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Case Report

Case Report: Obstructed Hemivagina and Ipsilateral Renal Anomaly (OHVIRA) Syndrome

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Abstract

OHVIRA syndrome is a rare variety type III mullerian duct congenital anomaly. The syndrome includes uterine didelphys, obstructed hemivagina and ipsilateral renal anatomy, also known as Herlyn-Werner Wunderlich (HWW) syndrome. It is characterized by congenital birth defect of the lower abdominal organs and pelvic organs.

Mullerian malformation results from defective fusion of the mullerian duct during development of the female reproductive system. American fertility society classification of mullerian anomalies is the most commonly used standard classification.

We report a case of 14 year old young girl presented in outpatient clinic with complain of irregular vaginal bleeding associated with abdomino pelvic pain for 4 months. She had menarche 4 months back. The history was suggestive of developmental anomaly of vagina. Trans abdominal ultrasound done, followed by magnetic resonance imaging (MRI) of pelvis with contrast MRI is most sensitive diagnostic modality for pelvic abnormalities. After confirmation of the diagnosis surgical intervention was suggested, initially parents refused but later agreed. Surgical correction done successfully with drainage of menstrual blood and partial resection of the vaginal septum. Detailed informed consent was recorded in file.

Keywords: *Obstructed Hemivagina and Ipsilateral Renal Anomaly (OHVIRA); Herlyn-Werner Wunderlich (HWW) syndrome; Transvaginal Septum; Renal Agenesis; Laparoscopy and Hysteroscopy*

Introduction

The prevalence of congenital Mullerian duct anomalies is reported to be 1 % (Ashton et al., 1988)[1]. OHVIRA is a rare variety of congenital anomaly of mullerian duct. The syndrome includes uterus didelphys, obstructed hemivagina and ipsilateral renal anomaly, also known as Herlyn-Werner-Wunderlich (HWW) syndrome. As patient may have normal menses so diagnosis is challenging. It was first reported in 1922 [2]. Usually patients present after menarche with pelvic pain and/or a pelvic mass and rarely, in later years, with primary infertility. Strong suspicion and knowledge of this anomaly are essential for a precise diagnosis [3]. The triad of uterine didelphys, obstructed hemivagina, and ipsilateral renal agenesis, known as OHVIRA syndrome or HWW syndrome, is a congenital anomaly of Müllerian duct with associated mesonephric duct anomaly [4,5]. This is a very rare but well documented variety of mullerian anomaly. A very few cases has been reported in the literature. They generally present at puberty with nonspecific symptoms, such as pelvic pain, recurrent severe dysmenorrhea, and a palpable mass associated with hematocolpos or hematometra [6]. Hematocolpos or hematometra occurs with occlusion of vaginal canal due to the fusion of the septum with the vaginal wall [7].

Case Report

114 years young girl belonging to low socioeconomic status presented in OPD with complaint of irregular vaginal bleeding for 4 months. She had menarche 4 month back since then she had irregular vaginal bleeding and abdomino pelvic pain. On physical examination normal looking female, clinical abdominal examination was unremarkable. No obvious abnormality of external genitalia. Investigation included complete blood count, random blood sugar and detailed urine examination. Hormonal and coagulation profile were with in normal range. Transabdominal ultrasound of pelvis revealed two uterine horns along with two separate cervices, the endometrial

thickness of right uterine horn 0.6 cm & endometrial thickness of left uterine horn was 0.3 cm. Vagina appears dilated & filled with fluid containing dense internal echoes representing blood, it approximately measures 6.1x 1.8 cm. Dilated vagina had displaced the uterus & cervix posteriorly & to the left, another tubular hypoechoic Structure is seen anterior to vagina slightly to right, it measures 3.7x0.9 cm, its lower end is seen in communicating with lower 1/3rd of vagina. Both ovaries normal. Ultrasound also revealed absence of right kidney while left kidney was normal. Continuous vaginal bleeding further evaluation was done by MRI.

