Obstructed Hemivagina and Ipsilateral Renal Agenesis (OHVIRA)

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Figure 1. USG shows two widely separated uterine cornua, a non-obstructed normal appearing left uterus [Fig. 1a], and hematometra, hematosalpinx in right uterus [Fig. 1b], normal right ovary and hematometra in right uterus [Fig. 1c].

Figure 2. Axial T2W AND T1W images show widely divergent uterine horns, non-obstructing normal left uterus, and hematometra and hematosalpinx in obstructed right uterus.
Figure 3. Sagittal T2WI images reveal two separate uterine corpi with right blind ended uterus. Figure 3b shows non-obstructed uterus opening into vagina.

Figure 4. Coronal T2WI images of abdomen show ipsilateral renal agenesis on right side.
Figure 5. Schematic diagram shows uterine didelphys with unilateral vaginal agenesis and ipsilateral renal agenesis.

Diagnosis: Uterine didelphys with unilateral vaginal agenesis and ipsilateral renal agenesis (OHVIRA)
Findings: Ultrasound findings show two widely separated uterine corpi, a non-obstructed normal appearing left uterus and cervix, and hematometra, hematosalpinx in right uterus.
MRI findings include widely divergent uterine horns, non-obstructing normal left uterus and cervix opening into vagina which communicates with external opening, and hematometra and hematosalpinx in obstructed right uterus which ends as a blind end cavity and does not show any communication with vagina suggestive of vaginal agenesis.
Additional imaging of urinary system shows renal agenesis on right side.
Discussion: Mullerian duct anomalies are congenital anomalies which results from non-development, defective vertical or lateral fusion, or resorption failure of mullerian (paramesonephric ) ducts. MDA have been classified by American Society of Reproductive Medicine according to patients clinical presentation, treatment and prognosis(1).
I Müllerian agenesis or hypoplasia
II Unicornuate uterus
III Uterus didelphys
IV Bicornuate uterus
V Septate uterus
VI Arcuate uterus
VII DES-related uterine anomalies

Classification of Vaginal Anomalies
I Transverse vaginal septum (obstructing or non-obstructing)
II Longitudinal vaginal septum (obstructing or non-obstructing)
III Stenosis or iatrogenic cause

OHVIRA syndrome is typically associated with type III MDA (uterine didelphys) with two uteri, two cervixes and two vaginas, one of which is obstructed, and associated with renal agenesis on the side of hemivaginal obstruction, but this case is a variant of described OHVIRA as it was renal agenesis with ipsilateral complete vaginal agenesis instead of vaginal obstruction by transverse (type I) or longitudinal (II) vaginal septum.

OHVIRA results from embryonic arrest at eight weeks of gestation which affects adjacent mullerian and metanephric ducts. The defect may also occur as early as the fourth week of gestation and can affect both the mesonephric ducts and ureteral buds. The mesonephric duct, which is maldeveloped, does not allow crossover of the mullerian duct and consequent fusion results in a didelphys uterus and obstruction of the ipsilateral horn and the vagina.

Clinically, patients with OHVIRA usually present due to symptoms from obstruction. Almost uniformly this presentation occurs at or after menarche when the hemiuterus or hemivagina becomes symptomatic and the patient experiences cyclical dysmenorrhea. This results in retrograde menstrual flow and may present as a unilateral abdomino-pelvic mass on physical examination. Complications associated with this disorder are endometriosis, pyosalpinx, pyocolpos and pelvic adhesions.

Imaging is usually done when patient presents with complaints of abdominopelvic pain and cyclic dysmenorrheal. Since the other non obstructive uterus is normally functional, infertility is a rare presentation. Ultrasound findings include two widely separated uterine corpii, a non-obstructed normal appearing uterus and cervix opening into vagina, hematometra and hematosalpinx in other uterus, however exact cause of obstruction is difficult to evaluate on sonography and further imaging is required.

MRI is the imaging investigation of choice in such cases due to multiplanar capabilities, high soft tissue contrast and no risk of radiation. MRI findings include non-obstructing normal uterus and cervix opening into vagina which communicates with external opening. Hematometra and hematosalpinx in obstructed uterus which ends as a blind end cavity, and does not show any communication with vagina suggestive of vaginal agenesis.

Imaging of urinary system is imperative to perform in presence of mullerian duct anomalies or vice versa. Differential diagnosis is uterine didelphys with hemivaginal obstruction by transverse or longitudinal vaginal septum or imperforate hymen.

References
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