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Case Report

Uterus Didelphys with Obstructed Hemivagina and Ipsilateral Renal Agenesis (OHVIRA Syndrome): A Rare Case Report

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Abstract

The triad of uterine didelphys, obstructed hemivagina and ipsilateral renal anomaly known as OHVIRA syndrome, formerly known as Herlyn-Werner-Wunderlich syndrome, is a rare congenital urogenital malformation. It represents a diagnostic dilemma because of the regular menstruation and nonspecific abdominal pain. We present the case of a 13 year old pubertal girl presenting with severe dysmenorrhea. After evaluation with history and investigations she was diagnosed to have bicornuate uterus with hematometrocolpos, left sided hematosalpinx and left renal agenesis. After laparotomy she was diagnosed to have uterine didelphys with left sided hematometra, hematocolpos and ahematosalpinx due to left sided obstructed hemivagina and left renal agenesis (OHVIRA Syndrome). Surgery was done by abdominoperineal route. She had drainage of hematometra, hematocolpos and hematosalpinx and repair of obstructed hemivagina through perineal route. In conclusion, awareness of such anomaly is a prerequisite to early and prompt diagnosis. Surgery should not to be delayed after diagnosis of OHVIRA syndrome, so that complications can be avoided and reproductive function can be preserved.

Keywords: OHVIRA Syndrome, Uterine didelphys, obstructed hemivagina, renal agenesis, hematocolpos, hematometra, Mullerian malformation.

INTRODUCTION

OHVIRA Syndrome (Obstructed hemivagina with ipsilateral renal anomaly) is a rare congenital malformation consisting of an obstructed hemivagina, associated ipsilateral renal anomalies and various types of uterine malformations [1]. It was first reported by Purslow in 1922 and was formerly known as Herlyn-Werner-Wunderlich Syndrome when renal agenesis and ipsilateral blind hemivagina was described in 1971 by Herlyn and Werner, followed by Wunderlich's description of associated uterine malformation in 1975[2]. It amounts to 0.16%-10% of all cases of Müllerian malformations [3]. Prevalence varies between 0.1%-3.8% [4]. OHVIRA syndrome comprises a variety of clinical presentations including an obstructing hemi-vagina combined with various Mullerian and renal anomalies such as a uterus didelphys, unilateral obstructed hemi-vagina, ipsilateral renal agenesis, dysplastic kidney, pelvic kidney or ectopic ureter [5]. Rock and Jones [6] classified it into three separate categories (Figure1). Type I includes blind hemi-vaginal septum without an opening. In this case the uterine horn behind the septum has no connection to the outside or into the contralateral uterus. Menstrual blood accumulates in the cavity behind the vaginal septum. Type II is blind hemi-vaginal septum with an opening. In this case there is a pinpoint-size hole in the septum through which a limited amount of menstrual blood drains out. The ipsilateral uterus is separate from the other horn. Type III comprises complete hemi-vaginal septum with

cervical fistula. In this case a fistula connects the two cervices, the one on the side of the obstructed vagina and the contralateral cervix.

