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Diverse Presentations of OHVIRA Syndrome: A Case Series

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Introduction

OHVIRA (obstructed hemivagina and ipsilateral renal anomaly) is a rare condition first reported in 1925 and subsequently described in detail by Herlyn, Werner, and Wunderlich after whom it was initially named HWW syndrome. It is characterized by the triad of uterine didelphys, obstructed hemivagina, and ipsilateral renal anomaly. Delays in diagnosis are common due to normal onset of puberty and menstruation. Patients usually present with progressive dysmenorrhea and nonspecific lower abdominal pain, though other presentations such as urinary retention and pelvic mass are possible. The incidence is unknown but has been quoted at 0.1% and 3%,[1] and the etiology is poorly understood. There are a range of phenotypic variables; hence, when patients present with dysplastic or absent kidneys, an attempt to establish uterovaginal anomalies should be made to allow early recognition and management. However, because the presentation is variable prompt recognition can be difficult. Though renal agenesis is the classic presentation, other anomalies include renal dysplasia and ectopic ureters.[2] As well as renal anomalies ipsilateral to the obstructed hemivagina, contralateral renal anomalies have been reported in up to 50%.[2,3] Renal physicians need to be aware of this condition to allow appropriate referral to a pediatric gynecologist.

Variants of OHVIRA have been recognized where the septum has a fenestration resulting in only a partial obstruction as reported in a series by Fedele and colleagues.[4] Occasionally the septum is thin and retained menstrual contents can cause it to perforate resulting in a communication between the 2 vaginas.[5] This can also predispose to ascending bacterial infections with development of a pyometra or a pelvic infection. Cervical atresia and a double cervix with hemicervical obstruction with a rudimentary hemiuterus may present as OHVIRA syndrome.

In most patients with OHVIRA, the septum is thick and limits the ability of the hemivagina to distend resulting in retrograde bleeding causing endometriosis. Endometriosis has been reported in 17% to 35% of patients with this condition.[5-7] This can be avoided by timely decompression.

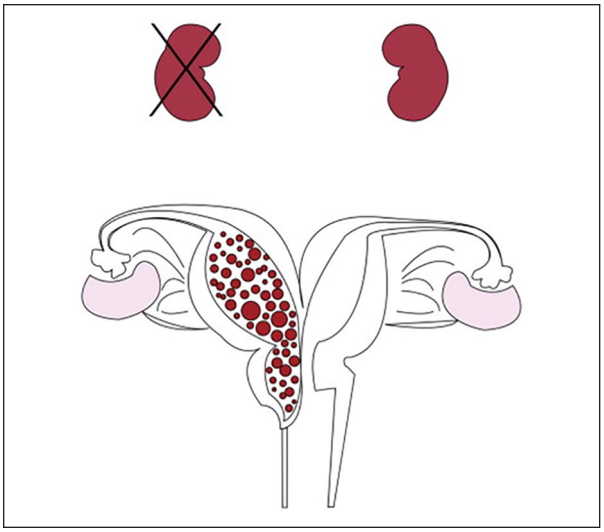

Figure 1. Graphical representation of the triad of OHVIRA (obstructed hemivagina and ipsilateral renal anomaly) syndrome.

In patients with OHVIRA, ovarian function is normal. The impact of this condition on fertility is not well known, but problems may arise due to endometriosis, development of pelvic abscess, and pelvic adhesions. There are case reports of pregnancy occurring in the obstructed hemiuterus.[8]

Figure 1 shows the graphical representation of the condition.

In this case series we report on 3 patients who were referred to the Tertiary Pediatric and Adolescent Gynecology clinic over a 12-month period, all with OHVIRA syndrome. The clinical presentation of all 3 patients was completely different though the underlying condition and management was the same.

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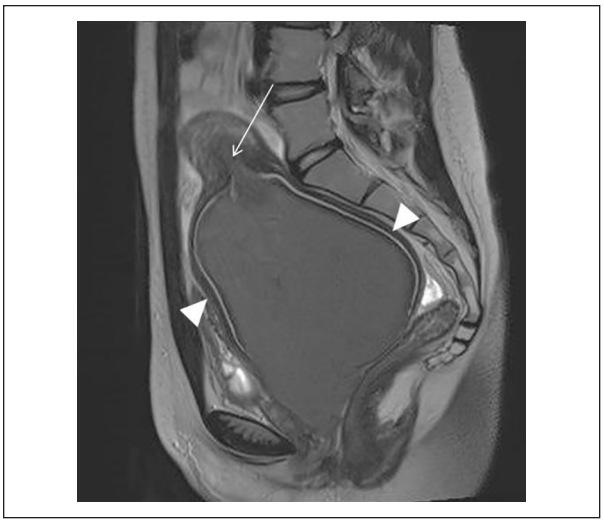

Figure 2. T2-weighted sagittal image of the distended cervix. The uterine cavity (arrow) is only slightly distended. The cervical stroma (arrow heads) is considerably stretched.

