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Case report

Herlyn Werner Wunderlich syndrome: A case of obstructed hemivagina with ipsilateral renal agenesis (OHVIRA syndrome) and didelphic uterus



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KEYWORDS

Obstructed hemivagina;
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Abstract

We report a rare case of obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) syndrome with a didelphic uterus. A 22-year-old female presented with infertility for one year. Imaging studies revealed an absent left kidney, massive hematometrocolpos on the left side with normal other side. She was diagnosed with OHVIRA syndrome. She underwent septostomy of the vaginal septum, drainage of hematometrocolpos and hysterolaparoscopy that showed a bicornuate uterus and hematosalpinx. Given the rarity of the syndrome it is frequently misdiagnosed. Delayed diagnosis can lead to endometriosis.

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Introduction

Obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) syndrome with uterine anomaly is a rare disorder. The incidence of

Müllerian duct anomalies ranges from 0.8% to 4%. The incidence of the OHVIRA syndrome is not precisely known, but according to the literature it is estimated as 0.1–3.5% of all Müllerian anomalies [1,2]. The patient is usually young and presents with dysmenorrhea. The diagnosis is frequently delayed. Early diagnosis and excision of the vaginal septum will relieve the patient of her symptoms and prevents subsequent development of endometriosis and infertility.

Case report

22-year-old female presented to our clinic with primary infertility after one year of marriage. The patient's menarche was at the age of 13 years. She had regular menstrual cycles associated with dysmenorrhea and there was a history of dyspareunia. General examination showed well-developed breasts and normal feminine axillary and

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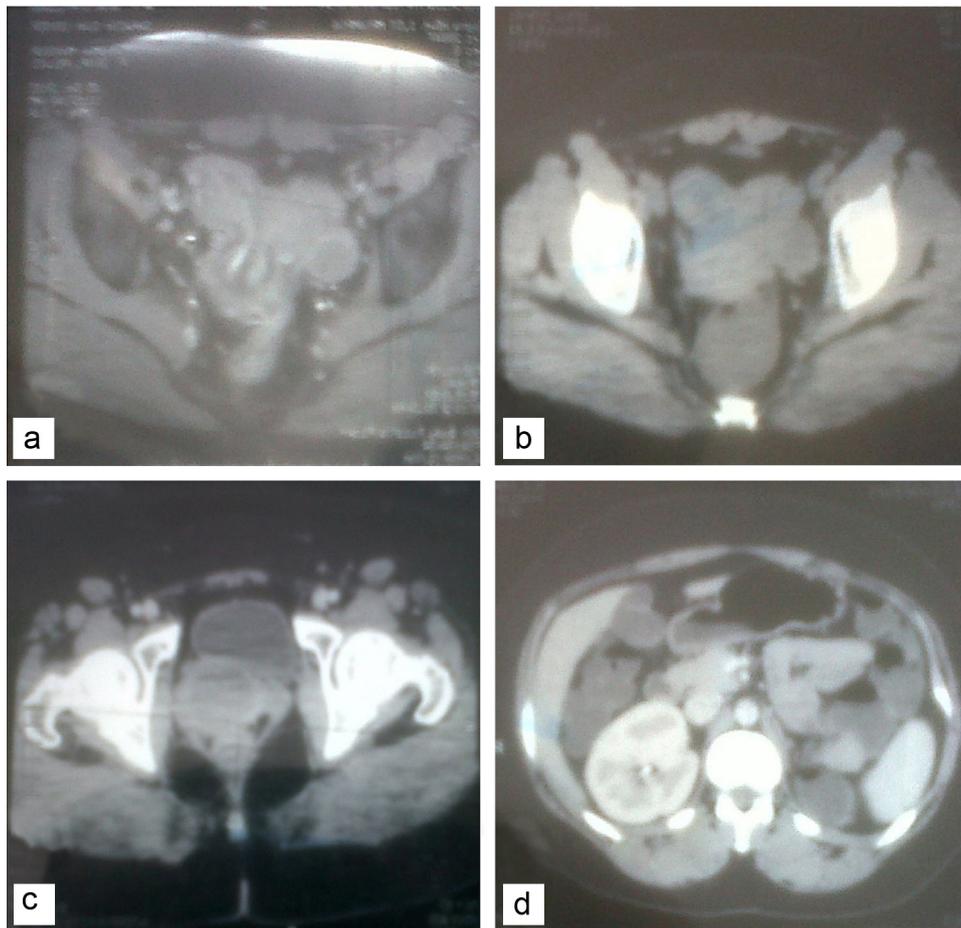


Figure 1 CT showing (a) didelphic uterus and left adnexal cystic structure, (b) the cystic vaginal swelling, (c) the vaginal swelling between the bladder and anal canal, (d) absent left kidney.

pubic hair distribution. Abdominal examination revealed no tenderness or swelling.