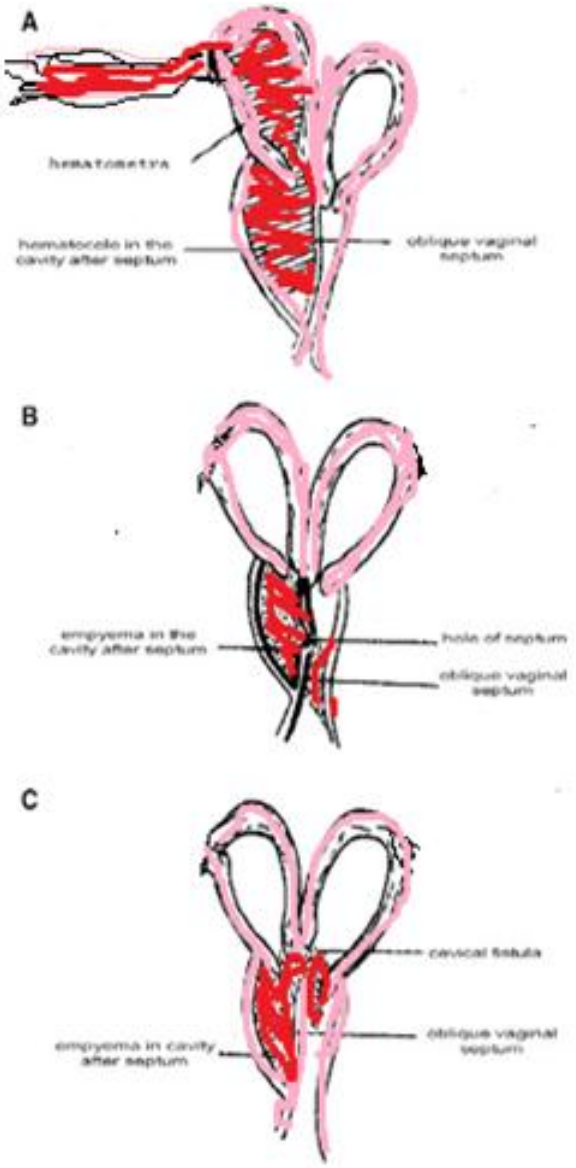

Fig-1: Classification of OHVIRA Syndrome: a. Type I, b. Type II, c. Type III

Patients usually present after menarche with progressive dysmenorrhea, lower abdominal pain, and a vaginal or pelvic mass and normal menstrual periods. The didelphys uterus in these cases is associated with reproductive issues such as miscarriages, preterm labor and placental dysfunction. Rare presentation may include foul mucopurulent discharge, intermenstrual bleeding, acute retention of urine, fever, vomiting and abdominal swelling [7-9]. Accurate identification is important as a delay or lack of treatment may increase the risk of endometriosis, pelvic adhesions and infertility [10]. The diagnosis of OHVIRA syndrome requires a multi-modal approach, integrating the patient's history, clinical presentation, imaging studies including ultrasonography (USG), computed tomography (CT) and magnetic resonance imaging (MRI) and direct visualization via hysteroscopy or laparoscopy [11].

Once the diagnosis of OHVIRA syndrome is made, surgical management should be carried out as soon as possible in order to relieve symptoms and to prevent long term complications related to retrograde menstrual flow like hematocolpos, pyocolpos, endometriosis and pelvic adhesions. The obstructing vaginal septum is usually oblique and varies in thickness from very thin to quite thick. Resection of the vaginal septum is the treatment of choice [12]. In some situations where surgical correction is not readily available or contraindicated for other reasons, menstrual suppression with hormonal medications is a reasonable alternative. However, only surgery provides definitive management.

CASE PRESENTATION

A 13 year old pubertal girl of middle class family was admitted in Reproductive Endocrinology and Infertility Department of Bangabandhu Sheikh Mujib Medical University with the complaints of severe dysmenorrhea for 4 months and a history of laparotomy 3 months back in a local hospital. According to the statement of the patient she had menarche at 10 years of age. She was regularly menstruating since her menarche and had no dysmenorrhea in first two cycles. Then she developed left sided lower abdominal pain during menstruation which was dull aching in nature, continuous, severe in intensity with no radiation, no specific aggravating or relieving factor not associated with vomiting or dysuria, started from one day before menstruation and continued throughout the menstrual period. Gradually the pain was increasing in intensity and duration for 2½ years. For last 4 months the pain was severe, intense, not subsided with analgesics and continued beyond menstruation. With these complaints she went to a gynecologist and sonologically diagnosed as a case of left sided endometrioma with hydrosalpinx. Explorative laparotomy was done 3 months back in a local hospital but no definitive diagnosis was made. Her dysmenorrhea did not subside. She was treated with dienogest and combined oral contraceptive pill for menstrual suppression for last 2 months and referred to Bangabandhu Sheikh Mujib Medical University with an ultrasonogram reporting bicornuate uterus with a single vagina. Right horn was normal in size and the left horn including cervical region appeared distended with echogenic collection suggesting hematometrocolpos and hemosalphinx (Figure 2). Left kidney was not located in left renal fossa or in the abdomino- pelvic cavity.

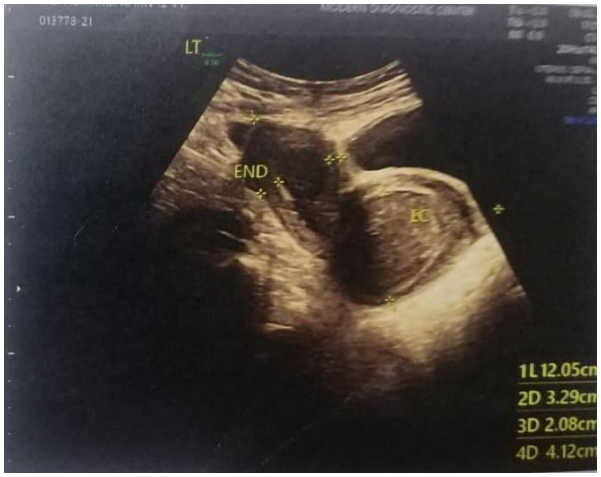

Fig-2: Hematometrocolpos and Hemosalphinx

With due consent, the patient was examined after admission. On per abdominal examination, there was mild tenderness on left iliac fossa. No palpable mass was found. On per rectal examination anterior rectal wall was bulged on left side.

So, from history, examination and USG report our provisional diagnosis was bicornuate uterus, left horn associated with transverse vaginal septum. MRI abdomen reported agenesis of left kidney and bicornuate uterus with hematometra at left horn, left adnexal cystic lesion suggestive of hydrosalphinx (Figure 3).

