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CASE REPORT

HYSTEROSCOPIC RESECTION OF VAGINAL SEPTUM UNDER ULTRASOUND GUIDANCE IN OHVIRA SYNDROME WITH PRESERVATION OF HYMEN

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A B S T R A C T

Background/Aims: To report a patient who had hysteroscopic vaginal septum removal in Obstructed Hemivagina with Ipsilateral Renal Anomaly (OHVIRA) syndrome, with preservation of the hymen under ultrasound guidance.

Methods: In a tertiary academic referral center, hysteroscopic vaginal septum resection was performed for a 12 year-old girl with uterus didelphys, obstructed hemivagina, and renal agenesis along with hypothyroidism.
Hysteroscopic resection of a vaginal septum in a single adolescent female, who had uterus didelphys and obstructed hemivagina that was preformed twice.

Results: Hematometrocolpos drainage after hysteroscopic resection of vaginal septum under ultrasound guidance, along with preservation of the hymen, is a more conservative alternative to conventional vaginal septum excision. This resulted in symptomatic relief of the cyclic dysmenorrhea .after 4 months follow up. However, repeat surgery was necessary after 7 months due to closure of the vaginal septum incision.

Conclusion: Hysteroscopy is a safe alternative in resecting a vaginal septum in OHVIRA syndrome. However, repeat surgery might be indicated in few cases when the vaginal septum is high and thick. Longer follow up is warranted in such cases due to the risk of closure of the vaginal aperture along with the unintended consequences of hematometrocolpos and abdominal pain.

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INTRODUCTION

Herlyn-Werner-Wunderlich (HWW) syndrome is a rare congenital anomaly of the Müllerian and Wolffian ducts [1]. HWW, is also known as Obstructed Hemivagina with Ipsilateral Renal Anomaly (OHVIRA). OHVIRA syndrome is characterized by, the triad of uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis [2]. This rare syndrome, was first described by Purslow in 1922 [3]. Another German report, by Herlyn and Werner on the Simultaneous occurrence of an open Gartner-duct cyst, a homolateral aplasia of the kidney and a double uterus, has later on ensued [4]. In 1976, Wunderlich, further reported different management options in patients with bicornuate uterus who had simple vagina and isolated hematocervix on right without connection of the right uterus to the vagina [5]. These patients had aplasia of the right kidney and ureter [5]. Renal agenesis is the most common urologic anomaly in OHVIRA syndrome [6]. Other malformations are renal duplication and multicystic dysplastic kidney [6].

The commonest presentation for patients with OHVIRA syndrome is periodic pelvic pains or dysmenorrhea due to the effect of the progressive distention of the obstructed hemivagina[7]. The mean age of presentation is about 14 to 15 years [8].

Both ultrasonography and MRI can aid in the diagnosis of OHVIRA [9]. Excision of the vaginal septum and drainage of hematometrocolpos is the standard treatment option to relieve the pain [8]. A Korean group first described Hysteroscopic resection of the vaginal septum, for OHVIRA syndrome in 1998 [10]. This was followed by few other studies reporting successful excision of the vaginal septum via the hysteroscope [11, 12]. In the Middle East, preservation of the hymen is important for patients and their families due to the social, religious and cultural beliefs.

We, hereof, report a case of hysteroscopic resection of a vaginal septum in a single adolescent female with hypothyroidism, who had uterus didelphys and obstructed hemivagina.The procedure was performed under ultrasound guidance without laparoscopy along with the preservation of the hymen integrity. The procedure was repeated due the

closure of the initial incision. Previous case reports have described follow up of patients up to 12 months [10]. Few other reports have followed patients between 4 and 6 months [13]. We believe follow up periods should be more frequent and over one year.IRB approval was obtained.

Case report

A 12 year-old single girl presented to the emergency department with one-week history of dysmenorrhea and abdominal pain mainly in the right iliac fossa. She was on her fifth day of her menses. Her medical history revealed hypothyroidism, which she was treated for, and her surgical history was unremarkable. She had her menarche five months earlier, which was regular initially and started becoming irregular in the past two months. She had already received treatment from several gynecologists to relieve her abdominal pain. Upon examination, her abdomen was tender particularly on the right lower quadrant. She had normal vulva and urethra and there was no obvious vaginal bleeding. The laboratory tests revealed a normal white blood cell count and hemoglobin of 13.3g/dl. An abdominal ultrasonography concurred with the pelvic MRI findings that confirmed the absence of the right kidney and showed two separate uteri with two separate cervices and two proximal vaginas.

