A Rare Case of Herlyn Werner Wunderlich Syndrome - Role of MRI in Diagnosis

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ABSTRACT

Herlyn werner wunderlich syndrome (HWWS) is a rare congenital mullerian ductal Anomaly (MDA) characterised by the triad of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis. It generally occurs at puberty and exhibits non-specific and variable symptoms with acute or chronic pelvic pain shortly following menarche, causing a delay in diagnosis. Moreover, the diagnosis is complicated by the infrequency of this syndrome, because mullerian ductal anomalies are infrequently encountered in a routine clinical setting. We report here a 15-year-old female presenting with secondary amenorrhoea and pain abdomen where magnetic resonance imaging (MRI) suggested the diagnosis of HWWS.

Keywords: Herlyn-Werner-Wunderlich syndrome, Magnetic resonance imaging, Mullerian ductal anomaly

INTRODUCTION

Herlyn werner wunderlich syndrome (HWWS) is rare and often a misdiagnosed entity and high index of suspicion is required in patients with mullerian and mesonephric duct anomalies. HWWS represents a type of mullerian duct anomalies (MDA) associated with mesonephric duct anomalies. MDA are congenital entities resulting from non developmental (agenesia or hypoplasia), defective vertical or lateral fusion, or resorption failure of the mullerian ducts.

The incidence of didelphys uterus, related to HWWS, is approximately 1/2,000 to 1/28,000, and it is accompanied by unilateral renal agenesis in 43% of cases. The incidence of unilateral renal agenesis is 1/1,100 and 25-50% of affected women exhibits associated genital abnormalities.

It usually presents at puberty with pelvic pain, but rarely can present in neonates or in adulthood in the form of pyometra, ischiorectal swelling, urinary obstruction, and primary infertility. Magnetic resonance imaging (MRI) is the modality of choice for the diagnosis of HWWS and other such anomalies because of better anatomic delineation of pelvic structures and higher sensitivity for blood products.

CASE REPORT

A 15 year old unmarried girl came with chief complaints of chronic pelvic pain, on and off amenorrhoea and pelvic mass since 8 months. She attained menarche at the age of 14 years. A bimanual physical examination indicated a right sided cystic and tender pelvic mass, movable, mildly tender to palpation. Routine haematological laboratory tests were normal. Further she was advised ultrasonography, which revealed a bulky distended
Figure 1: Sagittal and axial ultrasound images showing a bulky uterus with multiple thick internal echoes in endometrial cavity.

Figure 2: Sagittal T2 W image showing elongated and distended right cavity.

Figure 3: Sagittal T2 W image showing normal left uterine horn (arrow).

Figure 4: Axial T2 W images showing right and left uterine horns. Right uterine horn is distended with hyperintense collection, left uterine horn was normal.

Figure 5: Coronal T2 W image showing normal left kidney (arrow) with absent right kidney.

Figure 6: Postoperative axial image showing normal right and left uterine horns (arrows).
uterus with multiple low level internal echoes in the endometrial cavity (Figure 1) with absent right kidney. So, in view of uterine pathology and absent right kidney, Magnetic Resonance Imaging (MRI) was performed in order to evaluate the possible genito-urinary anomaly. The MRI examination was performed on a 1.5 Tesla MR clinical scanner (GE). MRI revealed two separate uterine cavities, cervices and vaginas, suggestive of uterus didelphys. The right uterine cavity and cervical canal were dilated and filled with fluid which was hyperintense on both T1W and T2W images (s/o blood products) [Figures 2 and 4].

In addition, there was absent right kidney [Figure 5]. The left uterine cavity, cervix, and vagina were normal [Figure 3]. Hence, a diagnosis of uterus didelphys with right hematometra, hematocolpos, and right hemivaginal obstruction was made. Considering that she also had absent right kidney, a final diagnosis of HWWS or obstructed hemivagina ipsilateral renal anomaly (OHVIRA) syndrome was made. A Cruciate incision was made in right vaginal septum and the margins of incision were everted and about 1000 ml of chocolate-coloured fluid was drained. Subsequently an MRI was done post-operatively 10 days after the surgery, which revealed normal right uterine cavity and cervix (Figure 6) with minimal T2 hyperintense collection in right cervical canal (Figure 7).

**DISCUSSION**

The most frequently used system for classification of Mullerian duct anomalies (MDAs) was proposed by Buttram and Gibbons, which classifies them into six Categories [7] [Table 1]. HWWS is a combination of Type III Mullerian anomaly with mesonephric duct anomaly with vaginal septum. The classic renal manifestation of OHVIRA syndrome is ipsilateral renal agenesis, but reports of duplicated kidneys, dysplastic kidneys [8], rectovesical bands [9] or crossed fused ectopia [10] have also been described. Uterus didelphys with obstructed hemivagina is due to lateral non-fusion of the Mullerian ducts with asymmetric obstruction, and it is almost always associated with renal agenesis ipsilateral to the side of obstruction. [8] Embryopathogenesis of HWWS

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**Table 1: Mullerian duct anomalies classification**

<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 Mullerian agenesis</td>
<td>Hypoplasia of the tubes, uterus, cervix, or vagina</td>
</tr>
<tr>
<td>II</td>
<td>Unicornuate uterus or unilateral Mullerian anomaly</td>
<td>No rudimentary horn</td>
</tr>
<tr>
<td>III</td>
<td>Uterus didelphys</td>
<td>Two separate uterine horns and two cervices</td>
</tr>
<tr>
<td>IV</td>
<td>Bicornuate uterus</td>
<td>Complete (septum extends to the internal or external os)</td>
</tr>
<tr>
<td>V</td>
<td>Septate uterus</td>
<td>Complete (septum extends to the internal os)</td>
</tr>
<tr>
<td>VI</td>
<td>T-shaped uterus</td>
<td>Results from in utero exposure to diethyl stilbestrol</td>
</tr>
</tbody>
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![Figure 7: Postoperative axial image showing minimal T2 hyperintense collection in right cervical canal (arrow).](image-url)
CONFLICT OF INTEREST

The authors declare no conflict of interest.

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REFERENCES