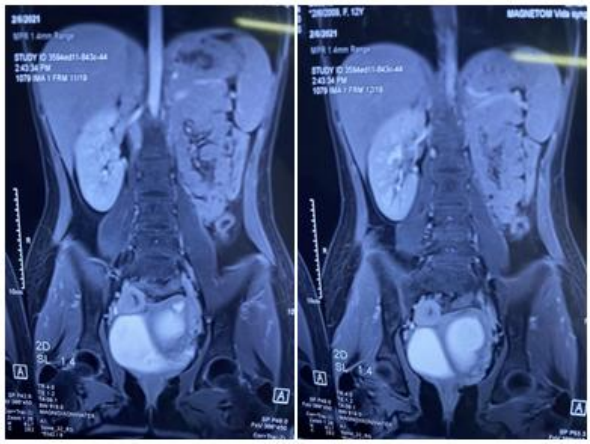

Fig-3: MRI suggestive of OHVIRA syndrome

With full consent patient underwent surgical management. During operation she was diagnosed as a case of uterus didelphys with left sided hematometra, hematocolpos, hematosalpinx and left sided obstructed hemivagina, left renal agenesis (OHVIRA Syndrome) and operation was done by abdomino- perineal route. Under general anesthesia laparotomy was done followed by evacuation of hematometra, hematocolpos and hematosalpinx. Repair of obstructed hemivagina was done in perineal route.

Examination under anesthesia revealed that right sided cervix was small and left sided obstructed hemivagina was high up and could not be reached properly. No altered blood was retrieved when explored through syringing. Decision for laparotomy was taken. Two separate uterus was found with one fallopian tube and one ovary on each side (Figure 4). Left sided hematometra, hematosalpinx, hematocolpos were identified. After separating peritoneum from bladder an incision was given over the upper segment of left obstructed hemivagina and dark thick chocolate colored material was evacuated. Then vaginal wall was repaired (Figure 5) and abdomen was closed in layers. Right sided uterus, ovary and fallopian tube were found healthy and separated from the left sided uterus by an omental band (Figure 6). Next through perineal route lower part of obstructed vaginal septum was incised and marsupialization was done (Figure 7). A Foley's catheter was kept inside the left vagina for 7 days. A vaginal pack was given for 1 day.

