Obstructed Hemivagina and Ipsilateral Renal Anomaly: 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 obstructed hemivagina and ipsilateral renal anomaly (OHVIRA). 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 are desirable. The objective of the study was to report on the management of the obstruction due to HWWs. Patient was suspected hematocolpos with uterus didelphys from ultrasound and suggested right renal agenesis from BNO IVP. Diagnosis then confirmed by laparoscopy diagnostic.

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.

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, fever, or vomiting [5], [6], [9], [10]. The aim of this article is to describe a case of OHVIRA [11], [12].

Introduction

Hemivaginal obstruction syndrome and ipsilateral renal anomalies are known as Herlyn-Werner-Wunderlich syndrome (HWWs) or better known by the acronym obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) [1], [2]. It consists of a lateral fusion defect in the caudal portion of the Müllerian tract at 8–12 weeks of gestation and also includes abnormalities associated with failure of absorption from the septum at 20 weeks of gestation [2], [3], [4], [5]. According to the American Fertility Society classification, uterine didelphys is classified as a class III anomaly and described as partial or complete uterus, vaginal and cervical 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 was 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%. The prevalence of uterine congenital anomalies is approximately 6.7% on fertile women and 7.3% on infertile woman. 16.7% women with this anomalies had history of recurrent pregnancy loss [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. Patient
had intermittent abdominal pain since 4 months before hospitalized. 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 were within normal limits.

**Gynecology record**

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, and there was a palpable mass in the right upper quadrant as big as the chicken egg, with solid consistency, no tenderness, and empty ampulla. Pyocolpos was found during the complication.

From the laboratory examination of the patient, they had hemoglobin 11.5 g/dl, leukocytes 6190/mm$^3$, platelets 180,000/mm$^3$, 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, and 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 form ultrasonography suspected hematocolpos with uterus didelphys. From the

**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, and investigations, the patient was correct. A lot of blood coming out of the vagina (+), blackish color, a little smelly since the past 1 month. Menstrual history, menarche in February 2021, regular, 5 days, number of pads 2–3x/day, menstrual pain (+) since past 4 months.

The basis for the diagnosis in this patient was a syndrome, namely, OHVIRA syndrome, a
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 magnetic resonance imaging (MRI) are widely and effectively used in the diagnosis of genitourinary anomalies, with close to 100% accuracy being reported for MRI. Ultrasound is a non-invasive, 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. MRI 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], [19].

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], [18], [19].

**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 and obstructed hemivaginal. Treatment options are surgical resection of the obstructed vaginal septum and drainage.

**References**

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