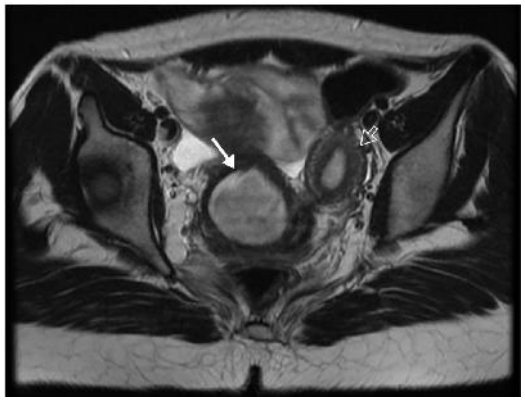

Figure 1 This Sagital T2-weighted MRI fast spin-echo image shows blood filled right hemiuterus-hematometra (long arrow) and upper 1/2 vagina- hematocolpos (short arrow) and normal lower 1/2 of vagina (arrowhead)

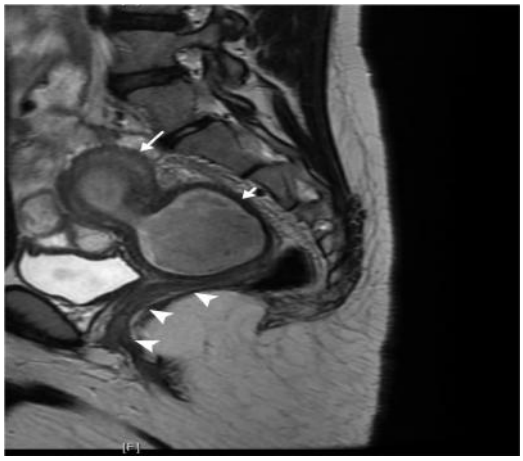

Figure 2 This Short Axis T2-weighted MRI fast spin-echo image shows two separate uterine bodies, right is distended with blood-hematometra (long arrow) and left normal uterus (open arrow).

The right vagina was distended and filled by a large hypoechoic mass slightly hypointense material on T2-weighted images (figure 1 and 2) and hyperintense material on T1-weighted image sequences suggesting a blood collection, haematocolpos measuring 5.6 x 4 cm on the right side. Only the distal portion of the left vagina was identified suggestive of a longitudinal septum, which was over one centimeter in thickness. The bladder and both ovaries were normal. There was no pelvic collection.

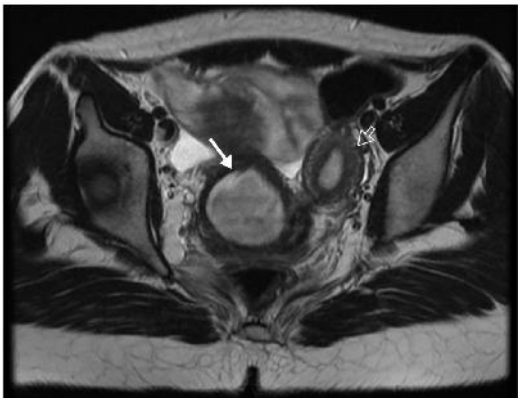

Figure 3 This image shows the Hysteroscopic resection of the vaginal septum and drainage of hematometrocolpos (short arrow) under Transabdominal ultrasound by 9 mm bipolar hysteroscope (arrowheads) The right hemiuterus-hematometra (long arrow)

The patient was diagnosed to have Obstructed Hemivagina with Ipsilateral Renal Anomaly (OHVIRA) syndrome. Hysteroscopic resection of the vaginal septum, drainage of hematometrocolpos under ultrasound guidance was performed along with the preservation of the hymen. Under general anesthesia, a 9 mm bipolar hysteroscopy (Gynecare, Ethicon, U.S.A.) was introduced into the vagina without a speculum to preserve the integrity of the hymen; saline was used as a distending medium. A 16F Foley's catheter was inserted and the bladder was filled with saline for better visualization by ultrasound. Hysteroscopic view identified the bulging mass on the right side and as well as the uterine cervix on the left side (figure 3). Under abdominal ultrasound, the obstructed vaginal septum and the hysteroscope tip were identified. The vaginal septum was resected to about 3 cm length by the hysteroscopic loop using a cutting electrode. Dark chocolate colored material was drained through the septum and the size of the hematometrocolpos was markedly reduced as has been identified by the ultrasound. The right and left cervices were delineated by the hysteroscope. Post operatively, a 12F Foley's catheter was inserted and was kept for one day to keep the outflow tract patent. After 4 months the patient had was asymptomatic and had regular periods. Unfortunately, the patient had presented to the emergency department after 7 months complaining of acute abdominal pain mainly in the right iliac fossa, repeat ultrasound and MRI had revealed re-accumulation of hematometrocoplos on the right side. A second hysteroscopic resection of the vaginal septum was performed where a larger incision along the old sealed incision was done. The incision in the second time was increased to 4 cm and the chocolate material was completely drained. Thorough rinsing of the vagina was performed by saline via the hysteroscopy. A 16 F Foley's catheter was kept intravaginally for one day. Patient was asymptomatic after follow up and frequent monthly visits were arranged for the patient in order to ensure her wellbeing.

