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(RESEARCH ARTICLE)

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Hysteroscopic versus laparoscopic management of two different variants of OHVIRA syndrome with preservation of hymen integrity

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Abstract

OHVIRA syndrome is a rare mesonephric duct-induced mullerian anomaly characterised by the triad of uterus didelphys, obstructed hemivagina and ipsilateral renal anomaly.

I present here 2 cases of OHVIRA syndrome. One with the mentioned classic features and another rare variant with a high/proximal vaginal septum which made the diagnosis and management even more challenging.

The first case was a 12-year-old girl who presented with dysmenorrhea in the last 2 months since menarche. Ultrasound and MRI scans confirmed the diagnosis of right haematocolpos, double uterus and right renal agenesis. Trans-hymenal vaginoscopic septoplasty was performed using a hook and loop of a bipolar resectoscope.

The second case was a 14-year-old girl who was referred to us with left iliac fossa pain, absent left kidney and a pelvic cystic structure. After full assessment and investigations including MRI, the differential diagnosis was unicornuate uterus with functioning non-communicating horn versus OHVIRA syndrome with cervicovaginal atresia (class 1.2). However, the final diagnosis of OHVIRA syndrome (class 1.1) with a high/proximal vaginal septum was confirmed by combined hysteroscopy and laparoscopy. She was managed by laparoscopic ipsilateral hemihysterectomy and septoplasty. The integrity of the hymen was preserved as per the patient and her parents' wishes.

OHVIRA syndrome is generally a very rare condition. However, there are even more rare forms and variants of this anomaly which need more vigilance to reach timely accurate diagnosis and more experience and skills to perform proper management.

Keywords: OHVIRA syndrome; Herlyn-Werner-Wunderlich syndrome; Uterus didelphys; Obstructed hemivagina; Renal agenesis; Vaginal septoplasty

1. Introduction

OHVIRA syndrome is a rare complex congenital anomaly of the genitourinary tract characterised by the triad of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis. These findings were first described by Purslow in 1922 [1]. However, an association between renal agenesis and ipsilateral blind hemivagina was reported by Herlyn and Werner in 1971 [2]. The condition was then named Herlyn-Werner-Wunderlich (HWW) syndrome in 1976 when Wunderlich reported an association between the previously mentioned two components with double or Bicornuate uterus [3]. In 2007, Smith and Laufer suggested the name of OHVIRA syndrome which is more descriptive of the condition [4].

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In literature, OHVIRA syndrome is found as isolated case reports or a few case series. However, the estimated incidence is about 1 in 20,000 [5]. Although the exact aetiology is unknown, multiple factors such as genetic, environmental and endocrine factors may affect the normal development of mullerian and mesonephric ducts during embryological life [6].

The classic mullerian theory in 1830 stated that fallopian tubes, uterus, cervix and upper 1/3 of vagina are derived from mullerian ducts, while lower 2/3 of vagina and vestibule are derived from the urogenital sinus [7,8]. This theory failed to completely explain the development of OHVIRA syndrome in two ways; first is the oblique or longitudinal vaginal septum which suggests that vagina is supposed to develop from a paired structure, unlike the urogenital sinus which is an unpaired structure. Second is the associated ipsilateral renal anomaly in 100% of OHVIRA cases. This assumes that kidney and vagina may arise from a common origin. Hence, Acien theory was postulated in 1992 which stated that fallopian tubes, uterus and cervix are derived from mullerian ducts. Vaginal plate is derived from the mesonephric ducts (paired structure explaining the oblique vaginal septum and associated renal anomaly) and vaginal mucosa is derived from mullerian tubercle. Urogenital sinus is responsible only for the development of vaginal vestibule [9].

OHVIRA syndrome is classified as "class III" mullerian anomaly according to American Fertility Society (AFS) classification in 1988 [10] and as "U3b C2 V2" according to European Society of Human Reproduction and Embryology (ESHRE) classification in 2013 [11]. According to the American Society for Reproductive Medicine (ASRM) classification in 2021, it was named "uterus didelphys and obstructed hemivagina" [12]. Although the classic description of OHVIRA syndrome is with uterus didelphys, this anomaly occurs also with Bicornuate and septate uterus [13].

