A Rare Case of OHVIRA Syndrome with Urethral Stenosis

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ABSTRACT
Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) is a rare anomaly of the urogenital system. The characteristic triad of this syndrome, which was initially reported in 1950, is didelphys uterus, obstructed hemivagina, and ipsilateral renal agenesis (Embrey). The prevalence of congenital Müllerian duct anomalies is reported to be 1%. A 24-year-old girl got admission to MGH on 15.3.2019 with lower abdominal pain and difficulty in the passing of urine. Her menarche was established 6 months back with a regular cycle and associated with dysmenorrhea. Magnetic resonance imaging (MRI) established the diagnosis of OHVIRA. She underwent surgery for drainage of the hematocolpos and excision of the vaginal septum and urethral dilatation with cystoscopy followed by an uncomplicated recovery and the patient had normal menstrual cycles after surgery.

Keywords: Hemivagina, Ipsilateral renal agenesis, Müllerian anomalies, Uterus didelphys.

INTRODUCTION
The prevalence of congenital Müllerian duct anomalies is reported to be 1%. The syndrome of obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) is a rare congenital anomaly of the Müllerian ducts (paramesonephric) and Wolffian structures (mesonephric). It usually includes unilateral renal anomalies and uterine didelphys. Moreover, the menstrual flow that comes from the patent hemivagina resembles normal menses leading to delay in diagnosis and surgery. The exact cause and pathogenesis are uncertain; however, the diagnosis and treatment at an early stage can relieve acute symptoms and preserve normal fertility.

CASE DESCRIPTION
A 24-year-old unmarried girl admitted in Gynae and Obs Department of Mahatma Gandhi Medical Hospital with lower abdominal pain and difficulty in passing urine. The pain was not related to fever, vomiting, dysuria, or loin pain. Her menstruation is normal with a regular cycle, scanty flow, and associated with dysmenorrhea. She did not have any significant medical or surgical history. Abdominal examination revealed mild lower abdominal tenderness without rebound or guarding. Vaginal examination demonstrated a bulge in the right upper vagina. USS findings were suspicious for right-sided renal agenesis and uterus didelphys. A subsequent magnetic resonance imaging (MRI) demonstrated the classical findings of right-sided OHVIRA.

After proper counseling with the patient’s family, cystoscopy examination was a plan under anesthesia, but there was urethral stenosis present. So cystoscope could not be negotiated, hence urethral dilatation was performed with female urethral dilators serially. After that cystoscopy was done. In cystoscopy, only left side hemitrigone was found, RT ureteric orifice and hemitrigone not seen, only left ureteric orifice visualized, a ureteric catheter placed in the left ureter, RGP has done.

After that following procedure was done by the gynecology department—an incision was made over the vaginal wall. A 700 mL of inspicted blood was escaped out. A Foley catheter was introduced through the cervix to keep patency of the outflow tract (Figs 1 to 5).

The patient was discharged on the third postoperative day with antibiotics and an intrauterine catheter. She came for a follow-up after 10 days when the catheter was removed. The patient came

Fig. 1: Image shows urethral stenosis
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for follow-up after 1 month. Her menstruation was regular with normal flow and duration.

**Discussion**

Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) is a rare form of Müllerian tract abnormality. The prevalence of congenital Müllerian duct anomalies is reported to be 1%. The incidence of OHVIRA syndrome is still unknown, with only few published case reports so far. Müllerian (paramesonephric) duct anomalies are congenital anomalies of the female genital tract which result from non-development or non-fusion of the Müllerian ducts or failed resorption of the uterine septum during the sixth to ninth weeks of fetal life causing a wide-ranging series of reproductive ducts malformations. The most common renal anomaly associated with this syndrome is renal agenesis, although other forms of renal malformations have been reported, including dysplastic kidney and renal duplication.
The clinical presentation of Müllerian duct anomaly varies widely in symptoms and age at onset. Several presentations have been reported, including pre-pubertal with urinary incontinence,8 adolescence with dysmenorrhea, difficulty with sexual intercourse or using a tampon, as well as a delayed diagnosis in pregnancy due to cervical incompetence9 and a rare case of a ruptured gravid uterus.10 Early diagnosis is important to relieve symptoms and to avoid adverse effects. Timely treatment will prevent adverse obstetric outcomes such as miscarriage, preterm birth, malpresentation, fetal growth restriction, and cesarean delivery.11,12 Preconception counseling is important, as is antenatal care in a high-risk setting. Associated renal anomalies also confer an increased risk of hypertensive disease in pregnancy as well as long-term implications for chronic renal and cardiovascular disease.13

Preoperative imaging with MRI is essential to define the Müllerian duct structures and renal tract. It also assists with surgical planning.14 Laparoscopy is still the gold standard for investigation of gynecological congenital abnormalities but is only used if MRI fails to establish a diagnosis or MRI is unavailable.15 Treatment of OHVIRA is vaginoplasty. It is ideally a single surgery with drainage of the obstructed hemivagina and resection of the septum to restore normal vaginal function.

CONCLUSION

OHVIRA syndrome is an uncommon congenital anomaly with clinical significance. It is typically associated with didelphys uterus with two cervices and two vaginas, one of which is obstructed. The obstruction usually occurs on the same side as the renal anomaly. Imaging particularly MRI, as in this case plays a major role in diagnosis which could be missed clinically. An early correct diagnosis is a goal to relieve the symptom and prevent complications.

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