CASE REPORT

Regular Menstruation with Outflow Track Obstruction (Herlyn-Werner-Wunderlich Syndrome)

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Abstract

Backround: Uterus didelphys with obliterated half-vagina and absent kidney on the same side is a syndrome with very low incidence known as Herlyn-Werner-Wunderlich syndrome (HWW) and also known as obstructed half-vagina and ipsilateral renal anomaly (OHVIRA).[1,2]

Case report: Sixteen-year-old female presented with cyclical pain in abdomen mostly from Day 1 on her periods since menarche since 1–2 years. She had regular menstrual cycle with dysmenorrhea. Clinical examination was done, and on ultrasound, we diagnosed it as HWW syndrome.

Intraoperatively, a bulge was arising from right vaginal wall, and on left side, cervix was felt with normal size uterus. Incision was taken on the bulge, about 60cc of mucous discharge was drained. We confirmed uterine didelphys. Later suturing was done on the opening of the right vagina, to keep it patent.

Conclusion: We should think of HWW syndrome in patients who have cyclic pain in abdomen and also in newborn cases with any renal abnormalities. Early and prompt surgical therapy prevents further complications.

Clinical significance: Early diagnosis and treatment prevents complications such as endometriosis and infertility.

Keywords: Didelphys, Hemivagina, Herlyn-Werner-Wunderlich syndrome.

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Backround

HWW syndrome is a type of Müllerian duct anomaly with very low incidence and consists of uterus didelphys with obstructed half-vagina and same side kidney being absent. Most commonly, patients present with pain in lower abdomen, pain during menstruation, and abdominal lump because of hematocolpos.[3,4] Purslow first described this syndrome in a young woman who presented with gradually increasing pelvic pain and a pelvic mass with regular menstruation.[5] The diagnosis of HWW syndrome is difficult as the incidence is less. Early diagnosis and surgical treatment can avoid complications. The chances of conception are good after treatment. A differential diagnosis should be kept in mind in patient with renal anomalies. Complications such as endometriosis and infertility can occur in future if diagnosis is delayed.

Detailed investigation has to be done to diagnose the type of anomaly. When a definitive diagnosis is concluded, many treatments are available.

Case Report

A sixteen-year-old girl presented to out patient department (OPD) with complains of dysmenorrhoea mostly from Day 1 on her periods since menarche (3 years) and also had difficulty in passing urine for 1–2 years.

Her menarche being 13 years, every month she had bleeding for 3–4 days with pain in abdomen. Last menstrual period (LMP) was 15 days back.

On examination, she was stable and also examined for her secondary sexual characteristic which was found to be normal. On per abdominal examination, the abdomen was soft and non tender. No mass was felt. Local examination of external genitalia was normal. A small bulge was visible at introitus.

On ultrasonography, there was absence of right kidney, didelphic uterus, hypoechoic collection with coarse internal echoes, and fluid in right half of vaginal canal which was suggestive of hematocolpos (Fig. 1).

Our preoperative diagnosis—hematocolpos? We decided to examine her under anesthesia. Intraoperatively, per vaginal examination was done and we noted a bulge arising from right vaginal wall (Fig. 2) and cervix felt on left side along with uterus which was normal in size and anteverted. Per speculum was done, and same findings were noted.

Foleys was inserted. Decision was made to open the bulge. Fifty to sixty cc of dirty mucoid collection drained. Scope was introduced through vagina and uterus on the right side. We found that there were two vaginas, and there was no connection in between.

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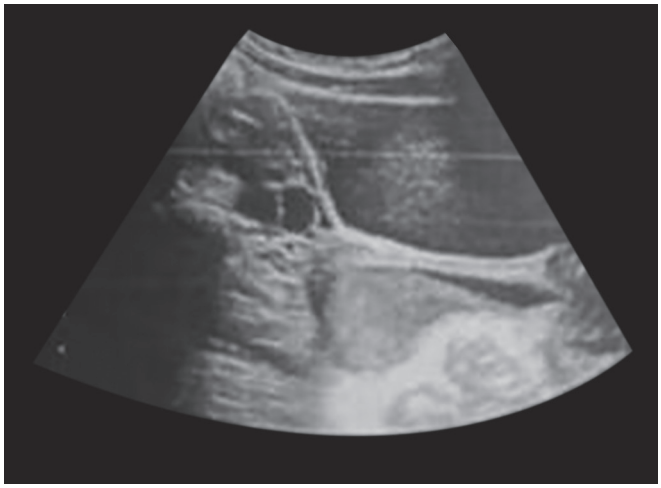

