This triad is known as "Herlyn–Werner–Wunderlich syndrome," and it was first described by Purslow [1]. It is a rare obstructed Mullerian duct anomaly. The incidence of Mullerian duct anomalies ranges from 0.8 to 4%, and the incidence of OHVIRA syndrome is estimated to be 1/20,0000 in the general population [2, 3]. Review of literature in 15 reported studies consisting of 1771 patients during 1965–1996 period revealed mean incidence of uterine didelphys to be 11.1% [4]. Patients with didelphic uterus, 29.1% had associated renal anomalies with 23.6% diagnosed to have renal agenesis and obstructed hemivagina [5]. It usually presents in a post-pubertal adolescent female after menarche with nonspecific symptoms like lower abdominal pain, urinary retention, pelvic pain and a mass.

In this study, we report a case of a 14-year-old girl from India who presented with acute onset lower abdominal pain. Abdominal ultrasound (US), magnetic resonance imaging (MRI) and intraoperative findings are reported in this study.

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Case presentation

A 14-year-old female presented with abdominal pain since 5 days, which was sudden in onset, gradually progressive, non-radiating, intermittent and relieved on medication. The patient attained menarche at 12 years of age. Menstrual history: 5/30, regular, normal flow with presence of clots and dysmenorrhea, 3 pads/day. General examination was unremarkable. Secondary sexual characters were attained. On examination per abdomen was soft and non-tender; local examination showed no bulge. On per rectal examination posterior vaginal wall bulge was felt.

Ultrasound abdomen and pelvis showed absence of right kidney in right renal fossa (Fig. 1a) and compensatory hypertrophy of contralateral left kidney measuring 133×55 mm (Fig. 1b). Evidence of two uterine horns (Fig. 2a) and two uterine cavities noted measuring 57×25 mm on the right side (Fig. 2b) and 64×22 mm on the left side (Fig. 2c)—uterine didelphys. The right horn of uterus and cervical canal are seen extending into a large well-defined hypoechoic collection with internal echoes measuring approximately 100×75 mm suggestive of hematometrocolpos (Fig. 3). The left horn of uterus and cervical canal appear collapsed. No gross adnexal pathology is seen. Both ovaries are visualized normally (Fig. 4).

MRI pelvis was done which revealed complete duplication of uterine horns as well as duplication of the cervix and vagina (Fig. 5). The right hemivagina was distended by T1 hyper intense, T2 hypo intense collection measuring 74×74×95 mm with low-lying debris displacing the left hemivagina, rectum to the left and compression of the urinary bladder anteriorly—likely obstructed right hemivagina with subacute blood secondary to longitudinal vaginal septum (Fig. 6). The left hemivagina appeared collapsed. No communication between the duplicated uteri and vagina could be established. Bilateral ovaries were normal. Right kidney is absent in right renal fossa. Left kidney showed compensatory hypertrophy (Fig. 7). Additionally, a T1 hyperintense cystic structure with T2 shading was noted in the right adnexa inserting into the upper vagina—likely a blind right ectopic ureter with hematic reflux from hematocolpos (Fig. 8). All these features are suggestive of OHVIRA syndrome.

The patient underwent drainage of the hematocolpos. Around 150 ml of chocolate colored fluid was drained. The Intraoperative findings are shown in Fig. 9. Postdrainage ultrasound 2 months later showed minimal fluid in the lower uterine cavity (Fig. 10).

