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CASE REPORT

Obstructed Hemivagina and Ipsilateral Renal Anomaly (OHVIRA):

Case report

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Abstract

Background: Hemivaginal obstruction syndrome and ipsilateral renal anomalies are known as Herlyn-Werner-Wunderlich syndrome (HWWs) or better known by the acronym OHVIRA (Obstructed Hemivagina and Ipsilateral Renal Anomaly). It occurs as a result of complete failure of the Müllerian ducts to fuse and accounts for about 5% of Müllerian duct anomalies. Initial manifestations usually appear as a result of secretions accumulating within the hemivaginal obstruction. Patients with HWWs can come with complaints of lower abdominal pain, severe dysmenorrhea, pelvic or vaginal mass, abnormal vaginal discharge, acute urinary retention, fever or vomiting. Therefore, careful diagnosis and appropriate management of this condition is desirable. The objective of the study was to report on the management of the obstruction due to HWWs. **Case Report:** A 12-year-old patient was admitted to the gynecology ward of Dr. M. Djamil Central General Hospital from polyclinic with abdominal pain since 4 months before being admitted to the hospital. The pain gets worse every day before menstruation and decreases when given painkillers. Bleeding from the vagina often recurs with blackish color, a little smelly since the 1 month ago. **Conclusion:** OHVIRA syndrome is a triad characterized by hemivaginal obstruction and agenesis of the ipsilateral kidney (OHVIRA) and uterus in the delphys. Clinical symptoms in general are cyclic dysmenorrhea, palpable mass due to accumulation of menstrual flow, and severe pelvic pain which can later develop into a persistent form as a result of prolonged retention of menstrual secretions, obstructed hemivagina. Management options are surgical resection of the obstructed vaginal septum and drainage.

INTRODUCTION

Hemivaginal obstruction syndrome and ipsilateral renal anomalies are known as Herlyn-Werner-Wunderlich syndrome or better known by the acronym OHVIRA (Obstructed Hemivagina and Ipsilateral Renal Anomaly).^{1,2} It consists of a lateral fusion defect in the caudal portion of the Müllerian tract at 8 to 12 weeks of gestation and also includes abnormalities associated with failure of absorption from the septum at 20 weeks of gestation.²⁻⁵ According to the American Fertility Society classification, uterine didelphys is classified as a Class III anomaly, described as partial or complete uterus, cervical, and vaginal duplication.^{5,6} It occurs as a result of complete failure of the Müllerian ducts to fuse and accounts for about 5% of Müllerian duct anomalies.^{2,4,6,7} Independent formation of the hemiuterus and cervix of each of the Müllerian ducts without clear endometrial communication of the cavity, and at the cervical level, a small degree of fusion can be seen.^{1,3,6,7}

HWWs syndrome is a very rare inherited disorder reported in the literature. Initial



manifestations usually appear as a result of secretions accumulating within the hemivaginal obstruction.^{7,8} The etiology and pathogenesis of HWWs are unknown. The incidence of HWWs until 2011, only about 200 were documented in the world literature. It has a low prevalence and an undetermined rate.^{1,3,7,8} For HWWs the incidence of bilateral agenesis is 1/600 - 1/1200 and the prevalence of genital anomalies associated with kidney disorders in women is found to be 25-89%. While the prevalence of congenital uterine anomalies is approx. 6.7% of fertile men, 7.3% and 16.7% of infertile women with a history of repeated abortions.^{2,8,9} Unilateral renal agenesis occurs when one or both ureteral cysts fail to form, and the metanephric blastema does not differentiate into nephrons.^{7,8} This congenital anomaly is rare. Patients with HWWs can come with complaints of lower abdominal pain, severe dysmenorrhea, pelvic or vaginal mass, abnormal vaginal discharge, acute urinary retention, fever or vomiting.^{5,6,9,10} The aim of this article is to describe a case of OHVIRA.^{11,12}

CASE REPORT

A 12-year-old patient was admitted to the gynecology ward of Dr. M. Djamil Central General Hospital on July 27, 2020, sent from polyclinic. Abdominal pain since 4 months before being admitted to the hospital. The pain gets worse every day before menstruation and decreases when given painkillers. Bleeding from the vagina often recurs with blackish color, a little smelly since the 1 month ago. The patient had menarche in February 2021 and last menstrual period was July 25, 2021, with regular cycle every month which last for about 5 days each cycle with the amount of 2-3 times pad changes/day with menstrual pain has been felt since 4 months ago. Defecation and urination was within normal limits.

