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Case Report

Uterus Didelphys with Obstructed Right Hemivagina and Ipsilateral Renal Agenesis (Herlyn-Werner-Wunderlich Syndrome). A Rare Congenital Urogenital Anomaly

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Abstract

Herlyn-Werner-Wunderlich syndrome is a rare congenital urogenital anomaly of the Müllerian duct. It is presented with double uterus, unilateral obstructed hemivagina and ipsilateral renal agenesis, also known as OHVIRA syndrome. The potential complications of Herlyn-Werner-Wunderlich syndrome are either acute complications as pyohematocolpos, pyosalpinx, or peritonitis, or long-term complications as endometriosis, pelvic adhesions which increase the risk of abortion or infertility.

Keywords: *Herlyn-Werner-Wunderlich Syndrome; OHVIRA Syndrome; Uterus Didelphys; Renal Agenesis; Mullerian Duct, Ultrasound; MRI*

Background

Herlyn-Werner-Wunderlich syndrome is a rare congenital urogenital anomaly of the Mullerian duct. It is presented with double uterus, unilateral obstructed hemivagina and ipsilateral renal agenesis (OHVIRA syndrome). The exact incidence is not known, but it has been reported between 0.1% and 3.8 % [1].

Case Report

An 18-year old female was a known case of congenital left renal agenesis and solitary right kidney. The patient came to the emergency department complaining of severe lower abdominal and pelvic pain. Transabdominal ultrasound demonstrated right sided uterine body measuring 4.3 x 1.8 cm with central endometrium measuring 0.3 cm, ipsilateral cervix and vagina (Figure 1). Left sided uterine shape structure with a cyst-like appearance containing echogenic fluid was seen on the left side of the lesser pelvis, posterior to the distended bladder. It measured 14 x 8.2 cm. It represented distended cervix and vagina due to obstructed left hemivagina (Figure 2). Both ovaries were presented normal. The right kidney was demonstrated normal but the left kidney was absent (Figure 3). MRI examination was recommended for further evaluation. It showed uterus didelphys (Mullerian duct anomaly type III), associated with transverse vaginal septum causing obstruction of a single vagina and extending to a dimension of 13 cm by retained hemorrhagic fluid. Two cervices and two uteri were identified separately. On the left side, the cervix and uterus were distended with hemorrhagic fluid causing thinning out of the myometrium of the left uterus (Figure 4 and 5). On the right side, the normal uterine horn was seen (Figure 6). Moderate amount of fluid was seen within the pelvic peritoneal recesses. The ovaries were seen bilaterally. The left kidney was confirmed absent (Figure 7). The right kidney showed signs of compensatory hypertrophy (Figure 7).

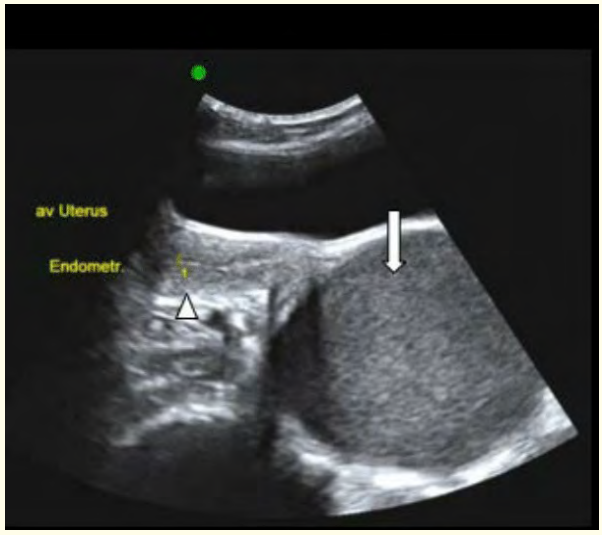

Figure 1: *Ultrasound: Dilated cervix filled with turbid fluid (arrow). Normal right uterine horn (arrowhead).*

