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# Uterus didelphys with obstructed hemivagina and ipsilateral renal anomaly (OHVIRA syndrome): A case report<sup>☆</sup>



Mohamed Amin El-Gohary\*

Department of Surgery, Division of Pediatric Surgery, Burjeel Hospital, P.O. Box 7400, Abu Dhabi, UAE

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## ABSTRACT

Uterus didelphys with obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome is a rare congenital urogenital anomaly. We report a case of a 14-year-old female who presented with acute lower abdominal pain and tender pelvic mass. The initial diagnosis of twisted left ovarian cyst was confirmed by abdominal ultrasonography (US), which showed an associated absent left kidney. The findings at laparoscopy were that of uterus didelphys with obstructed left hemivagina. Parents were concerned about hymnal integrity and refused hemivaginal septal resection.

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The triad of uterine didelphys, obstructed hemivagina, and ipsilateral renal anomaly, known as OHVIRA syndrome, is a rare congenital anomaly (Fig. 1). It represents a diagnostic dilemma because of the regular menstruation and nonspecific abdominal pain. Awareness of such anomaly is prerequisite for early and prompt diagnosis and prevention of potential complications.

## 1. Case report

A 14-year-old girl was admitted to our institute with 2 days colicky left lower abdominal pain. The pain started with her menstruation with gradual increase in severity. She had milder episodes of lower abdominal pain following her menses which responded to oral anti-inflammatory medication. Examination revealed an ill-defined tender mass at the left iliac fossa with guarding and limited mobility. The clinical diagnosis of twisted left ovarian cyst was initially made and confirmed by ultrasonography (Fig. 2), with the additional finding of an associated absent left kidney.

However the findings at laparoscopy were that of uterus didelphys with left sided enlarged hemivagina and hematometocolpos (Fig. 3). A needle was inserted percutaneously during laparoscopy

inside the dilated hemivagina and 200 ml of altered blood aspirated. It was decided to stop at this stage and discuss further management with our gynecologists and the parents.

The working plan of doing magnetic resonance imaging (MRI), followed by vaginal septotomy was suggested, but refused by the parents for fear of infringing hymnal integrity. When given the available alternatives the parents opted to seek hysteroscopy excision of the vaginal septum at another facility.

## 2. Discussion

The triad of uterine didelphys, obstructed hemivagina, and ipsilateral renal anomaly, known as OHVIRA syndrome, is a rare congenital anomaly of the Mullerian ducts and Wolffian structures [1]. The combination of obstructed hemivagina and uterus didelphys was first reported in 1922 [2]; however the triad of obstructed hemivagina and uterus didelphys as well as an ipsilateral renal anomaly (OHVIRA syndrome) was initially reported in 1950 [3]. Incidence of these anomalies is believed to be between 0.5 and 5.0% [4,5].

The uterus, Fallopian tube, cervix and upper two thirds of the vagina develop from the paired mullerian ducts while the lower third of the vagina develops separately from the urogenital sinus. The association of uterine anomalies with renal anomalies ipsilateral to the side of obstruction can be explained by embryologic arrest at the 8th week of gestation, which simultaneously affects the mullerian and metanephric ducts. Certain other renal

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\* Corresponding author. Tel.: +971 2 508 555; fax: +971 2 508 556.

E-mail address: [amingoh@gmail.com](mailto:amingoh@gmail.com).