GYNECOLOGY REPORT

Inspection of V/U was normal. There was a yellowish exudate appears from vagina. From the rectal touché examination, the anus was normal, the anal sphincter tone was normal, the rectal mucosa was smooth, there was a palpable mass in the right upper quadrant size as big as the chicken egg, with solid consistency, no tenderness, and empty ampulla.

From the laboratory examination of the patient, they had hemoglobin 11.5 g/dl, leucocytes 6190/mm³, platelets 180000/mm³, hematocrit 32 %, basophil 0%, eosinophil 1%, segmented neutrophil 59%, monocyte 10%, lymphocytes 30%, prothrombin time 10.6s, activated partial thromboplastin time 25.9s, urea 43 mg/dl, creatinine 0.5 mg/dl, albumin 3.1 g/dl, globulin 3.6 g/dl, serum glutamic oxaloacetic transaminase 26 u/l, serum glutamic pyruvic transaminase 18 u/l, sodium 138 Mmol/L, potassium 4.5 Mmol/L, chloride 104 Mmol/L.

From the ultrasonography (Figure 1) there was Didelphys uterus, with endline, the size was 6.66 x 2.19 cm, and 5.20 x 3.56 cm. There was a well-defined hypoechoic mass appears in the cervical region. The size left and right adnexa within normal limits. The conclusion from ultrasonography suspected hematocolpos with uterus didelphys. From the BNO-IVP examination suggest agenesis Ren Dextra dd/ Non function ren dextra (Figure 2). The patient

underwent diagnostic laparoscopic surgery on July 2, 2021 (Figure 3) suggest hematocolpos with uterus didelphys.



Figure 1. Ultrasonography examination.



Figure 2. BNO-IVP examination

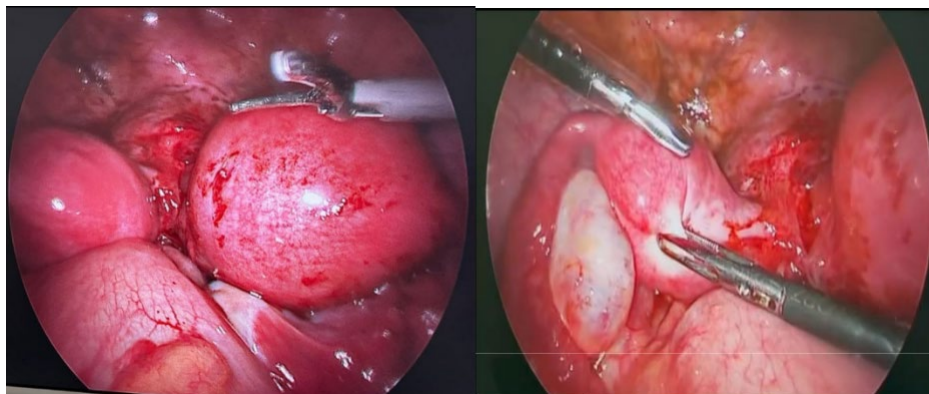


Figure 3. Diagnostic laparoscopic surgery



From anamnesis, physical examination, laboratory examination, ultrasonography examination and diagnostic laparoscopic, the patient was diagnosed with hematocolpos ec sindroma OHVIRA (HWWs). The operative management of the patient was hematocolpos evacuation. Post-operative management was catheter urine for 24 hours, vaginal tampon for 24 hours, vulva hygiene twice a day, fluid with IVFD ringer lactate 20 drops per minute, injection ceftriaxone 2x1 gr, metronidazole 3x500 mg orally, paracetamol 3x500 mg orally, sulfas ferosus 2x180 mg orally and vitamin C 2x50 mg orally.

DISCUSSION

A 12-year-old female patient was admitted from the patient who was consulted from the gynecological endocrinology polyclinic to the urogynecology department with a diagnosis of hematocolpos + uterine didelphys. Based on the history, physical examination, investigations, the patient was correct. A lot of blood coming out of the vagina (+), blackish color, a little smelly since the last 1 month. Menstrual history, menarche in February 2021, regular, 5 days, number of pads 2-3x/day, menstrual pain (+) since last 4 months.

