Radiological Manifestations of Obstructed Hemivagina and Ipsilateral Renal Anomaly Syndrome: A Rare Complex Müllerian and Wolffian Duct Anomaly

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ABSTRACT

Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome is a rare complex Müllerian and Wolffian duct anomaly. It is also known as Herlyn–Werner–Wunderlich syndrome (HWWS). It includes unilateral renal anomalies and uterine didelphys. It generally occurs at puberty and exhibits non-specific and variable symptoms with acute or pelvic pain shortly following menarche, causing a delay in the diagnosis. We report here a 16-year-old female presenting with progressive cyclical pelvic pain, where magnetic resonance imaging (MRI) suggested the diagnosis of the OHVIRA syndrome. She was managed by surgical resection of the septum and draining of the obstructed vagina.

Keywords: Herlyn–Werner–Wunderlich syndrome, Müllerian duct anomaly, OHVIRA syndrome, Renal agenesis, Uterus didelphys.


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Conflict of interest: None

INTRODUCTION

Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome is a rare congenital anomaly of Müllerian (paramesonephric) and Wolffian (mesonephric) ducts. It is also known as the Herlyn–Werner–Wunderlich syndrome (HWWS). Purslow1 first reported it as a combination of obstructed hemivagina and uterine didelphys. Classical OHVIRA syndrome with triad was reported in 1950.2 Müllerian anomalies are present in 0.8 to 4% of the female population and the OHVIRA syndrome comprises about 0.1 to 3.5% of those Müllerian abnormalities.3 Diagnosis in most cases is late due to its rare incidence and non-specific clinical presentation,4-7 and also menstrual flow that comes from the patent unobstructed hemivagina gives the appearance of normal menses.

CASE REPORT

A 16-year-old female presented with complaints of acute lower abdominal pain and mild dysmenorrhea. Her menarche had occurred at the age of 12 and she had regular menstruation, once every 30 days for 4 to 5 days. Physical examination revealed tenderness over the right lower quadrant of the abdomen. External genital inspection showed an intact hymen and menstrual discharge. She underwent ultrasonographic evaluation, which revealed two widely separated uterine horns and large heteroechoic cystic mass at the lower aspect of both uterine horns (Figs 1A and C). Right uterine horn was distended with heteroechoic fluid and was clearly connected to the cystic mass, consistent with hematometocolpos (Figs 1B and D). Communication
between the left uterine horn and hematocolpos was not clearly visualized (Figs 2A to C), which suggested the possibility of bicornuate uterus or uterine didelphys. A dilated tubular structure filled with slightly heteroechoic fluid and in close proximity to the right uterine horn was also observed, consistent with hematosalpinx (Fig. 2D). Right kidney was not visualized, and left kidney was relatively hypertrophied. Magnetic resonance imaging (MRI) was performed for further evaluation. Magnetic resonance imaging showed two completely separate uterine horns with normal zonal anatomy, two separate cervices, and large hematocolpos connected to the right uterine horn, corresponding to obstructed right hemivagina (Figs 3 to 5). Right-sided hematometra and hematosalpinx (Figs 3) were also seen. Left uterine horn had its own opening into the left hemivagina (Figs 6 and 7). Both ovaries appeared normal, and right kidney was absent. Based on imaging findings of uterine didelphys, unilateral obstructed hemivagina with resultant hematometrocolpos and hematosalpinx, and unilateral renal agenesis, the case was diagnosed as OHVIRA syndrome.

Vaginal septotomy and drainage of 150 mL of hematometrocolpos and hematosalpinx were performed, and patient was discharged without any immediate complications. On follow-up, patient showed no complications and had normal menstrual flow.
**Figs 4A and B:** (A) Axial fat saturated T1W image of the pelvis shows hyperintense signal of the right hemivaginal collection (asterisk). The collapsed hypointense left hemivagina is seen adjacent to it (arrow), and (B) Axial T2W image of the pelvis showing heterogenous hypointense right hemivaginal collection (asterisk) and collapsed hyperintense left hemivagina with minimal fluid (arrow).