MRI showed evidence of didelphys uterus showing duplication of uterine cavity with obstructed endometrial cavity seen on left side, however heamatocolpos is seen on right side. It appears low or iso intense on T1W1 and high intensity on T2w2 images.

Both ovaries appear normal with no evidence of abnormal signal in both ovaries.

No free fluid seen in pelvic cavity.

No pelvic Lymphadenopathy seen.

The visualized muscles appear normal.

The Visualized bones are normal with no evidence of marrow replacement

Right kidney is not visualized while left kidney is normal

After taking permission from parents it was decided to do laparoscopy & examination under anaesthesia in 2015. Laparoscopy confirmed the presence of uterus didelphys with left sided enlarged hemi vagina and haematocolpos. Also vaginal examination under anaesthesia done, nick was given on hymen at 4 and 12 o'clock position, bulging was felt in vagina on right side gentle dissection with scissors created a space, left side cervix felt, partial incision of vertical septum released dark coloured collected blood, which was drained, a free communication between the patent upper portion of right hemi vagina and normal left canal secured. Her postoperative recovery was uneventful.

She has been on regular follow-up every 6 months. Last follow-up was in March, 2020. She had regular, periods rather heavy flow & dysmenorrhea controlled by NSAID (Non-Steroid Anti-Inflammatory drugs).

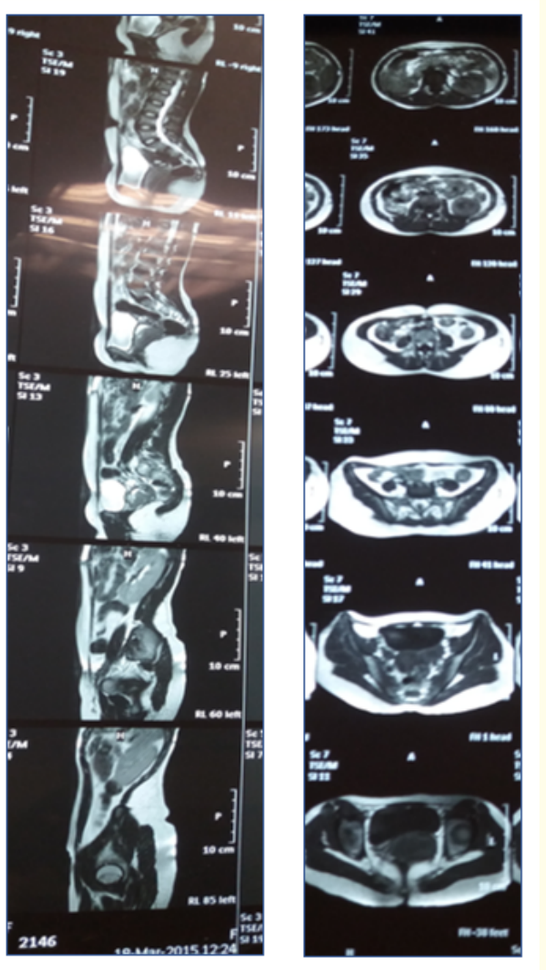

Figure 1: *MRI.*

Discussion

Although the true incidence of mullerian defects is about 1.1 - 3.5%. It is believed to be much higher (around 25%) in women with recurrent miscarriages and subfertility. Renal agenesis and uterus didelphys are strongly associated with each other (81%) [8].

The triad of uterine didelphys, obstructed hemivagina, and ipsilateral renal anomaly, known as OHVIRA syndrome, is a rare congenital anomaly of the Mullerian ducts and Wolffian structures.(9) The combination of obstructed hemivagina and uterus didelphys was first reported in 1922; (10) however the triad of obstructed hemivagina and uterus didelphys as well as an ipsilateral renal anomaly (OHVIRA syndrome) was initially reported in 1950. (11) Incidence of these anomalies is believed to be between 0.5 and 5.0%.(12, 13) The uterus, Fallopian tube, cervix and upper two thirds of the vagina develop from the paired mullerian ducts while the lower third of the vagina develops separately from the urogenital sinus.