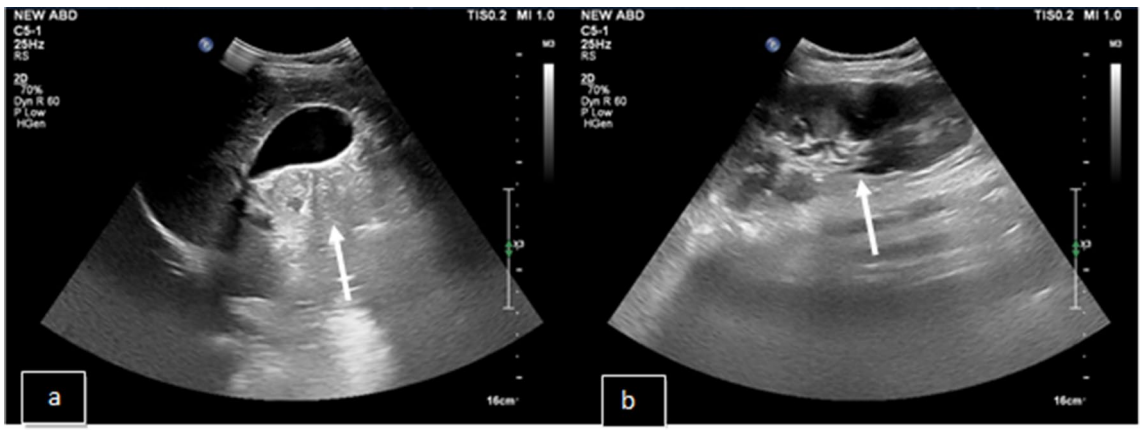

Fig. 1 **a** and **b** USG abdomen and pelvis of the patient showing absent right kidney in the right renal fossa (arrow in **a**) and compensatory hypertrophy of the contralateral left kidney (arrow in **b**)