Laboratory tests, including complete blood count, urinalysis and hormonal profile were normal, and the pregnancy test was negative. The husband's semen analysis was normal. Vaginal examination revealed left antero-lateral cystic swelling in the upper vagina adjacent to the cervix. Bimanual examination showed irregular uterine contour with left adnexal swelling.

Hysterosalpingography showed a picture suggesting a unicornuate uterus, because the contrast medium was injected in one cervix at a time. Abdominal/pelvic ultrasound revealed a didelphic uterus with the left horn larger than the right one. There was a partial longitudinal vaginal septum enclosing a hematocolpos with the cervix in it. The left horn and the Fallopian tube were filled with echogenic fluid. The left kidney was not visualized. Intravenous pyelography (IVP) showed no visualization of the left kidney and left ureter with mild compensatory hypertrophy of the right kidney. Computed tomography (CT) and magnetic resonance imaging (MRI) showed two separate uteri and cervixes with a longitudinal vaginal septum and an obliterated left side that was seen distended and filled with blood (Figs. 1 and 2).

On the basis of the imaging findings of unilateral renal agenesis, uterus didelphys, unilateral obstructed hemivagina with resultant

hematometrocolpos and hematosalpinx, the case was diagnosed as OHVIRA, or Herlyn–Werner–Wunderlich syndrome. Laparoscopy was performed to determine the extent of hematometra and hematosalpinx and to assess the adnexa for any endometriosis from retrograde menstrual flow.

Laparoscopy confirmed the ultrasound and MRI findings and did not reveal any endometrial peritoneal deposits. Methylene blue instilled through the visible cervix was seen exiting only from the left Fallopian tube, demonstrating a lack of communication between the uterine cavities. The vaginal septum was excised up to the level of the cervix. The post-operative recovery period was uneventful. The patient became pregnant after two months.

Discussion

Herlyn–Werner–Wunderlich syndrome is a triad of obstructed hemivagina, uterus didelphys and ipsilateral renal agenesis. The acronym OHVIRA is used to describe the triad of obstructed hemivagina and ipsilateral renal agenesis and any other uterine anomaly other than uterus didelphys [2,3]. Both syndromes are rare and are discussed as the same entity in many case reports. The presentation and management of the two conditions do not differ very much, so using different names only adds to the prevailing confusion. We are of