**Figs 5A and B:** (A) Coronal T2W image of the pelvis shows communication of hyperintense right hematometra with the right hematoccolpos (black arrow). The collapsed hyperintense left hemivagina is seen adjacent to it (white arrow), and (B) Coronal T2W image of the pelvis showing heterogenous hyperintense right hematoccolpos and collapsed hyperintense left hemivagina with minimal fluid (white arrow). No communication is visualized in these two images.

**Figs 6A and B:** (A) Sagittal T1W image of the pelvis shows hyperintense right hematometra (RH) with the right hematoccolpos (asterisk), and (B) Sagittal T1W image of the pelvis showing hypo to isointense left horn (LH).
DISCUSSION

Obstructed hemivagina and ipsilateral renal anomaly is rare Müllerian duct anomaly (MDA).Incidence of Müllerian defects is about 0.8 to 4%.\footnote{1} In women with recurrent miscarriages and subfertility, it is much higher – around 25%.\footnote{8}Mean incidence of uterus didelphys was 11.1%\footnote{8} with strong association of renal agenesis with uterus didelphys about 81%.\footnote{9} Until 2011, only about 200 documented cases were there in the world literature.

Herlyn–Werner–Wunderlich syndrome is characterized by uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis, i.e., combination of Type III MDA with mesonephric duct anomaly with vaginal septum. Obstructed hemivagina and ipsilateral renal anomaly syndrome is a broader term than HWWS, including other types of renal abnormalities, such as, renal agenesis, double collector, duplication renal system, horse-shoe kidney, duplicated kidneys, dysplastic kidneys,\footnote{10} rectovesical bands,\footnote{11} or crossed fused ectopia.\footnote{12} Same sided renal anomalies as obstructed hemivagina are always seen, which is explained by embryology. Right side is affected twice as often as the left side.\footnote{13} In the case of unicornuate uterus, renal anoma-

lies are ipsilateral to rudimentary or absent uterine horn.\footnote{14}

There is no universally accepted MDA classification system. Each system has its limitations. Buttram and Gibbons proposed a system for classification of MDAs in 1979, which classifies them into six categories. American Society of Reproductive Medicine (ASRM) revised it in 1988 into seven categories. Now it is the most frequently used classification\footnote{15} (Table 1). Limitation of these classification systems\footnote{36} is their lack of categorization of vaginal

<table>
<thead>
<tr>
<th>Class</th>
<th>Name</th>
<th>Description</th>
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<tbody>
<tr>
<td>I</td>
<td>Mayer-Rokitansky-Kuster-Hauser syndrome or Müllerian agenesis</td>
<td>Hypoplasia of the tubes, uterus, cervix, or vagina</td>
</tr>
<tr>
<td>II</td>
<td>Unicornuate uterus or Unilateral Müllerian anomaly (15%)</td>
<td>Complete (septum extends to the internal or external os) Partial (septum is confined to the fundal region) Two separate uterine horns and single cervix Fundal depression separating the two horns greater than 1 cm Intercornual distance greater than 4 cm</td>
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| III   | Uterus didelphys | No rudimentary horn (35%) Non-cavitatory rudimentary horn (33%) Cavitatory rudimentary horn [communicating (10%) or non-communicating (22%)]
| IV    | Bicornuate uterus | Complete (septum extends to the internal or external os) Partial (septum does not reach the internal os) Fundal depression separating the two horns less than 1 cm Intercornual distance less than 4 cm |
| V     | Septate uterus | Complete (septum extends to the internal os) Partial (septum is not divided) Fundal depression separating the two horns greater than 1 cm Intercornual distance greater than 4 cm |
| VI    | Arcuate uterus | Normal variant Endometrial cavity is not divided External contour of uterus is flattened or minimally concave |
| VII   | T-shaped uterus | Results from in utero exposure to diethylstilbestrol |
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Table 2: Embryological-clinical classification for female genitourinary malformations (revised and updated from Acién, 1992; and Acién et al., 2004a)