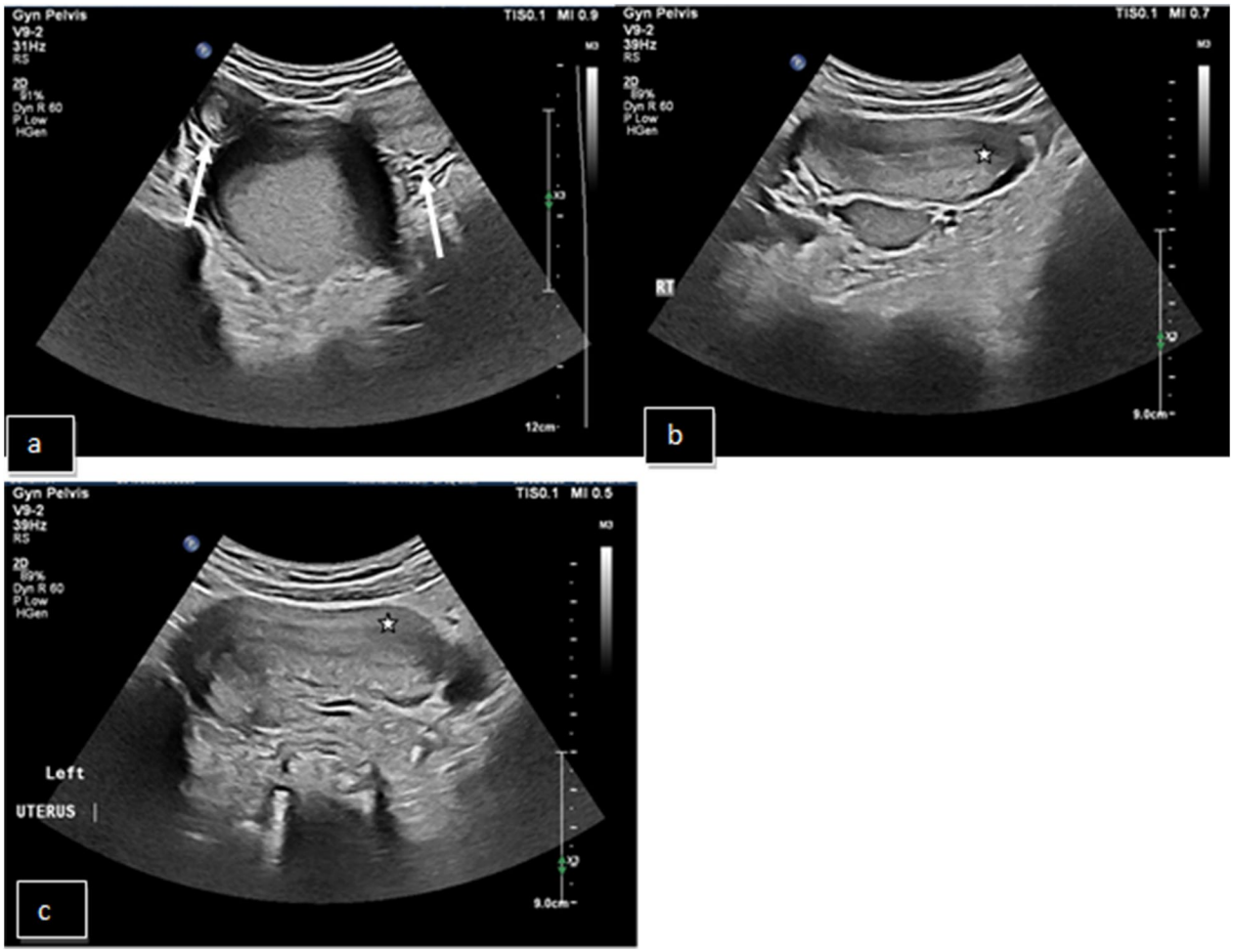

Fig. 2 **a**–**c** USG abdomen and pelvis showing two uterine horns (arrows in **a**). Right uterine cavity (star in **b**) and left uterine cavity (star in **c**)

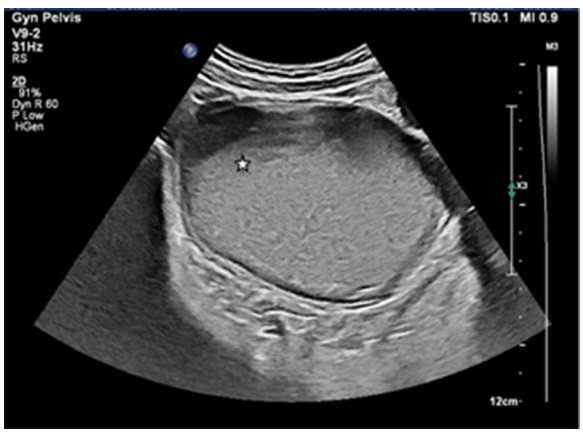

Fig. 3 USG abdomen and pelvis. The right uterine horn and cervical canal are seen extending into a large well-defined hypoechoic collection with internal echoes (**star**) measuring approximately 100 × 75—s/o hematometrocolpos

Discussion

OHVIRA syndrome is often misdiagnosed. The patient generally presents at puberty with pelvic pain. In the adulthood, they present with primary infertility, hydrometrocolpos, ischiorectal swelling and urinary obstruction. There can be a delay in diagnosis due to normal onset of puberty and menstruation [6].

Herlyn–Werner–Wunderlich syndrome (HWWS) is a combination of type III Mullerian anomaly with mesonephric duct anomaly with vaginal septum. Renal anomalies can occur in 50% of patients with Mullerian anomalies. The classic renal manifestation of OHVIRA syndrome is ipsilateral renal agenesis, but cases of dysplastic kidneys, duplicated kidneys, recto vesical bands, or crossed fused ectopia and ectopic ureters [7] should be looked for. Contralateral renal anomalies have been reported in up to 50% of the cases [8]

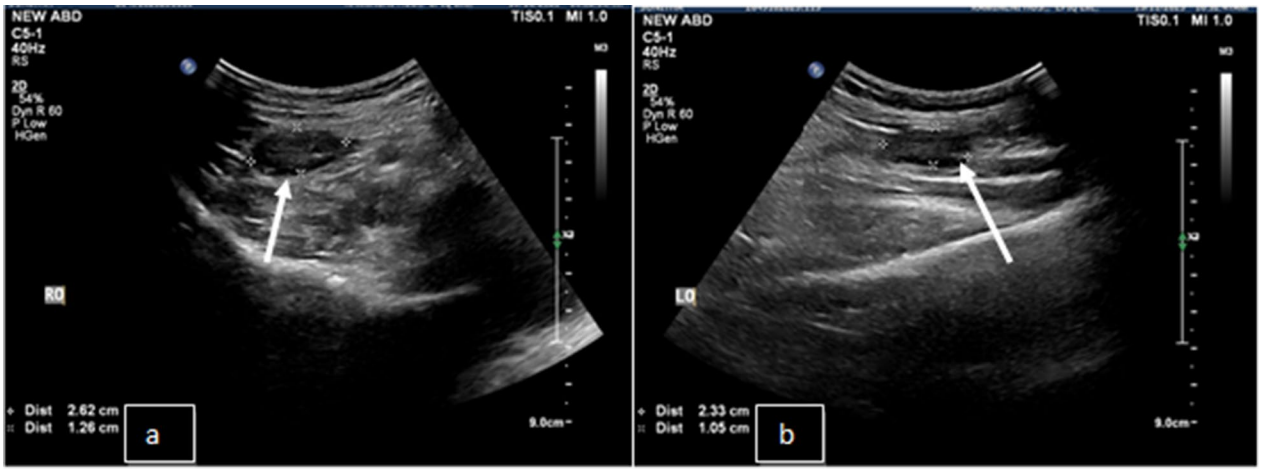

Fig. 4 **a** and **b**. USG abdomen and pelvis showing normal sized right ovary (arrow in **a**) and normal sized left ovary (arrow in **b**)