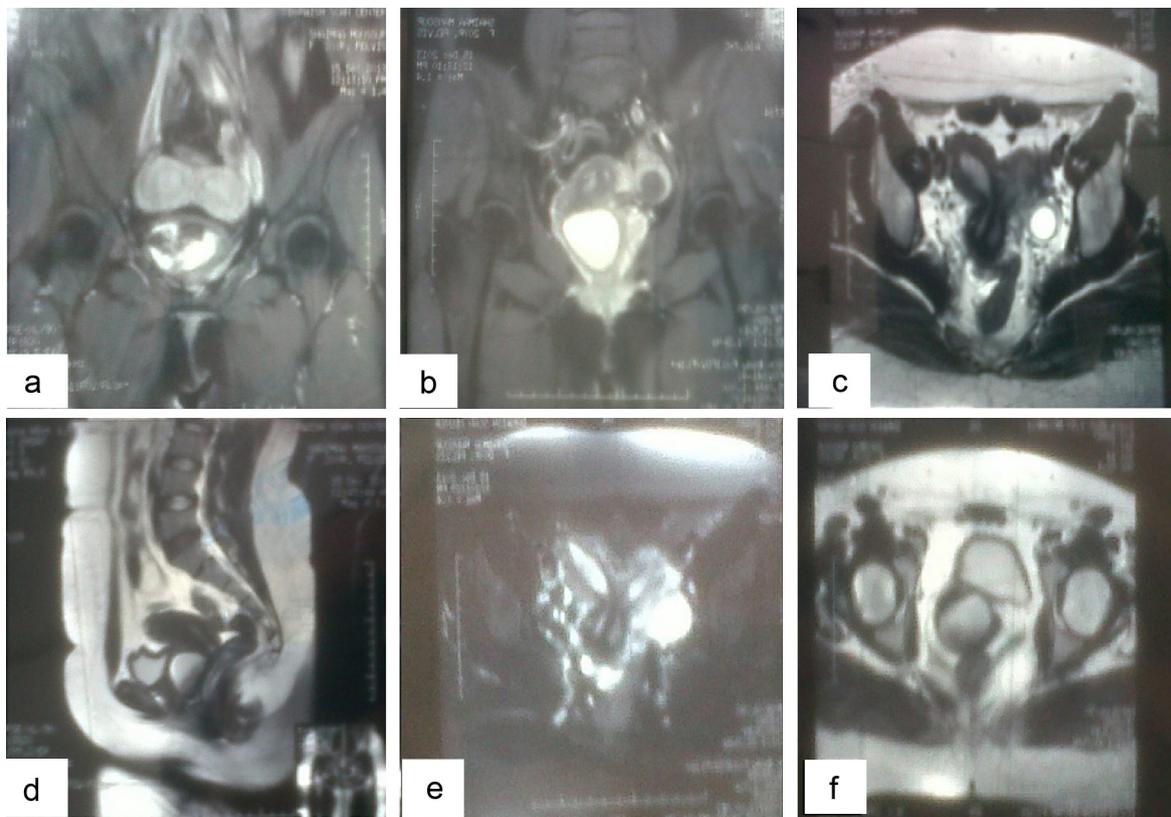


Figure 2 MRI showing (a) didelphic uterus, (b) left uterine horn with cystic vaginal swelling below and left adnexal cystic structure, (c and e) didelphic uterus, (d) uterus and vaginal swelling between bladder and rectum, (f) cystic vaginal swelling between bladder and anal canal.

the opinion that this should be referred to as OHVIRA syndrome, including all forms of uterine anomalies [3].

The aetiology is not known. The syndrome can be associated with urogenital sinus, bladder exstrophy and other renal anomalies. The simultaneous insult to the paramesonephric system and metanephros could suggest a multifactorial origin [4]. Bajaj et al. have summarized the embryological development as related to OHVIRA syndrome. An early failure of the metanephric diverticulum to develop from the mesonephric duct results in agenesis of the ureteric bud, leading to ipsilateral agenesis of the ureter and kidney. The mesonephros is responsible for the development and positioning of paired paramesonephric ducts. Due to failed positioning of the paired paramesonephric ducts, the two hemiuteri and hemicervixes fail to unite, resulting in Müllerian anomalies associated with OHVIRA syndrome [5].

There is a lack of awareness of the OHVIRA syndrome due to its rarity. The classical presentation is that of a young girl with severe dysmenorrhea a few months to one year after attaining menarche. Usually they are treated symptomatically or as endometriosis until they develop an abdominal mass and pressure symptoms. Retrospective study of cases has shown that the mean age of presentation is about 15 years [6]. Pelvic pain is the most common presenting symptom (90%) followed by an abdominal mass (40%) and pressure symptoms. Patients can present at a later age with foul smelling vaginal discharge due to pyocolpos [7].

Ultrasonography is helpful in diagnosis, but MRI is usually conclusive. The management is excision of the vaginal septum and drainage

of the hematometrocolpos. Cetinkya et al. have discussed the use of hysteroscopy to excise the vaginal septum and preserve hymenal integrity [8].

Laparoscopy may be done in the same sitting to clearly identify the uterine anomaly. Endometriosis is frequent. After excision of the vaginal septum, precautions must be taken to keep the outflow tract patent [9].

The fertility of patients with OHVIRA syndrome is not substantially compromised. Septate uterus excision is indicated if there is infertility or adverse obstetric outcomes. Hysteroscopic metroplasty is the procedure of choice for uterine defects.

Conclusion

OHVIRA is a rare syndrome of Mullerian and Wolffian duct abnormalities. A simple excision of the vaginal septum can relieve the patient of her symptoms. Given the rarity of this syndrome it is frequently misdiagnosed or diagnosed late.

Conflict of interest

There is no conflict of interest.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Acknowledgments

I acknowledge the cooperation of EL-Shatby Maternity University Hospital residents who participated in appointing the patient and following up. I also appreciate the commitment and compliance of the patient who reported the required data and attended for regular follow up.

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