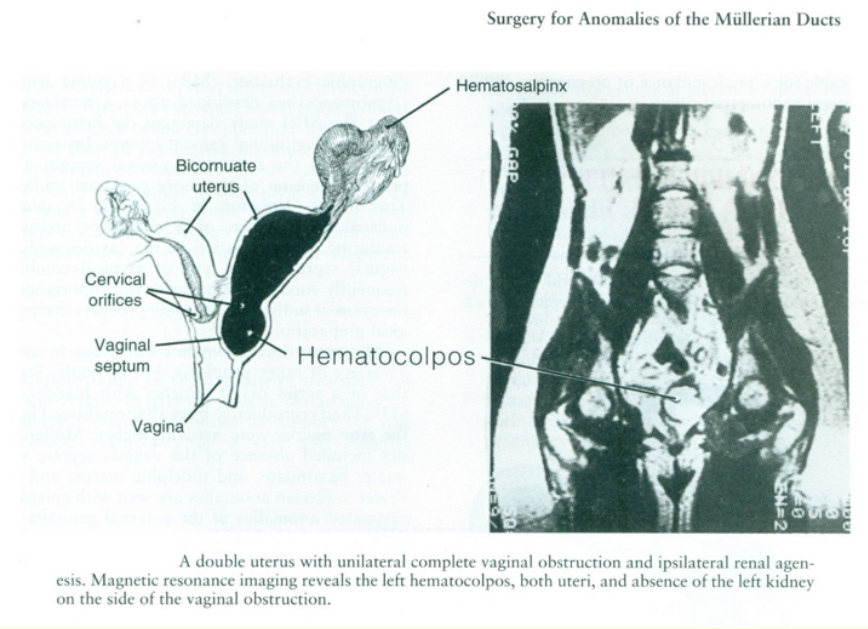

Figure: *Reference: Adapted from "Te Linde's Operative Gynecology" Tenth Edition.*

The association of uterine anomalies with renal anomalies ipsilateral to the side of obstruction can be explained by embryologic arrest at the 8th week of gestation, which simultaneously affects the mullerian and metanephric ducts. Certain other renal anomalies may also be associated, such as renal dysplasia, double collecting system and ectopic ureter [13]. The condition has also been reported to be associated with high-riding aortic bifurcation, Inferior vena cava (IVC) duplication, intestinal malrotation and ovarian malposition [14]. The most common clinical presentation is pelvic pain initiating shortly after the menarche, associated with 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 presentations may include intermenstrual bleeding, acute retention of urine, fever, vomiting, and abdominal swelling [15-18].

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 patent unobstructed hemivagina gives the impression of normal menses. Consequently accurate diagnosis and surgical treatment may be delayed for several months or even years. Imaging modalities used to diagnose this condition include ultrasonography, and MRI. Computed tomography (CT) has a limited role in evaluation of the female pelvis.

The standard surgical treatment for obstructed hemivagina is surgical excision of vaginal septum. In cases where incomplete excision is done after sometime stenosis occur & further treatment is required. Laparoscopy is utilized to confirm diagnosis or for concurrent endometriosis treatment. some surgeons utilize abdomino pelvic approach. In this patient we mainly use vaginal approach. Some studies also reported Hysteroscopic resection of vaginal septum avoiding hymenotomy if it is of concern.

Conclusion

OHVIRA syndrome is a rare variety of female urogenital anomaly, requires knowledge of this abnormality to raise suspicion when young girls present with dysmenorrhea. Generally Ultrasonography (USG) and MRI clinch the diagnosis. A surgical correction restores normal menstrual cycle, sexual and reproductive functions and to avoid long-term complications i.e. pyocolpos, chronic cryptomenorrhea and endometriosis.

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