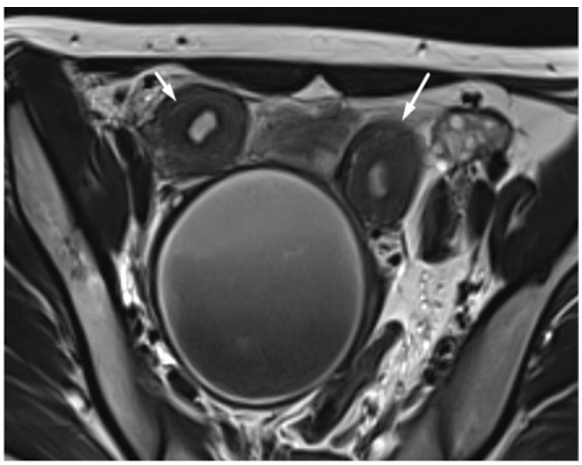

Fig. 5 MRI pelvis T2-weighted axial images showing two uterine cavities (arrows)

The incidence of uterus didelphys, related to HWW, is approximately 1/2,000 to 1/28,000, and it is accompanied by unilateral renal agenesis in 43% of cases [9].

Embryology of uterovaginal development is essential to understand the pathogenesis of Mullerian anomalies. Two theories have been described, the classic theory and acien theory. The classic theory described that the vagina is divided into upper and lower parts, where upper part of vagina is developed by Mullerian ducts and lower part is developed by sinovaginal bulbs of the urogenital sinus. This theory talks about Mullerian duct anomalies, but not the development of anomalies such as OHVIRA and the association of Mullerian duct anomalies with kidney pathology [10]. Acien theory explains that there are two ducts—Mullerian and paramesonephric ducts. Another duct is Wolffian duct, also known as mesonephric duct. Uterus and cervix are formed from joined Mullerian ducts, and vagina is formed from Wolffian duct. The vaginal lining consists of paramesonephric cells, derived from the paramesonephric tubercle, but the entire vagina is not developed from paramesonephric ducts. And since vaginal and renal development occurs from the Wolffian duct, if there is any error in derivation from one side of the Wolffian duct, it will affect the same side of the kidney and vagina or both. This theory helps in understanding complex anomalies such as OHVIRA [11, 12].

Near fifth week of gestation, Wolffian duct gives rise to the metanephric diverticulum. If this does not form, it will lead to the agenesis of the same side of the kidney and hemivagina. Mullerian development is lateral to the Wolffian duct at around nine weeks. After crossing the Wolffian duct, they come in midline and fuse to form the cervix and uterus. If they don't fuse, it results in uterine didelphys. Hence anomalies in the Wolffian duct and the fusion of two Mullerian ducts will give rise to OHVIRA.