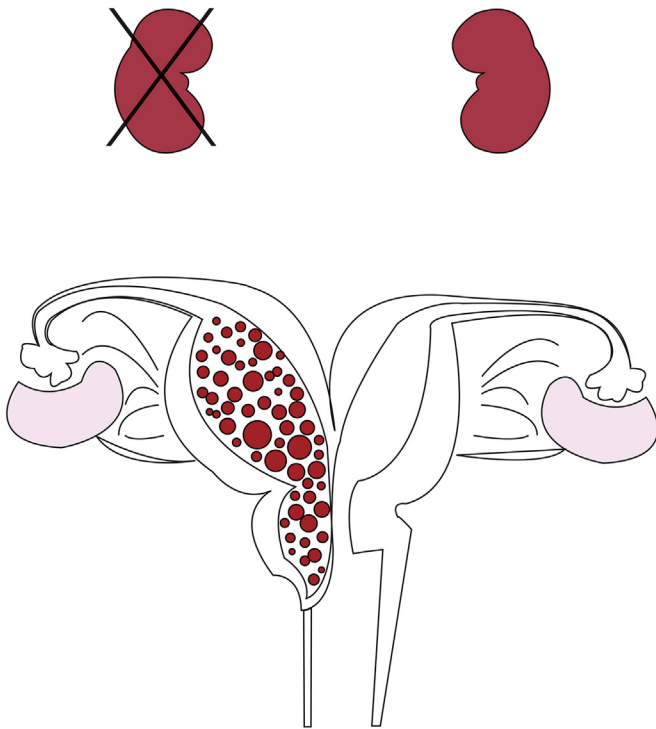


Fig. 1. Diagrammatic illustration of OHVIRA syndrome.

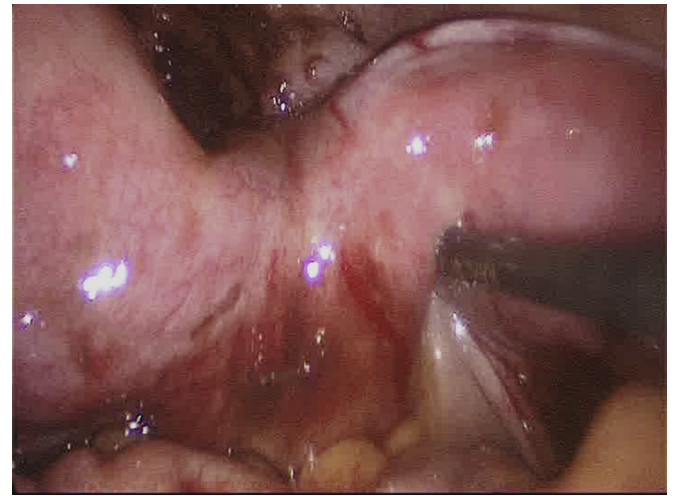


Fig. 3. Laparoscopic finding of uterus didelphys.

anomalies may also be associated, such as renal dysplasia, double collecting system and ectopic ureter [5]. The condition has also been reported to be associated with high-riding aortic bifurcation, Inferior vena cava (IVC) duplication, intestinal malrotation and ovarian malposition [6].

The most common clinical presentation is pelvic pain initiating shortly after the monarch, associated with a vaginal or pelvic mass and normal menstrual periods. The didelphys uterus in these cases is associated with reproductive issues such as miscarriages, preterm labor, and placental dysfunction. Rare presentations may include intermenstrual bleeding, acute retention of urine, fever, vomiting, and abdominal swelling, [7–10].

Most of the patients suffering from this syndrome are diagnosed late due to its rarity and the nonspecific clinical presentation.



Fig. 2. Ultrasound showing a large pelvic cyst diagnosed an ovarian cyst.

Moreover, the menstrual flow that comes from the patent unobstructed hemivagina gives the impression of normal menses. Consequently accurate diagnosis and surgical treatment may be delayed for several months or even years.

Imaging modalities used to diagnose this condition include ultrasonography, and (MRI). Computed tomography (CT) has a limited role in evaluation of the female pelvis.

The obstructing vaginal septum seen in this condition is usually oblique and varies in thickness from very thin to quite thick. Resection of the vaginal septum is the treatment of choice [11].

The percutaneous aspiration of the altered blood from the hematocolpus done in our case during laparoscopy is not a standard treatment and it was meant to temporary alleviate the obstructive symptoms until investigation is completed and full drainage is achieved.

The integrity of the hymen represent a major cultural issues in our community, The hysteroscopic excision of vaginal septum in uterus didelphys has been recommended for the management of those patients with good outcome [12]. The parents of our patients opted to seek this option in another facility.

It was unfortunate that our case was diagnosed after laparoscopic findings, but it was based on common expected pathology and it was the first time for the surgeon and radiologist to encounter such rare case.

### 3. Conclusion

OHVIRA syndrome is a rare anomaly with potential short and long term complication. The diagnosis is likely to be missed because of the normal menstruation and nonspecific abdominal pain. Reporting such cases increases awareness of the syndrome and helps to achieve early diagnosis and avoid potential complication.

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