OHVIRA is then sub-classified into 4 classes; class 1.1 with blind hemivagina, class 1.2 with cervicovaginal atresia, class 2.1 with vaginal septal perforation (incomplete vaginal septum) and class 2.2 with communicating uteri [14]. Another rare variant of this anomaly is a high or proximal vaginal septum causing more difficulty in diagnosis and management [15] (Fig. 1).

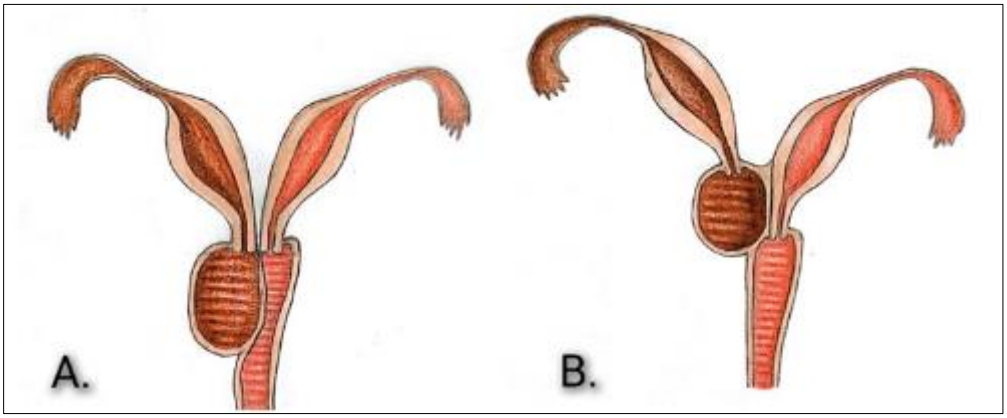

Figure 1 A. Diagram demonstrating the classic description of OHVIRA syndrome. B. Diagram demonstrating a case of high/proximal vaginal septum

Associated ipsilateral renal anomalies include renal agenesis (in 65% of cases), dysplastic kidney, atrophic kidney, pelvic kidney, ectopic ureter and ureterocele. Contralateral renal anomalies are also recorded and they include vesicoureteral reflux and hydroureteronephrosis [4]. Endometriosis is found in around 50% of cases of obstructive mullerian anomalies [16].

I report in this article 2 cases of different variants of OHVIRA syndrome. The first was the classic presentation of the syndrome and its management with the least invasive corrective surgery. The second case presented with a high/proximal vaginal septum showing the relative difficulty in diagnosis and management.

2. Case 1

A 12-year-old girl presented with dysmenorrhea in the last 2 months since menarche. Transabdominal ultrasound scan (USS) revealed a pelvic cystic structure (? haematocolpos). Magnetic resonance imaging (MRI) showed suspected right haematocolpos, double uterus and right renal agenesis (Fig. 2).

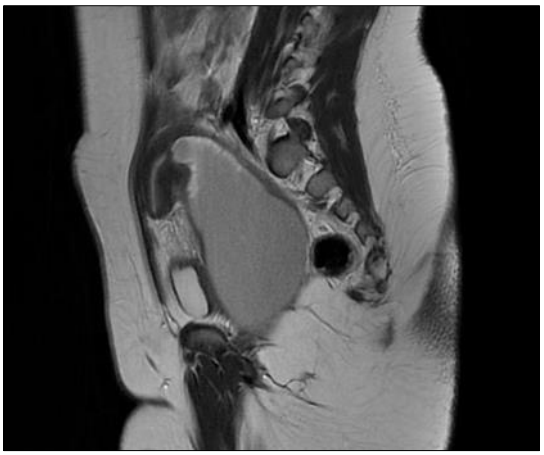

Figure 2 MRI showing the haematocolpos of the obstructed right hemivagina and the ipsilateral hemiuterus on the top

Due to cultural reasons, the patient and her parents requested preservation of hymen integrity. Vaginoscopic septoplasty was performed using a bipolar resectoscope. The resectoscope was introduced into the patent left hemivagina through the intact hymen where the bulging obstructive vaginal septum could be visualised on the right side. Performing the procedure immediately postmenstrual facilitated the identification of the maximally distended septum before the accumulated blood started to be absorbed. Holding labia minora together maintained vaginal distention allowing good visualisation for septal resection all through the procedure.