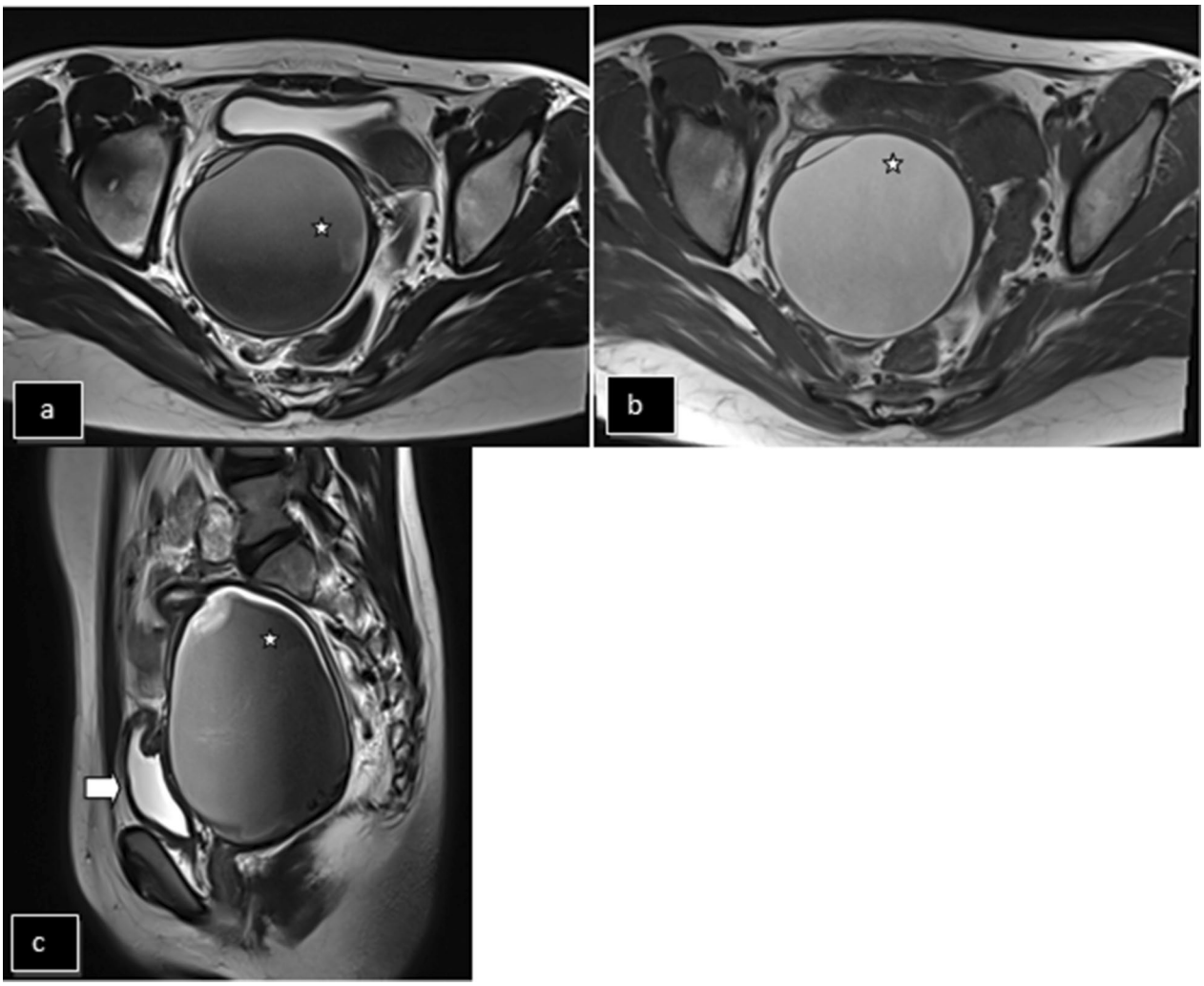

Fig. 6 **a**–**c** MRI axial T2W axial, T1W axial and T2W sagittal images. The right hemivagina is distended by T1 hyperintense (star in **b**), T2 hypointense (stars in **a** and **c**) collection measuring 7.4 × 7.4 × 9.5 cm and low-lying debris displacing the left hemivagina, rectum to the left and compression of the urinary bladder anteriorly (arrow in **c**)—likely obstructed right hemivagina with subacute blood secondary to longitudinal vaginal septum

The diagnosis of such a syndrome requires multimodality approach, including detailed history, examination and appropriate imaging studies. Ultrasound and MRI or the imaging modalities are used for diagnosing such a condition. 3D ultrasound has 93% sensitivity and 100% specificity in the assessment of Mullerian duct anomaly. However, it requires the expertise of the radiologist due to small size of the uterus, non-reactive endometrium and distended vagina in prepubertal and pubertal girls [13]. The Gold standard investigation is MRI which can confirm the diagnosis and provide detailed information about the internal and external uterine anatomy and can diagnose associated extragenital anomalies. It has 100% diagnostic accuracy [14].