<table>
<thead>
<tr>
<th>Group</th>
<th>Name</th>
<th>Description</th>
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<tbody>
<tr>
<td>1</td>
<td>Unilateral genitourinary agenesis or hypoplasia</td>
<td>1.1 With contralateral Müllerian agenesis</td>
</tr>
<tr>
<td></td>
<td></td>
<td>1.2 Without contralateral agenesis</td>
</tr>
<tr>
<td>2</td>
<td>Uterine duplicity with a blind hemivagina and ipsilateral renal agenesis</td>
<td>2.1 Large hematocolpos, blind hemivagina</td>
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<tr>
<td></td>
<td></td>
<td>2.2 Like Gartner’s pseudocyst</td>
</tr>
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<td></td>
<td></td>
<td>2.3 Partial reabsorption of the vaginal septum</td>
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<tr>
<td></td>
<td></td>
<td>2.4 Complete unilateral vaginal or cervicovaginal atresia with communicating uteri</td>
</tr>
<tr>
<td></td>
<td></td>
<td>2.5 Complete unilateral vaginal or cervicovaginal atresia without communicating uteri</td>
</tr>
<tr>
<td>3</td>
<td>Isolated or common uterine or utero-vaginal anomalies</td>
<td>A. Paramesonephric or Müllerian ducts including same 7 types as American Society of Reproductive Medicine (ASRM) classification of Müllerian duct anomalies (Table 1)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>B. Müllerian tubercle</td>
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<tr>
<td></td>
<td></td>
<td>B.1 Complete vaginal or cervicovaginal agenesis or atresia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>B.2 Segmentary atresias</td>
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<td></td>
<td></td>
<td>C. Both Müllerian tubercle and ducts - Complete utero-vaginal agenesis</td>
</tr>
<tr>
<td>4</td>
<td>Accessory uterine masses and other gubernaculum dysfunctions</td>
<td>Accessory and cavitated uterine masses with normal uterus</td>
</tr>
<tr>
<td>5</td>
<td>Anomalies of the urogenital sinus</td>
<td>Imperforated hymen</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Persistent urogenital sinus</td>
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<td></td>
<td></td>
<td>Congenital vesicovaginal fistula</td>
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<tr>
<td>6</td>
<td>Malformative combinations</td>
<td>Variable</td>
</tr>
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and other anomalies that bridge more than one classification. Acién et al in 2011 and Singh et al in 1993 proposed an updated embryological–clinical classification of genitourinary malformations, which integrates current embryological and pathogenic concepts and is clinically applicable and useful (Table 2).

Review of embryology of uterovaginal development is important to understand pathogenesis of MDAs and their association with renal anomalies. Classic theory of vaginal development was given by Koff. It was found inadequate in explaining complex MDA, such as, OHVIRA and their coexistence with renal anomalies. Pathogenesis of majority of these complex anomalies can be correctly explained and understood through embryological hypothesis for female genital tract presented by Acién and revised by Acién et al, Sánchez-Ferrer et al, and Acién and Acién. Acién proposed that vagina has complete mesonephric (Wolffian) origin although its lining did reveal Müllerian cells–derived from Müllerian tubercle, while uterus and cervix were derived from fused paired paramesonephric ducts. Thus, formation of vagina does not include paramesonephric ducts although Müllerian tubercle does explain vaginal lining. This has been proved to an extent by data published by Sánchez-Ferrer et al in his experiments with female rats using histoimmunochemistry techniques.

Embryopathogenesis of HWWS can be temporally located around 8 weeks of gestation. Agenesis of ureteric bud results due to early failure of development of metanephric diverticulum from mesonephric duct (around 5 weeks). This leads to agenesis of ipsilateral ureter and kidney. Development and positioning of paired paramesonephric duct in close proximity is done by mesonephros. At around 9 weeks gestation, paramesonephric duct is positioned in such a manner that it is lateral to the mesonephric duct in the first part, crosses it anteriorly, and lies medial to it in the converging portion. If the Wolffian duct is absent, the Müllerian duct moves laterally (in space where the Wolffian duct is absent) and may not be merged with the contralateral duct, giving a uterus didelphys. Failure of development of distal hemivagina due to developmental arrest of ipsilateral mesonephric duct results in obstructed hemivagina. This occurs in OHVIRA syndrome. Introtitis is not compromised, because its embryological origin comes from urogenital sinus. Mesonephric remnant can stay in the vaginal wall and sometimes become Gartner’s cyst. All 3 components of OHVIRA syndrome, i.e., uterus didelphys, unilateral low vaginal obstruction, and ipsilateral renal agenesis, are secondary to mesonephric duct-induced MDA.