Procedure started with visualisation of the contralateral cervix (on the left side in this case) as a landmark, then identification of the distended vaginal septum (on the patient's right side) (Fig. 3-A). The hook of a bipolar resectoscope was used to incise the septum at the most bulging site until drainage of retained pus. The pus was an unexpected finding, as there was no suggestive signs during the preoperative clinical assessment. Then, the resectoscope was passed through the created opening to visualise the ipsilateral cervix on the right side. Consequently, the septal opening was extended in cephalad then caudal directions (Fig. 3-B). The remaining edges of the septum were then resected by the loop to reduce the risk of adhesions and reobstruction (Fig. 3-C).

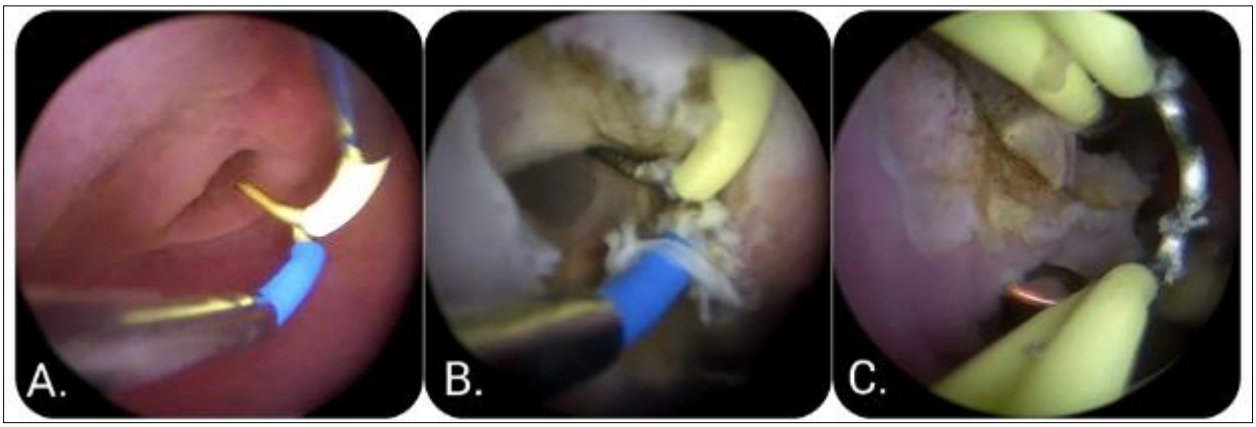

Figure 3 A. Vaginoscopic visualisation of the left cervix and bulging vaginal septum on the right side. B. Incision of the obstructive vaginal septum using the resectoscope hook. C. Resection of septal edges using the loop of the resectoscope

3. Case 2

A 14-year-old girl had been complaining of left iliac fossa pain for the last 1 year, presented to a urologist who requested a CT scan showing absent left kidney and pelvic cystic structures. For the later, she was referred to me as the gynaecologist. After clinical assessment, USS and MRI, 2 cystic structures on the left side of the pelvis could be visualised; one thin-walled rounded cyst and another smaller but thick-walled and elongated cyst in front of the first one. The radiologist claimed the first cystic structure to be haematometra based on the higher location in the pelvis, and the

second one to be haematosalpinx based on its oblong shape. I was inclined to believe that the first cystic structure was a haematocolpos based on its thin wall and the second one was the haematometra with the thicker uterine wall. Consequently, the preoperative differential diagnosis was one of 3 possibilities; unicornuate uterus with functioning non-communicating horn, OHVIRA syndrome with cervicovaginal atresia (class 1.2) or OHVIRA syndrome (class 1.1) with a high/proximal vaginal septum (Fig. 4).