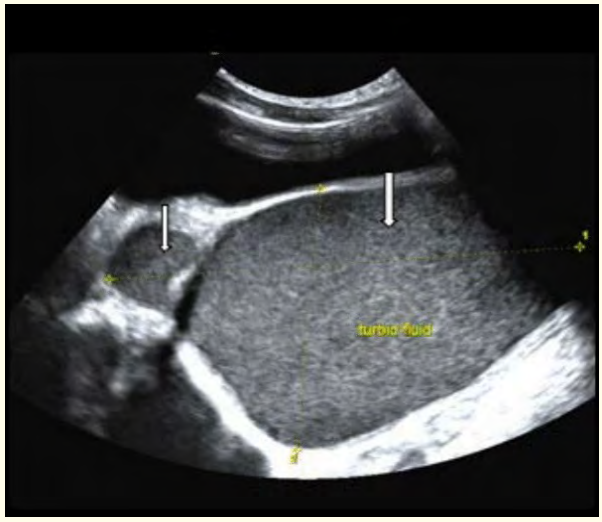

Figure 2: *Ultrasound: Dilated vagina and cervix (arrows) filled with turbid fluid due to obstructed left hemivagina.*

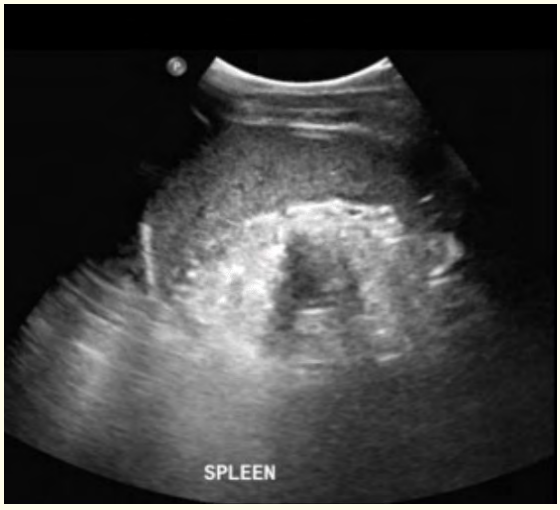

Figure 3: *Ultrasound: Absence of the left kidney in its usual anatomical location (N.B. the left kidney is not seen in the other anatomical locations).*

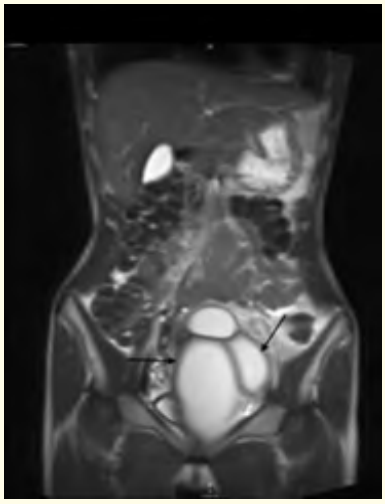

Figure 4: *MRI: Coronal T2WI shows dilated vagina, cervix and left uterine horn with high signal intensity blood (arrows).*

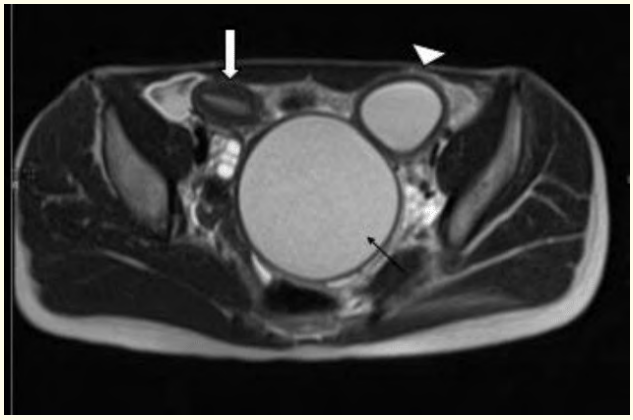

Figure 5: *MRI: Axial T2WI shows dilated vagina (thin arrow), left uterine horn (arrowhead) and normal right uterine horn (arrow).*