In our patient, there was a suspicion of a dilated ectopic ureter inserting into the obstructed right hemivagina. However, this finding requires further investigation like MR urography to confirm its presence, which was not

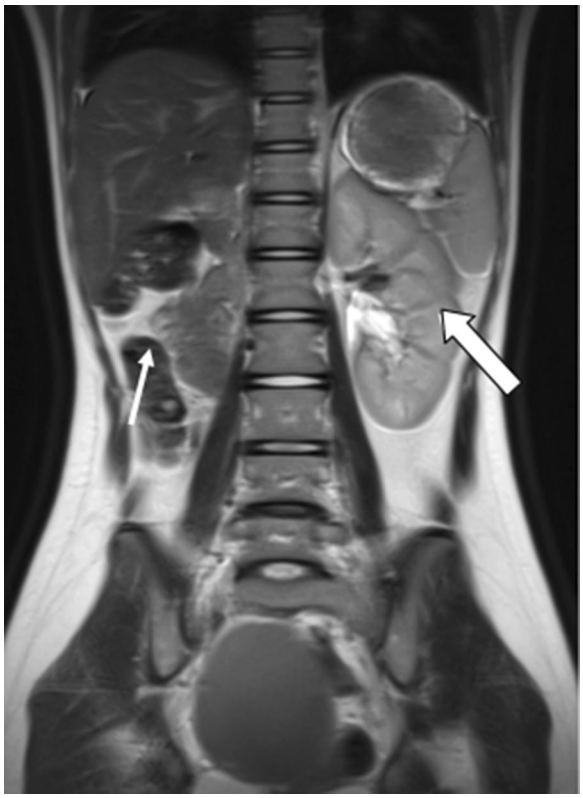

Fig. 7 MRI T2-weighted coronal images. Absent right kidney in right renal fossa (thin arrow) with left kidney showing compensatory hypertrophy (thick arrow)

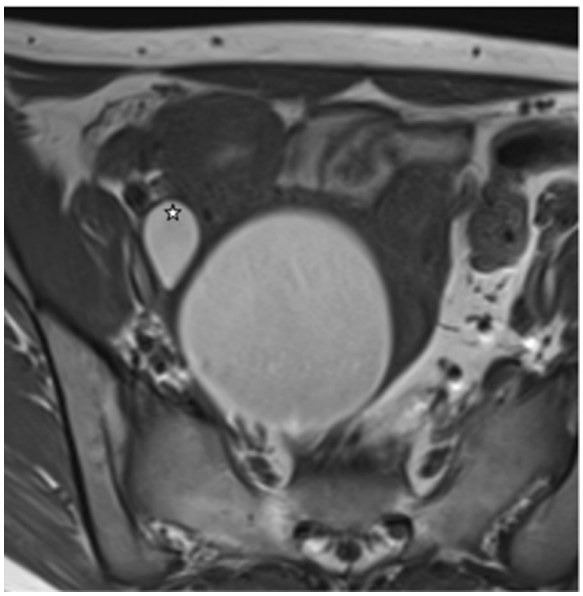

Fig. 8 MRI pelvis T1 axial images. A T1 hyperintense cystic structure (star) in the right adnexa inserting into the upper vagina—likely a blind right ectopic ureter with hematic reflux from hematocolpos

done in our case. The management of ectopic ureter in such a case would be transvaginal ligation of the ureter or laparoscopic transperitoneal nephrectomy [15].

The absence of ipsilateral absent of kidney, however, doesn't fully exclude the possibility of a small atrophic kidney. Additional investigations like intravenous urography, Technetium-99 m dimercaptosuccinic acid (DMSA) renal scan and contrast vaginography can be done to look for small atrophic kidney. However, these investigations were not performed in our case.

Primary management is vaginoplasty. It is done to relieve the obstruction. Before two-stage procedure used to be done, with the initial surgery to relieve the obstruction and the second to resect the septum. Currently, a single-stage vaginoplasty is done in which complete resection of the septum in a single procedure is carried out [16]. Surgery helps in relief of pain due to obstruction. It also reduces of the chance of pelvic endometriosis caused by retrograde menstrual seeding which can lead to infertility and chronic pelvic pain. Patients can have normal sexual life and can conceive and carry pregnancy to term. In a study, Altchek and Paciuc have reported pregnancy occurring twice in a previously obstructed didelphys uterus after surgical correction [17]. Therefore, effort must be made to preserve the obstructed uterus.

Conclusions

In conclusion, OHVIRA syndrome is an uncommon congenital anomaly. Imaging plays an important role in diagnosis. Surgery is the treatment of choice to resect the septum and relieve the obstruction. Early diagnosis helps to relieve the symptoms and prevent complications, which are caused by retrograde menstruation resulting in endometriosis and, also, preserve sexual and conception abilities [18].