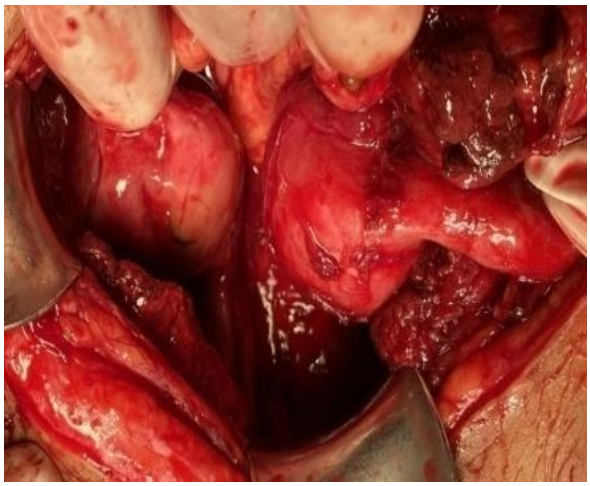

Fig-4: Uterus didelphys

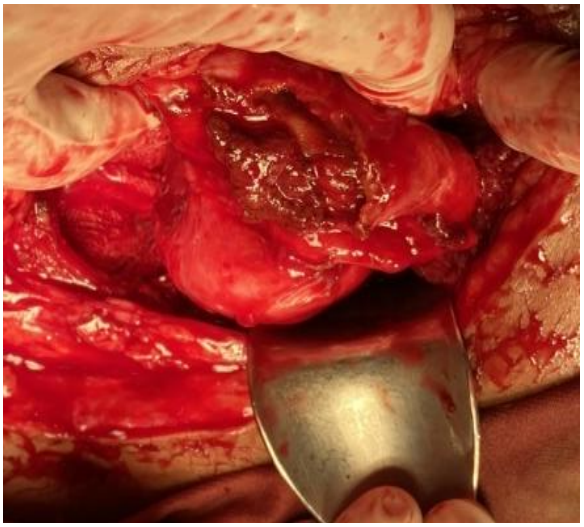

Fig-5: Hematocolpos drained

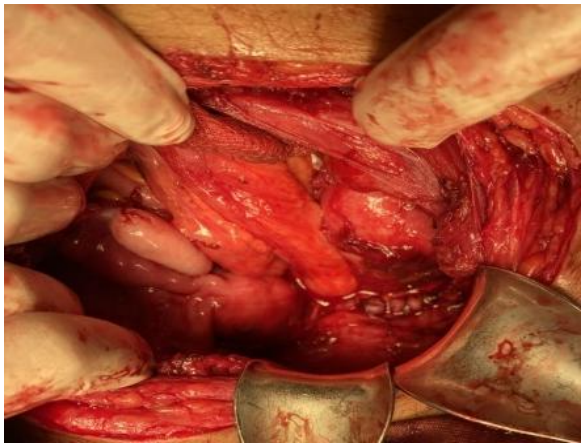

Fig-6: Omental band between two uterine horns

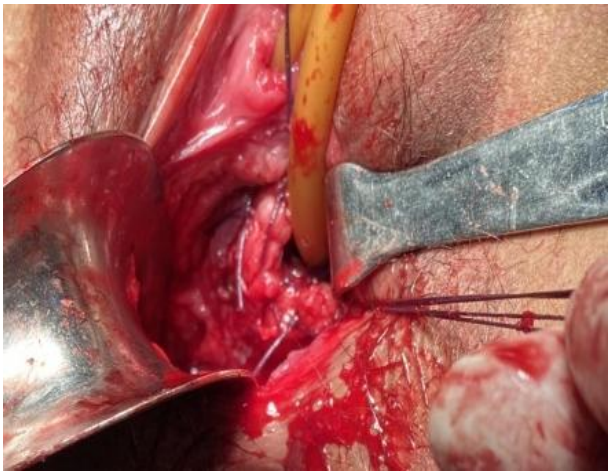

Fig-7: Marsupialization of transverse vaginal septum

The post-operative period was uneventful. Withdrawal bleeding occurred from first day of operation through normal right sided vagina and reconstructed left sided vagina. Patient was discharged on her 7th post-operative day. She was on combined oral contraceptive pill for 1 month and the withdrawal bleeding was pain free in next cycle.

DISCUSSION

The exact etiology and pathogenesis of OHVIRA syndrome is still unknown. It has been considered to represent anomalous Müllerian as well as Wolffian duct development [13]. 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 [14, 15]. On the side where the Wolffian duct is absent, the Müllerian duct is displaced laterally so it cannot fuse with the contralateral duct, resulting in a didelphic uterus, and cannot come into contact with the urogenital sinus centrally. The contralateral Müllerian duct gives rise to a vagina, while the displaced component forms a blind sac with an imperforate or obstructed hemivagina [16]. The vaginal introitus is not involved because lower ⅔rd of the vagina is originated from the urogenital sinus.

The most common clinical presentation is pelvic pain initiating shortly after menarche, associated with a vaginal or pelvic mass and normal menstrual periods. Recurrent and progressive pelvic pain is the chief complaint in most patients. Hematocolpos resulting from retained long-standing, partially clotted menstrual blood in the obstructed hemivagina is often clinically detected as a pelvic mass. Most of the patients suffering from this syndrome are diagnosed late due to its rarity and the nonspecific clinical presentation. Moreover the menstrual flow that comes from the unobstructed hemivagina gives the impression of normal menses. Consequently accurate diagnosis and surgical treatment may be delayed for several months or even years. Hematocolpos is suspected only months after menarche and the diagnosis is generally made only if the suspicion of the existence of this syndrome is raised [17]. A right-sided prevalence of the obstructed system has been described [13, 17]. Hematocolpos, hematometra and hematosalpinx may occur, as well as hemoperitoneum as a consequence of blood stasis and retrograde menstruation in the obstructed system. Endometriosis can result from blood reflux into the abdominal cavity and may have grave consequences [15, 18].

The evidence of hematocolpos on ultrasound, appearing as a fluid collection with low-level echoes, can make the detection of the uterine anomaly (didelphic/bicornuate bicollis uterus) easier [15]. Magnetic resonance imaging (MRI) with multiplanar image acquisition provides more detailed information. The accuracy of MRI for diagnosing uterine malformations is well-established [19].

Early recognition prompts surgical removal of the obstructing vaginal septum, with rapid relief of symptoms and prevention of complications related to chronic crypto menorrhea, such as endometriosis, pelvic adhesions and infectious collections like pyocolpos, pyometra and pyosalpinx [15, 17]. Surgical excision of the septum and drainage of the obstructed vagina is the definitive treatment [16, 17]. Resolution of the obstruction is also considered mandatory in order to prevent secondary endometriosis [20]. Moreover, fertility can be preserved as it is not significantly decreased in women with didelphic uterus.

CONCLUSION

OHVIRA Syndrome is a rare anomaly with potential short and long term complications. The diagnosis is likely to be missed due to normal menstruation and nonspecific abdominal pain. In girls with unilateral renal anomaly it is particularly important to consider OHVIRA Syndrome. Reporting such cases increases awareness of the syndrome and helps to

achieve early diagnosis and avoid potential complications.

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