OHVIRA syndrome is the result of disruption in the continuum of embryological development of the caudal

portion of one Wolffian duct with the subsequent involvement of the ipsilateralMüllerian duct [14]. On the same affected side, there is renal agenesis, which is the result of failure of regular ureteric budding [14]. Different types of double uterus and cervico-vaginal obstructions occur because of failure of the Müllerian duct to fuse with both its opposite counterpart and with the urogenital sinus [14].

OHVIRA syndrome is rare; the reported incidence has between the range of 0.1% and 3.8% [15]. The patients usually present with severe dysmenorrhea, few months to one year after menarche [7]. Other common symptoms are pelvic pain, abdominal mass and pressure symptoms [7]. Patients may present with foul smelling vaginal discharge due to pyocolpos[7]. Early diagnosis and treatment is essential in order to relieve the symptoms and prevent complications such as endometriosis [16]. Diagnosis is, commonly achieved clinically and with the help of both ultrasonography and MRI [9]. MRI is usually more conclusive [9]. Excision of the vaginal septum in order to drain the hematometrocolpos, is the typical management in patients with obstructed hemivagina. Vaginal excision with the use of retractors, scalpel, and scissors was the traditional method used in treating these patients [17]. Some reports have been described where laparotomy along with unilateral hysterectomy was performed [18].

However, with the new advancement and increasing experience with minimally invasive approach, hysteroscopic resection of vaginal septum became the mainstay of treatment. Several reports have shown hysteroscopic excision of the vaginal septum to be safe, easy and effective [10-12]. Additionally, this approach allows the preservation of the hymen, and thus virginity. This can be further seconded by visualization under ultrasound guidance, which is performed at the time of surgery similar to what was performed in this report. We performed hysteroscopic resection of the vaginal septum under ultrasound guidance and have kept a Foley's catheter intra-vaginal post operatively, for one day to keep the outflow tract patent. The patient was seen one and four months postoperatively where she was asymptomatic. However, repeat surgery was necessary due to closure of the previous incision after 7 months. Despite this fact, the parents were contended that preservation of the hymen was maintained and that neither laparotomy nor traditional vaginoplasty was warranted.

Previous reports have shown that patients had preserved their hymen and were symptom free after 4 months [13]. The longest follow up period with no recurrences reported in literature, was after one year [10]. Difficulty in achieving complete septal resection along with postoperative stenosis has been reported. This becomes more likely when the septum is highly located and is thick like in our patient. Patency of the outflow tract was maintained initially by a Foley's catheter of 16 F which was filled up with 15 ml of normal saline. The intention was to keep it for 6 weeks to allow for re-epitheliazation of the vaginal opening. Unfortunately, the Foley's had fallen off spontaneously the following day when the patient had gone to the toilet. This could indicate that the vaginal opening was wide enough to allow the release of the catheter. A successful attempt of placing a trachiobronchial stent in the orifice to maintain patency has been described by Cooper et al. [19]; however, this procedure had ended up by taking the patient three times to operating room obviating the need to remove the stent, in addition to disrupting the hymen. In the era of minimally invasive surgery, conservative surgery remains the mainstay of choices. However, surgical challenges can occur in cases of high and thick septae resulting in post-operative vaginal stenosis of the septal orifice and thus, the need to re-operate [20].

CONCLUSION

Obstructed Hemivagina with Ipsilateral Renal Anomaly (OHVIRA) syndrome is rare. Ultrasound and MRI can achieve the diagnosis. Hysteroscopic resection of the vaginal septum, drainage of hematometrocolpos under ultrasound guidance can be performed safely, along with the preservation of the hymen. However, repeat surgery might be indicated in few cases when the vaginal septum is high and thick. Longer follow up is warranted in such cases due to the risk of closure of the vaginal aperture along with the unintended consequences of hematometrocolpos and abdominal pain.

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