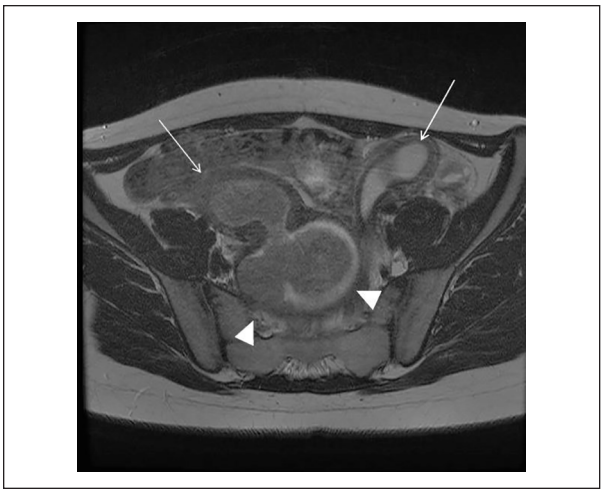

Figure 3. T2-weighted axial image of both uterine horns (arrows) in another patient. Note how separate the uterine horns are. The right-sided uterine cavity is more distended and is in direct communication with the dilated cervix (arrow heads).

Case Series

Case 1

A 15-year-old presented with acute urinary retention during her menses. Her menarche was at the age of 12 years and she complained of severe dysmenorrhea from the outset. Six months prior to the hospital admission, the patient had developed difficulties voiding urine at the time of her menstruation. An indwelling catheter was inserted on admission and an ultrasound scan followed by magnetic resonance imaging (MRI) was performed. The right kidney was absent and there was a uterine didelphys with a massive hematometra and hematocolpos or the right with an obstructed hemivagina. The cervix was grossly dilated and there was loss of normal cervical anatomy and the cervical canal and the obstructed hemivagina forming one continuous tract (Figure 2). The uterus on the left appeared normal and the patent vagina was displaced laterally by the obstructed hemivagina. The hematocolpos component alone measured 10.5 cm.

Case 2

A 13-year-old presented with progressive dysmenorrhea with menarche aged 12. The pain was initially worse during the menses but progressed to occur in between the menses too. She had to be home schooled due to severe menstrual pain. At the time of presentation, she had not had menstruation for 3 months, other than light bleeding and spotting.

The MRI showed that the right kidney was absent and an obstructed hemivagina with a large hematocolpos on the right. There was a patent vagina on the left, but due to the extent of the hematocolpos on the right, the left vagina was occluded with evidence of minor hematometra on the left side too (Figure 3).

Case 3

A10-year-old pre-pubertal child, excessively tall for her age was referred to the endocrine clinic for further investigations. She underwent further testing and was found to have triple X syndrome. The ultrasound showed a single kidney, 2 uterine horns, and presence of fluid on the right side of the vagina. A repeat scan 6 months later showed that the fluid on the right side had increased and was suggestive of a vaginal septum. An MRI was performed and OHVIRA was confirmed. A plan was made for the patient to return to the clinic following her menarche. This would allow better delineation of the septum for removal. The patient attended the clinic after her first menses. Further menstruation was suppressed and the septum removed electively. The association of triple X syndrome in this patient was incidental rather than causative as no link has previously been established.

Management

Investigation of this condition is best done by MRI imaging. This is because the obstructed hemivagina distorts the anatomy making it difficult to assess with an

ultrasound scan. In all 3 cases the menstrual flow was suspended by the use of progesterone (Norethisterone, 5 mg BD). There is no requirement for gonadotropin-releasing hormone analogues for menstrual suppression as this can be achieved with oral preparations of progesterone. Surgery was arranged electively and the patients were advised to allow a withdrawal bleed immediately before surgery. This allowed the vaginal septum to be distended making identification easier and excision easier.

The patients were informed of the risk of incomplete excision and the need for further surgery. They were also informed of the increased risks of endometriosis (due to retrograde menstruation) and miscarriage/preterm labor in later life if conception occurred in the obstructed uterine horn. The patients were advised they would require cervical screening from both cervices.

Summary and Conclusion

OHVIRA syndrome requires a high index of suspicion for diagnosis. It is important to educate renal physicians and pediatricians who are most likely to encounter the syndrome early, to consider this as a differential diagnosis. Complications such as pelvic infection due to ascending infection may require emergency excision of the septum. However, in the absence of acute symptoms treatment can be elective with initial suppression of menstruation. Renal function should be established and monitored as deterioration can occur around puberty due to rapid somatic growth. There are theoretical risks of hypertension, proteinuria, and chronic kidney disease in later life. Patients should be advised to avoid factors that may affect renal function such as nonsteroidal anti-inflammatory drugs.

Early diagnosis and removal of the obstructing septum can prevent complications and allow preservation of fertility in most patients. Where the condition is complicated by cervical atresia, surgery may be more complex. All cases should be managed in a tertiary center where there is adequate expertise and multidisciplinary approach to care.

Learning Points

- OHVIRA syndrome is a condition of obstructed hemivagina and ipsilateral renal anomaly.
- Presentation of OHVIRA syndrome is diverse and requires a high index of suspicion for diagnosis.
- Emergency department pediatricians and gynecologists as well as renal physicians should be educated about the condition.
- In young girls where renal agenesis or other abnormalities are detected an attempt to rule out OHVIRA prior to menarche should be made.
- Management should be in a tertiary center where there is experience of managing Mullerian duct anomalies.

Author Contributions

SJ: Patient care, outpatient review, and surgery; manuscript preparation; and background research.SA: Reported on the imaging and editing manuscript.

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