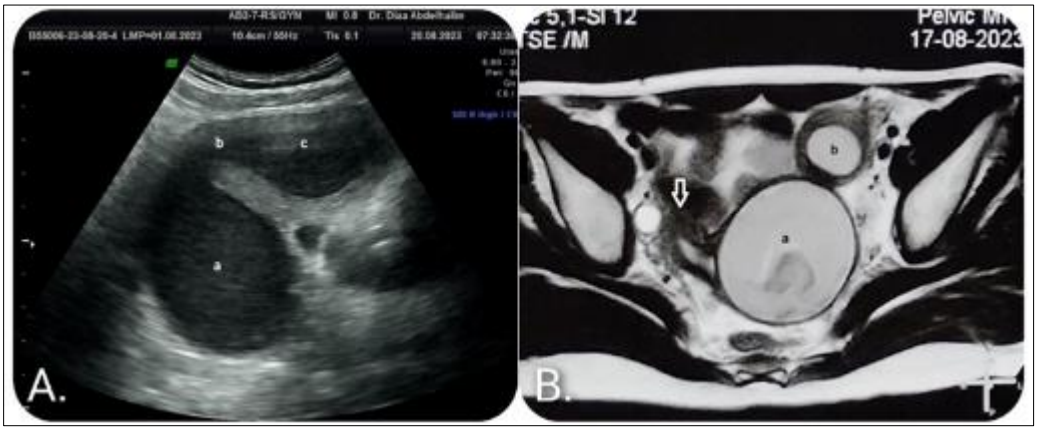

Figure 4 A. Ultrasound image showing haematocolpos (a) haematotrachelos (b) haematometra (c). B. MRI showing the right hemiuterus (white arrow), left haematocolpos (a) and left haematometra (b)

The final diagnosis could only be made after performing combined hysteroscopy and laparoscopy. The trans-hymenal hysteroscopy revealed completely normal vaginal canal with no evidence of a bulging vaginal septum and a normal-looking cervix leading to a narrow tubular cavity to the right side showing a single tubal ostium. Laparoscopy showed a hemiuterus on the right side (the one entered by hysteroscopy communicating with the vagina), a large swelling filling the pelvic cavity (proved to be the distended left hemivagina "left haematocolpos") and the distended left hemiuterus (left haematometra). What caused confusion and made the diagnosis more difficult is the higher level of the haematocolpos protruding cephalad secondary to a very proximal obstructive vaginal septum (Fig. 5-A).

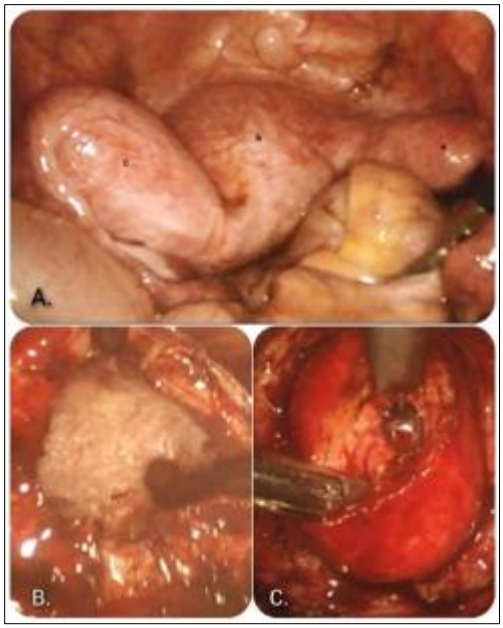

Figure 5 Laparoscopy showing A. the right hemiuterus (a), left haematocolpos filling the pelvic cavity (b) and left haematometra pushed cephalad by the large haematocolpos (c). B. the cavity of the obstructed hemivagina after suction of chocolate material, C. incision made to the high/proximal vaginal septum and tip of the sound

Laparoscopic left hemihysterectomy and opportunistic left salpingectomy was performed. The colpotomy done during the hemihysterectomy opened the obstructed hemivagina and the viscid blood and chocolate material were suctioned

(Fig. 5-B). Then, by the guidance of a sound introduced through the natural hymenal opening into the vagina, the obstructing vaginal septum was identified and incised by laparoscopy scissors (Fig. 5-C). To reduce the risk of re-closure of the incised vaginal septum, marsupialization of its edges was conducted. Finally, the vaginal vault of the colpotomy was closed by Intracorporeal suturing. A second-look hysteroscopy was performed on the same setting to show the final view of the septoplasty and to confirm the continuity between the main vaginal canal and the previously obstructed hemivagina.

