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Obstructed hemivagina with ipsilateral renal agenesis (OHVIRA) syndrome with imperforate anus

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ABSTRACT

OHVIRA syndrome (Obstructed hemivagina with ipsilateral renal agenesis) is a rare Mullerian duct anomaly that results in uterine didelphys, obstructed vaginal vault, and unilateral renal agenesis. We report an 11 year old girl with a history of imperforate anus presenting with progressive abdominal pain. This represents the first documented case of OHVIRA syndrome in association with an ano-rectal malformation.

OHVIRA syndrome (Obstructed hemivagina with ipsilateral renal agenesis), also known as Herlyn-Werner-Wunderlich (HWWS), is a rare Mullerian duct anomaly that results in uterine didelphys, obstructed vaginal vault, and unilateral renal agenesis. It may present at birth or soon after the beginning of menses as fluid collects in the obstructed vaginal vault. Literature review describes association with acute abdomen presentation and rare reports of intestinal malrotation or abdominal vascular anomalies [1,2]. We describe a case with the first documented association of OHVIRA syndrome with an anorectal malformation.

1. Case report

An 11-year-old girl presented with five days of acute colicky abdominal pain. At presentation, she was afebrile without back pain, dysuria, chills nor vaginal discharge. Menarche occurred six months prior and her last menstrual period was two weeks prior. She denied dysmenorrhea and menorrhagia. Prior history included primary anoplasty repair for imperforate anus, solitary left kidney with pyelonephritis and intermittent constipation. She was admitted for concern for subacute obstruction and bowel clean-out for constipation.

Laboratory examination indicated leukocytosis but otherwise normal renal and liver function. Urinalysis and urine culture were normal and urinary hCG was negative. Nuclear renogram (Fig. 1) and renal ultrasound confirmed the solitary left kidney with compensatory hypertrophy noted. Abdominal plain films showed a dilated rectum initially suspected for constipation (Fig. 2).

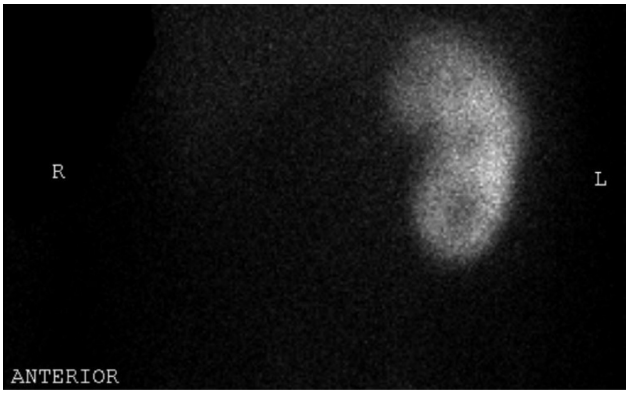

Fig. 1. TC-99 m DMSA kidney scan demonstrates absent right kidney.

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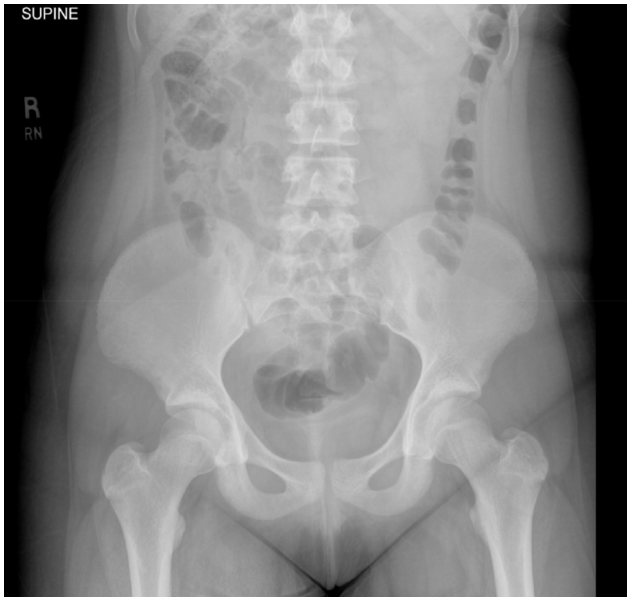

Fig. 2. Moderate stool burden with non-obstructive bowel gas pattern.