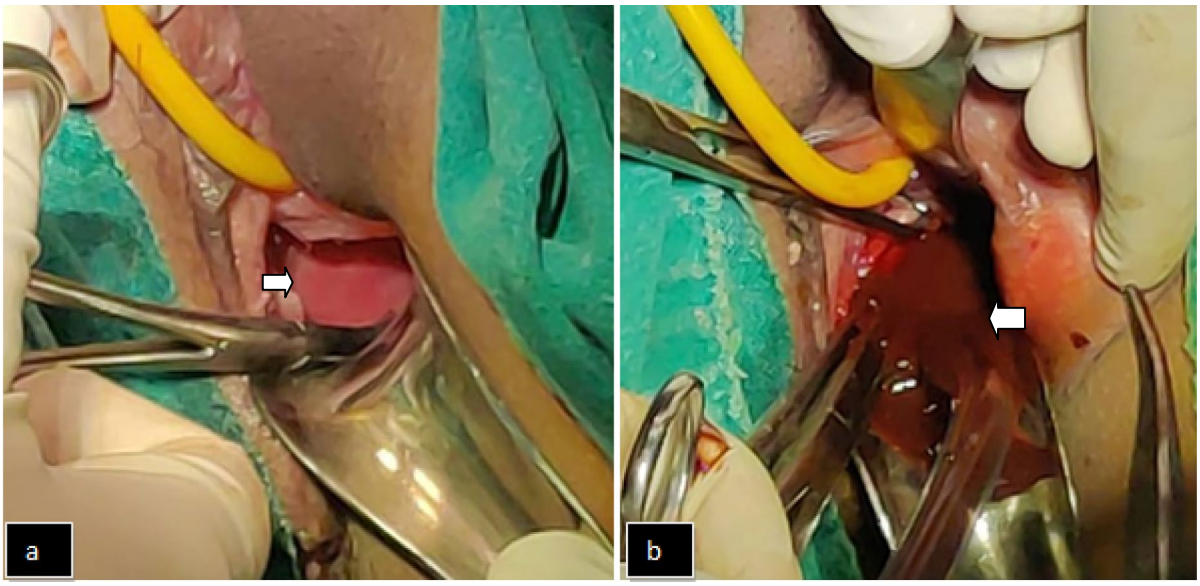

Fig. 9 **a** and **b** Intraoperative photograph showing smooth bulge (arrow in **a**) caused by right hematocolpos. Intraoperative photograph of septoplasty procedure showing drainage (around 150 ml) of chocolate colored fluid (arrow in **b**)

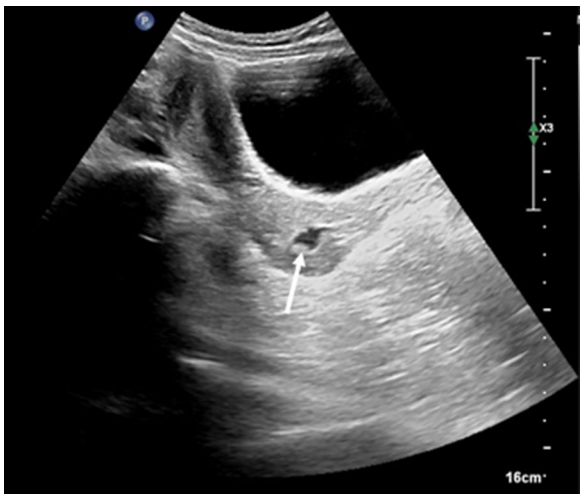

Fig. 10 USG abdomen and pelvis postdrainage showing minimal fluid in the lower uterine cavity of the right uterus (arrow)

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Availability of data and material
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Declarations

Ethics approval and consent to participate
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Consent for publication
An informed consent was obtained from the patient to use the necessary data for publication.

Competing interests
The author claims that there is no competing interest.

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Abbreviations

HWWS	Herlyn–Werner–Wunderlich syndrome
OHVIRA	Obstructed hemivagina, ipsilateral renal agenesis

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Author contributions
DJ did the major write up of this review article. The case in this review article was diagnosed and followed up by BB. The work was carried under the guidance BB and BV provided us the insight and knowledge to diagnose the syndrome. All the authors read the rough draft and provided valuable suggestions for the final draft. BB reviewed this article for corrections and final draft.

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