4. Discussion

Diagnosis of OHVIRA syndrome is usually difficult and delayed due to rarity of the condition which is not thought of by the clinicians. Eyes do not see what mind does not know. Menstrual flow from the non-obstructed hemiuterus and vagina deviates the attention from the obstructed side and further delays the diagnosis. Besides, dysmenorrhea - being the main symptom of OHVIRA syndrome – is a common complaint of girls at this age and is usually masked by simple analgesics.

In addition to the need for awareness of the condition and experience among clinicians, correct and timely diagnosis can be reached by focused history taking, good clinical examination and imaging modalities namely USS being the first-line and MRI being the gold-standard investigation [17,18].

The management is mainly corrective surgery in the form of vaginal septal resection for the objective of relieving symptoms, mainly in the form of dysmenorrhea, and preventing further complications such as pelvic inflammatory disease (PID) and endometriosis [19]. The surgeon should put into consideration preservation of future fertility and hymen integrity in virgins, should patients' cultural views make this necessary. As a result, trans-hymenal hysteroscopic septoplasty can be considered the best approach to perform this corrective surgery with the least minimally invasive intervention and the best outcome [20]. If hymenal integrity is not an issue, then the conventional vaginal approach could be considered. In some cases, when vaginoscopic or vaginal approaches alone do not allow adequate and safe resection of the septum, then laparoscopy or laparotomy may be required [19]. This was the situation in the second case I reported in this article with the high/proximal septum when laparoscopy was necessary to safely and adequately perform the colpotomy and septoplasty. Some may argue that hemihysterectomy was not mandatory and septoplasty alone would have been enough. In my opinion, laparoscopic colpotomy and septoplasty in the presence of the hemiuterus would have been technically difficult and challenging. Besides, leaving the hemiuterus behind with the monthly menstrual flow would increase the risk of re-accumulation of blood in case of stenosis or reobstruction of the septum, especially in case of proximal septum with narrow surface area. Another debate is that hemihysterectomy per se would have been enough without the need for the septoplasty. The answer for this is that leaving closed space of the obstructed hemivagina would result in mucocolpos or hydrocolpos due to accumulation of discharge from cervical or vaginal glands. Colpectomy of the obstructed vagina was described, however, it needs extensive dissection with more blood loss and higher complication rate. Other management options which may be necessary include the management of associated endometriosis by the specialised gynaecologist, in addition to nephrectomy of a multicystic dysplastic kidney (MCDK) and excision of an ectopic ureter by the urologist [21].

Post-operative follow-up of these cases is very important due to the incidence of reobstruction as a result of adhesions or stenosis, persistent mucous vaginal discharge and the possibility of ascending infection and PID. Follow-up of the associated endometriosis and possible infertility may also be required [4]. In addition, there is a higher risk of developing clear cell carcinoma of the vagina and adenocarcinoma of the cervix in these patients. This is why routine vaginal examination and cervical screening for each of the two cervices is of clinical importance [22]. From nephrology perspective, patients with solitary kidney need better care and timely proper management of urinary tract infections to avoid renal failure and potential need for dialysis [21].

5. Conclusion

Diagnosis of OHVIRA syndrome requires clinical awareness among gynaecologists, urologists and paediatric surgeons. Ultrasound and MRI scans are very helpful diagnostic tools. However, in some rare variants such as those with high/proximal obstructing vaginal septum, diagnosis may only be confirmed during laparoscopy.

In the era of endoscopy, the standard management of most cases of OHVIRA syndrome should be by vaginoscopic septal resection preferably using a bipolar resectoscope. This can be performed trans-hymenal to preserve the integrity of the hymen.

If this is not applicable, such as in cases with high/proximal vaginal septum, I recommend laparoscopic hemihysterectomy on the ipsilateral side, then colpotomy and septotomy guided by a sound passed through the hymenal orifice, followed by marsupialisation of edges of the septum to reduce risk of reobstruction.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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