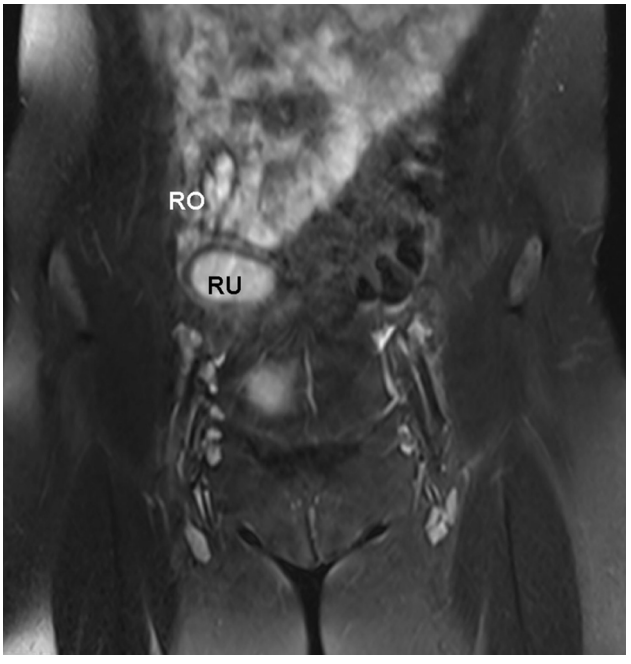

Fig. 4. T2-weighted, fat-saturated coronal view of the pelvis demonstrates the fluid-filled right uterus (RU) and the right ovary (RO).

Transabdominal pelvic ultrasound revealed a fluid-filled structure posterior to the bladder thought to represent hematocolpos. Abdominal magnetic resonance imaging (MRI) without contrast re-demonstrated the unilateral left kidney. Pelvic MRI without contrast demonstrated uterus didelphys with two vaginas, two uteri and two ovaries (Figs. 3–7). Both vaginas and the right uterus and right fallopian tube were distended with complex fluid.

She started a course of prophylactic antibiotics and started oral contraceptive pills to further suppress menstrual. Two weeks thereafter, she underwent cystoscopy, vaginoscopy and vaginoplasty under general anesthesia. Cystoscopy was normal except for deviation of the bladder anteriorly due to extrinsic compression and a single ureteral opening on the left consistent with her right renal agenesis. Vaginoscopy identified the patient's right hemivaginal bulge (Fig. 8) compressing the left cervix. The hemivaginal bulge was incised (Fig. 9) allowing drainage of the hematocolpos. The vaginal septum was then excised with primary closure to allow confluence of the formerly obstructed right hemivagina and the patent left hemivagina (Fig. 10). Her pain has subsequently improved and there is no current plan for further surgery.

2. Discussion

Obstructed hemivagina with ipsilateral renal agenesis (OHVIRA) syndrome is a rare entity among females resulting from failure of

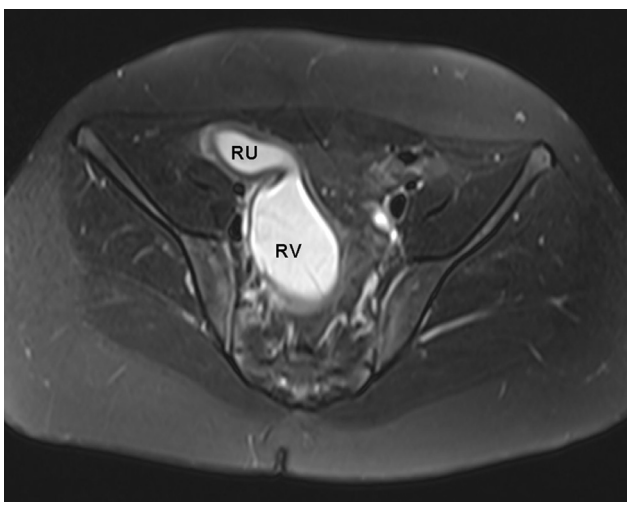

Fig. 3. T2-weighted, fat-saturated axial image of the pelvis demonstrates the right uterus (RU) and dilated right vagina (RV).

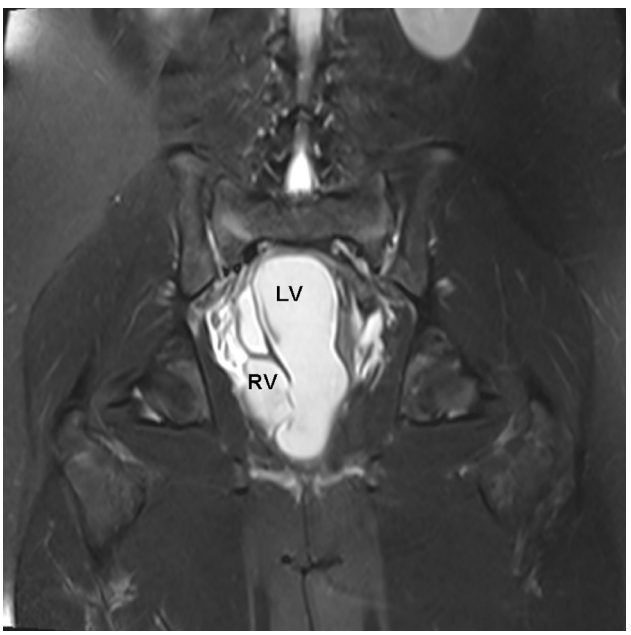

Fig. 5. T2-weighted, fat-saturated coronal view of the pelvis demonstrates the dilated right vagina (RV) and the more dilated left vagina (LV).