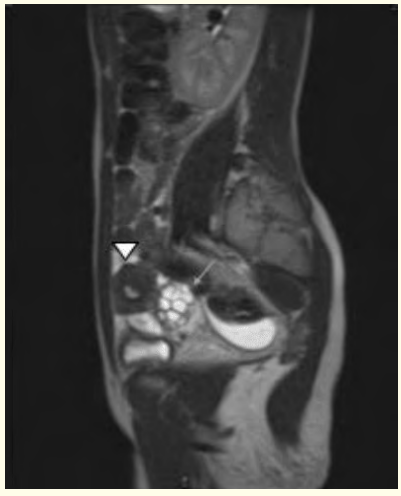

Figure 6: *MRI: Sagittal T2WI shows normal right uterine horn (arrowhead) and normal right ovary (arrow).*

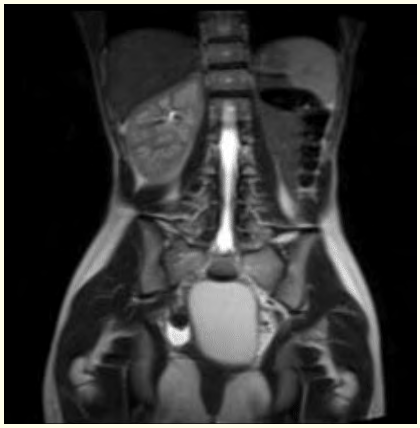

Figure 7: *Coronal T2WI shows absent left kidney and normal position right kidney.*

Based on ultrasound and MRI findings which showed uterus didelphys (Mullerian duct anomaly III) associated with transverse vaginal septum, hematocolpos left hematometra, normal ovaries and the unilateral renal agenesis, OHVIRA or Herlyn-Werner-Wunderlich syndrome was diagnosed.

Discussion

Mullerian duct anomalies incidence occurs in 2 - 3%, while if associated with obstructed hemivagina and ipsilateral renal anomaly, the incidence is 0.1 - 3.8% of all such anomalies [1].

Herlyn-Werner-Wunderlich syndrome is a congenital anomaly of the Müllerian ducts associated with defect in the development of the mesonephric ducts, so it is the triad of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis [2].

The definite cause and pathogenesis of Herlyn-Werner-Wunderlich syndrome is still not known, but it is considered to be anomalous Müllerian (paramesonephric) and Wolffian (mesonephric) duct development [3]. The Wolffian ducts giving origin to the kidneys and also is inductor elements for adequate Müllerian ducts fusion. Therefore, developmental anomaly of the caudal portion of one of the Wolffian ducts cause unilateral renal agenesis and associated with imperforate hemivagina [2].

Although ultrasound, CT, and hysterosalpingography are commonly used for the diagnosis of Herlyn-Werner-Wunderlich syndrome because their cost is low, MRI is the most accurate diagnostic method. It is helpful to the patients to maintaining their fertility [4]. MRI will demonstrate two completely separate uteri, each has normal zonal anatomy with endometrial cavity and normal endometrial myometrium ratio and it also shows two separate cervices and two hemivagina without communication between the each hemi uterus and hemivagina [4].

This rare syndrome is usually discovered at the time of puberty, especially after menarche. Recurrent and increasing pelvic pain is the clinical symptoms in most patients. Hematocolpos is a result of retained partially clotted longstanding menstrual blood in the obstructed hemivagina and clinically is detected as a mass in the pelvis [5].

Herlyn-Werner-Wunderlich syndrome is the least common anomaly of Müllerian duct anomalies, but it has the best prognosis [5].

The close differential diagnosis of Herlyn-Werner-Wunderlich syndrome is bicornuate bicollis uterus associated with hemivaginal atresia. In such cases imaging demonstrates two widely separated uterine horns, and the distance between two horns is greater than 4.5 cm with the presence of a deep fundal cleft greater than 1 cm [6].

The potential complications of Herlyn-Werner-Wunderlich syndrome are either acute complications as pyohematocolpos, pyosalpinx, or peritonitis, or long-term complications as endometriosis, pelvic adhesions which increase the risk of abortion or infertility [7].

Conclusion

Herlyn-Werner-Wunderlich syndrome or also named as OHVIRA syndrome is the least common anomaly of the Müllerian ducts but it carries the best prognosis. US and MRI play important role in the diagnosis of this syndrome. Early recognition prompts surgical excision of the obstructing vaginal septum, with rapid relief of symptoms and prevention of the complications.

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