The basis for the diagnosis in this patient was a syndrome, namely OHVIRA syndrome, a hemivaginal obstruction and ipsilateral renal agenesis (OHVIRA). Usually found in adolescence, immediately after menarche, with complaints of pain and a pelvic mass and rarely with infertility in adulthood. The association of renal agenesis with ipsilateral blind hemivagina was reported as HerlynWerner syndrome in 1971, and the association of renal aplasia, bicornuate uterus with isolated hematocervix, and a normal vagina was reported by Wunderlich in 1976.¹

The precise etiology and pathogenesis of OHVIRA syndrome is still unknown.^{5,6} It has been considered to represent the anomalous development of paramesonephric (Mullerian) and mesonephric (Wolffian) ducts.^{1,3,6,7,12-16} The Wolffian ducts, besides giving origin to the kidneys, are also inductor elements for adequate Mullerian duct fusion.^{1,3-6,12,16} Therefore, a developmental anomaly of the caudal portion of one of the Wolffian ducts may be the cause of unilateral renal agenesis associated with imperforate hemivagina.^{12,14,17}

On the side where the Wolffian duct is absent, the Mullerian duct is displaced laterally and cannot fuse with the contralateral duct, resulting in a didelphic uterus.^{15,17} The contralateral Mullerian duct gives rise to a vagina, whereas the displaced Mullerian duct that cannot come into contact with the urogenital sinus centrally forms a blind sac, leading to an imperforate or obstructed hemivagina. An obstructing longitudinal vaginal septum, which was also present in this case, is associated with 75% of these anomalies.^{12,13,17,18}

Renal agenesis is the most common associated nongenital anomaly and is always seen on the side with the obstructed hemivagina.^{18,19,20} Obstructed hemivagina and ipsilateral renal anomaly syndrome is usually discovered at puberty, shortly after menarche because of the cyclic, increasing lower abdominal pain secondary to hematocolpos resulting from longstanding, retained, partially clotted menstrual blood in the obstructed hemivagina.^{12,19} This syndrome may remain unrecognized at first because the menstrual flow from the patent unobstructed hemivagina gives the appearance of normal menses, and dysmenorrhea, if present, is a common complaint in this age group.^{17,19} Hematocolpos is suspected only months after menarche, and



the diagnosis is generally made only if this syndrome is suspected.²⁰

The most common clinical presentation is that of pelvic pain shortly after menarche in association with a vaginal or pelvic mass, and normal menstrual periods. Rarely, it presents as abnormal vaginal discharge, infertility, acute abdominal pain, vomiting, and fever. However, the syndrome in the present case presented with acute retention of urine, which has rarely been reported in published literature.^{13,14,20}

Early recognition and prompt surgical removal of the obstructing vaginal septum with drainage is the definitive treatment that will rapidly relieve the symptoms and prevent complications, including endometriosis.^{9,11,13} Ultrasonography and MRI are widely and effectively used in the diagnosis of genitourinary anomalies, with close to 100% accuracy being reported for MRI. Ultrasound is a noninvasive, widely available imaging modality that helps in the accurate diagnosis of this condition. However, the vaginal septum is difficult to visualize on ultrasound and is best shown on MRI. Magnetic resonance imaging is more sensitive in detecting the uterine contour, the shape of the intrauterine cavity, and the character of the septum compared to the other imaging modalities, but it is less adequate in diagnosing endometriosis, pelvic inflammation, and adhesions, so it has been suggested that the gold standard of diagnosis is laparoscopy.^{17,20}

The management is surgical resection of the obstructed septum and vaginal drainage, which allows relief of acute symptoms, treats obstruction to prevent endometriosis and preserves fertility.^{16,19-20}

CONCLUSION

OHVIRA syndrome is a triad characterized by hemivaginal obstruction and agenesis of the ipsilateral kidney (OHVIRA) and uterus in the delphys. Clinical symptoms in general are cyclic dysmenorrhea, palpable mass due to accumulation of menstrual flow, and severe pelvic pain which can later develop into a persistent form as a result of prolonged retention of menstrual secretions, obstructed hemivaginal. Treatment options are surgical resection of the obstructed vaginal septum and drainage.

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