CASE REPORT

OHVIRA Syndrome

Priya Pillai[1], Rana Abualsaud[2], Zainab Awni[3], Arash Rafii[4], Badreldeen Ahmed[5]

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A 14-year-old girl came to the gynecology clinic with recurrent periumbilical pain migrating to the right iliac fossa for the last 3 weeks. The pain was constant and more severe toward the end of her menstrual cycle. She did have a few episodes of vomiting over the last 3 weeks but she did not have any fever, diarrhea, melena, or hematochezia. However, she did mention episodes of increasing urinary frequency over the past few days but denied any dysuria or hematuria.

Menarche was achieved at the age of 13 and she has been having regular menstrual cycles every 28 days and lasting for 3–4 days. She denied any dysmenorrhea or menorrhagia.

She did not have any prior medical or surgical history.

Physical examination was remarkable for mild tenderness in the right iliac fossa but no rebound tenderness or guarding noted. Pelvic examination was not performed. Transabdominal pelvic ultrasound imaging (Fig. 1) revealed uterine didelphys with a right hematocolpos due to distal obstruction from a transverse vaginal septum. The renal ultrasound revealed an absent right kidney but a normal left renal architecture with compensatory hypertrophy. Magnetic resonance imaging (MRI) of the pelvis without contrast (Fig. 2) confirmed the uterine didelphys with two uterus, two cervices, and two vaginal canals.

The right vaginal canal was noted to be distended with a complex fluid suggestive of hematocolpos due to distal obstruction most likely from a transverse vaginal septum.

Laboratory examination was unremarkable with no leukocytosis and normal liver and renal function. Urinalysis was unremarkable and urinary beta hCG was reported as negative.

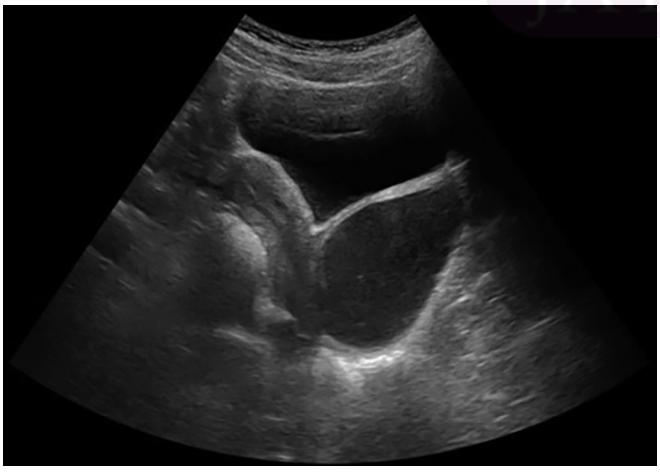

Fig. 1: Transabdominal pelvic ultrasound (sagittal view) demonstrating uterine didelphys with right hematocolpos due to distal obstruction from a transverse vaginal septum. The vagina containing the echogenic material

[1]Weill Cornell Medicine-Qatar, Education City, Doha, Qatar
[2]Qatar University-Medical School, Qatar
[3]Feto Maternal Center, Doha, Qatar
[4]Weill Cornell Medicine-Qatar, Education City, Doha, Qatar; Qatar University-Medical School, Qatar
[5]Weill Cornell Medicine-Qatar, Education City, Doha, Qatar; Qatar University Medical School, Qatar; Feto Maternal Center, Doha, Qatar
Corresponding Author: Badreldeen Ahmed, Weill Cornell Medicine-Qatar, Education City; University Medical School; Feto Maternal Center, Doha, Qatar, Phone: +974 55845583, e-mail: profbadreldeen@hotmail.com