Obstructed hemivagina and ipsilateral renal anomaly syndrome is generally detected in newborns or shortly after menarche usually based on suspicion. It is detected in neonates if fetal ultrasonography (USG) showed unilateral renal agenesis or multicystic dysplastic kidney. It is recommended to seek Müllerian duct obstruction and look for genital abnormalities. Uterine anomalies are best detected in the neonatal period where uterus is still under maternal hormonal stimulation (prominent echogenic endometrium and myometrium on USG) and worse in childhood to menarche, where the uterus size and shape do not allow diagnosis. In adolescents, it may be suspected clinically but usually detected on imaging while presenting with non-specific complaints. Patients usually present with pelvic pain or dysmenorrhea, with
or without palpable mass due to hematocolpos or hematometra. Accurate and early diagnosis is essential, as incidence of complications including endometriosis, menstrual disorders, infertility, and obstetric complications increases with time. Endometriosis is a frequent complication because of the obstructed outflow of part of menstrual blood that collects in obstructed Müllerian derivatives, leading to hematocolpos, hematosalpinx, and peritoneal endometriosis, confirming the theory postulated by Sampson.

Ultrasound and MRI are most commonly used imaging modalities in diagnosis of MDA. Hysterosalpingography (HSG) is typically indicated in initial stages of infertility work-up. Ultrasonography is frequently used as initial investigation and preferred as it is non-invasive, free of radiation, and widely available but cannot identify types of MDA. Magnetic resonance imaging is ideal imaging modality for noninvasive evaluation of female pelvic anatomy because it provides more detailed information regarding uterine morphology, continuity with each vaginal lumen, character of vaginal septum, and nature of fluid contents. It is more useful in detection of associated conditions, such as, endometriosis, pelvic inflammation and adhesions, and coexisting urologic anomalies. Laparoscopy is gold standard diagnostic modality with additional advantages of therapeutic drainage of hematocolpos/hematometra, vaginal septectomy, and marsupialization. However, it should be reserved when diagnosis is not clear after imaging or when MRI is not available and should not be performed as a routine procedure. Decision to perform laparoscopy is based on interval between menarche and diagnosis, severity of symptoms, and presence of hematometra or pyometra.

Resection of the obstructed vaginal septum to relieve obstruction is treatment of choice for symptom relief and preservation of reproductive capability, also reducing chances of pelvic endometriosis due to retrograde menstrual seeding. If surgery is to be delayed due to some circumstances, menstrual suppression with combined oral contraceptive pills is advised to prevent further accumulation of hematocolpos and further hematometra. Integrity of hymen represents major cultural issue in some communities. Hysteroscopic excision of vaginal septum in uterus didelphys is recommended for management of those patients with good outcome. Reconstruction of vagina can be accomplished by careful excision of the vaginal septum. Patients are able to have a normal sexual life. Altchek and Paciu have reported pregnancy occurring twice in a previously obstructed didelphys uterus after surgical correction. Successful pregnancy is achieved eventually in 87% patients, while 23% have risk of abortions. Hence, the surgeon must make every effort to preserve the obstructed uterus. Hemihysterectomy done earlier is now no more preferred as the reported incidence of pregnancy in both horns is almost equal. Hemihysterectomy is only used in selected cases for which resection of the vaginal septum is not enough to relieve hematometocolpos or recurrent vaginal stenosis develops.

In conclusion, OHVIRA is a rare congenital complex Müllerian and Wolffian duct anomaly with simple single-stage surgical management, and MRI plays a major role in its diagnosis.

REFERENCES

2. Embrey MP. A case of uterus didelphys with unilateral gyn.