Fig. 1: USG showing hematocolpos in right vaginal canal

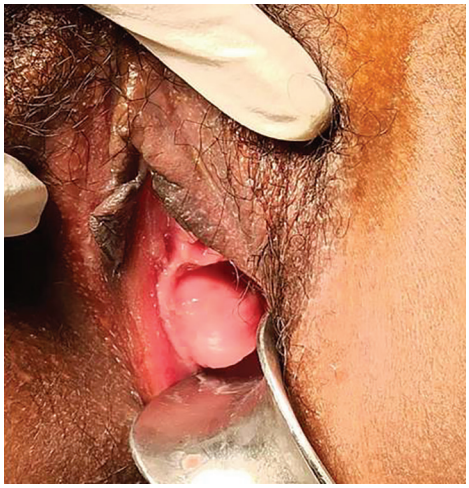

Fig. 2: Bulge arising from right vaginal wall

Hysteroscopy findings were on right side. There was no endometrium, and ostia was seen on left side. Vaginoscopy was done and left cervix was visualized. Later, circumferential suturing (interrupted) was done on the opening of the right vagina to keep it patent. Post-op was uneventful.

Discussion

Paramesonepheric duct anomalies incidence is more common in developmental anomalies, which may have absent uterus/vagina; the two ducts may not fuse, or septum between them may persist. HWW syndrome has very low incidence in which there are three things of two uterine cavities and vaginas, obliterated half-vagina, and same side kidney being absent. It is comes under type III Müllerian duct anomaly (MDA) with kidney abnormalities accounts for 5% of MDAs.

For the normal menstruation to occur, there should be intact hypothalamo–putitary axis which should act on ovaries and a patent outflow tract for the menstrual blood to flow out. But in this condition normal menstruation with outflow tract obliterated.

This was first described in 1922; the exact cause is not known. Incidence reported is 0.1–3.8%. Uterus didelphys constitutes 11% of paramesonephric duct anomalies. Small or absence of the uterine cavity and upper vagina accounts for 5–10% of paramesonephric duct. Associated kidney malformation is present in approximately 43%.

Seventy-five percent of cases with two uterine cavities have a minimum or maximum septum in between the vagina which is most commonly longitudinal in the HWW syndrome, which tells us that there is problem in lateral fusion in between the lower portion of two paramesonephric ducts. Hox gene and Wnt gene play an important role in uterine development.

The association of two uterine cavities with an obliterated half-vagina can be explained by a defect in development at the 8th week of gestation that involves the kidney and paramesonephric ducts; there is failed fusion of paramesonephric ducts, or septum may persist. Other anomalies may coexist renal dysplasia and doubling of the mesonephric duct.[6,7]

Often patients present with mass in the abdomen because of blood collected in vagina and uterus during menstruation, pain during menstruation, or as sudden-onset abdominal pain. Sometimes, the blood which is clotted becomes infected and pus gets collected which can lead to inflammation in the pelvis and mass in the tubes. It is hard to come to this diagnosis because menstrual cycle is normal, and when patient presents with pain during menstruation, they will be prescribed pain killers.[8–10]

Early diagnosis is important to prevent future complications caused by conditions such as retrograde menstruation further leading to infertility. Diagnosis is usually confirmed by ultrasonography and MRI. MRI is the most sensitive investigation.[11,12]

Surgical treatment that is removal of the vaginal septum is the only treatment in obliterated half-vagina. Patients with this condition have a better chance of conceiving, but failure of pregnancy is high (74%) and prematurity as complication (22%). Most of them land up in C-section.

Clinical Significance

HWW syndrome has very low incidence with obstructive malformation of paramesonephric ducts. Diagnosis should be made as early as possible so surgical therapy can relive abdominal pain and save her from complications like endometriosis. Fertility issues should be looked into later.

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