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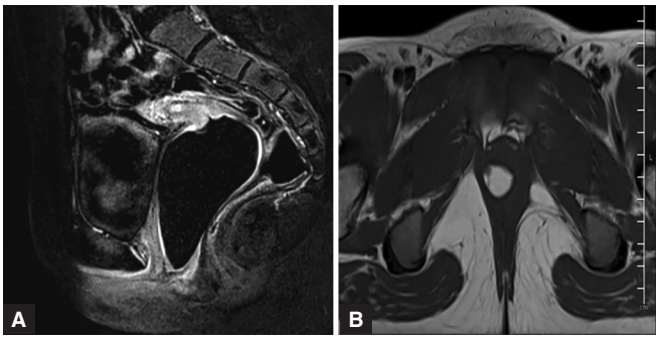
Upon imaging findings, she was offered a surgical excision of the transverse vaginal septum to relieve the hematocolpos and the subsequent pain from the blood accumulation. The procedure was done under general anesthesia where in the distended hemivaginal vault was incised to allow drainage of the old stagnant blood. The transverse vaginal septum was then excised with primary closure. Her abdominal pain subsided subsequently and she did not require any further surgeries.

Discussion

Obstructed hemivagina with ipsilateral renal agenesis syndrome is a rare condition among women which originates from the failure of the two Mullerian tubes to fuse during the 10th week of gestation.[1,2] Similar to many Mullerian tube anomalies, Obstructed hemivagina



Figs 2A and B: MRI pelvis T2 without contrast view (A) and sagittal view (B). MRI pelvis T2 with a contrast Coronal view demonstrating complex fluid hemivagina

and ipsilateral renal anomaly (OHVIRA) syndrome is usually diagnosed at the time of menarche when slow accumulation of blood from menses remains obstructed in one of the vaginal vaults. The resulting hematocolpos causes distention of the uterus and fallopian tubes subsequently. The local pressure as a result of this distention on surrounding structures causes the pain in the abdomen. Clinically, it is often misdiagnosed as appendicitis but the presence of unilateral kidney should prompt an inspection of OHVIRA syndrome.[3,4]

The urinary and genital systems arise from a common ridge of mesoderm arising along the dorsal body wall. The upper vagina develops from the paramesonephric duct and the lower vagina from the urogenital sinus. This development also relies on the normal development of the mesonephric system. Hence, abnormal differentiation of the mesonephric and paramesonephric ducts may also be associated with anomalies of the kidneys. Renal agenesis is the most common anomaly, although horseshoe or pelvic kidney, cystic renal dysplasia, duplication of the collecting system, and ectopic ureters have all been described.

Delay in the diagnosis can lead to complications that include endometriosis, adhesions, infertility, and infectious complications due to chronic cryptomenorrhea.[5] Delays in diagnosis have been attributed to lack of understanding of this condition by radiologists, gynecologists, urologists, nephrologists, pediatricians, and pediatric surgeons.

Diagnosis of OHVIRA syndrome is mainly from a clinical suspicion supported with imaging evidence. If any imaging modality reveals a unilateral renal agenesis in a prepubertal or peripubertal age, the clinical suspicion of Mullerian development anomalies (MDA) is high. These MDAs should be evaluated by a transabdominal ultrasound imaging of pelvis with careful visualization of the uterus and ovaries. The abdominal and pelvic ultrasound cannot visualize the vaginal septum. If improper visualization occurs with the pelvic ultrasound imaging, then MRI of pelvis with a 1.5 T or greater magnetic field strength can be used to assess the reproductive anatomy.[6,7] Furthermore, regular menstruation in the context of an incomplete vaginal outlet obstruction and slow extension of hematocolpos can also lead to delayed diagnosis.[8]

The management of OHVIRA syndrome begins with initial incision of the vaginal bulge to relieve the obstructed hematocolpos. Often, a vaginoplasty is required to excise the vaginal septum if it was the cause of the obstruction. However, hemihysterectomy of the obstructed uterus is not recommended anymore as studies have shown that pregnancy in a previously obstructed uterus is still viable.[9] Every effort should be made to preserve the uterus. Postoperative complications of vaginoplasty include vaginal stenosis, vaginal stricture, and potential re-closure of the vaginal septum.[10,11]

In conclusion, the case at hand is a unique presentation of OHVIRA syndrome with a transverse septum. The earlier the surgery, the better the outcome is. A simple vaginal incision, not affecting the integrity of the hymen, can be very effective in relieving the symptoms.

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