Research Paper

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Uterus Didelphys With Obstructed Hemivagina And Ipsilateral Renal Anomaly (Ohvira Syndrome)

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KEYWORDS : OHVIRA ,Uterus didelphys, obstructed hemivagina, Hematocolpos

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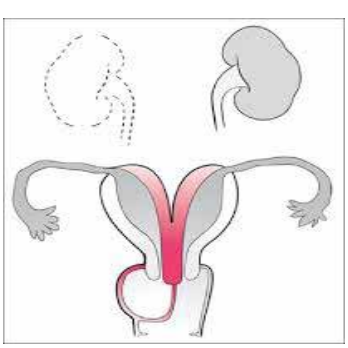
ABSTRACT

The syndrome of obstructed hemivagina and ipsilateral renal anomaly(OHVIRA) or Herlyn Werner Wunderlich is a rare congenital anomaly of the Mullerian ducts and Wolffian structure. We report a case of 27 year old female who presented with pain abdomen and bleeding per vaginum on and off diagnosed by Ultrasound and MRI and managed with excision of vaginal septum and drainage of hematocolpos.

INTRODUCTION

The syndrome obstructed hemivagina ipsilateral renal anomaly (OHVIRA) is a rare diagnosis in females that involves a combination of the triad of obstructed hemivagina, uterine anomaly (usually uterus didelphys), and ipsilateral renal anomaly. Incidence of mullerian duct anomalies ranges from 0.8% to 4% and the incidence of the OHVIRA syndrome is estimated to be between 0.1-3.5% of all mullerian anomalies.[1] The combination of obstructed hemivagina and uterus didelphys was first reported in 1922 [2]; however the triad of obstructed hemivagina and uterus didelphys as well as an ipsilateral renal anomaly was initially reported in 1950[3]. It represents a diagnostic dilemma because of the regular menstruation from unobstructed side and non specific abdominal pain. Consequently accurate diagnosis and surgical treatment can be delayed for several months or even years. OHVIRA could present with lower abdominal pain, severe dysmenorrhea, a pelvic or vaginal mass, abnormal vaginal discharge, intermenstrual bleeding [4,5], acute retention of urine, fever, vomiting [6], infertility and abdominal swelling or complication with pregnancy and labor[7] . Awareness of such anomaly is prerequisite for early and prompt diagnosis and prevention of potential complications. We present a case report of a patient with the triad of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis managed with excision of vaginal septum and drainage of hematocolpos.

FIGURE 1: . Triad of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis



CASE REPORT

A 27 year old nulliparous female with married life of 1 year presented to our institution with complain of bleeding per vaginum on and off with pain abdomen .Patient attained menarche at age of 11 year and had regular menstrual cycles, once in every 28-30 days with duration of 3-5 days menstrual flow along with irregular post menstrual discharge for 7-10 days.

General physical examination showed no abnormalities with well developed secondary sexual characteristics appropriate for age including breast and normal feminine hair distribution. Abdominal examination revealed no swelling/ tenderness. Laboratory tests were normal.

Vaginal and speculum examination revealed bulging cystic mass in right fornix , normal cervix on left side of mass.

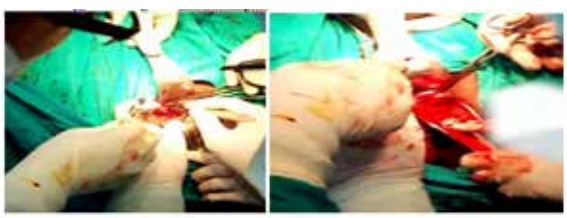
Abdominopelvic ultrasound showed normal liver, gall bladder, spleen, pancreas. Right renal fossa empty. Left kidney measured 118 mm. Pelvis showed 2 endometrial canals.Left sided canal continuing into normal cervix and vagina. Right side of canal continuing inferiorly into mild elongated collection with dense internal echos and fluid fluid level seen within it. Right ovary normal. Left ovary couldnot be clearly delineated. Features suggestive of Uterus didelphys with septate right hemivaginum with hematocolpos, non visualization of right kidney with cystitis. In presence of above findings possibility of Herlyn Werner Wunderlich syndrome (OHVIRA syndrome) should be considered.

MRI revealed 2 separate uterine cavities ,cervices and vaginas, suggestive of uterus didelphys.The left uterine cavity, cervix and vagina are normal. The right uterine cavity seen normal. Right hemivagina and cervical canal are dilated and filled with fluid which appear hyperintense on both T1W and T2W.MRI with few T2 hypointense areas suggesting blood products implicating the presence of an obstructing right vaginal septum. Right fallopian tube appears dilated and hyperintense on both T1W and T2W MRI suggestive of hematosalpinx. Left fallopian tube appears normal .Bilateral ovaries seen normal with a dominant follicle of size 20×23 mm in right ovary. Right kidney not visualized. Left kidney seen normal. The urinary bladder partially distended. The common iliac vessels, internal iliac vessels and external iliac vessels are unremarkable. Impression-Uterus didelphys with right

hematosalpinx, hematocolpos, right hemivagina obstruction and absent right kidney suggestive of Herlyn Werner Wunderlich syndrome (OHVIRA syndrome).

Hence final diagnosis of OHVIRA syndrome was made based on above clinical pictures and findings. The patient was treated by excision of vaginal septum and draining of hematocolpos. Dark altered coloured blood drained ≈60 ml. Antibiotics were continued for further 5 days. The postoperative course was uneventful.

FIGURE 2,3 : Drainage of hematocolpos.



DISCUSSION

The female reproductive tract develops at the same time and close to the urinary tract and kidneys from the intermediate mesoderm. As a result, developmental problems in the female

reproductive tract sometimes occur with problems in other areas, including the urinary tract, kidneys, such as pelvic kidney, absent kidney, duplication of the collecting system, or multicystic kidney[8] or ectopic ureters[9] . The OHVIRA syndrome is classically associated with uterus didelphys (type III, American Society for Reproductive Medicine classification)

or rarely a complete septate uterus (type V)[9,10] .The renal agenesis (mesonephric involution) on the side of the obstructed vagina associated with double uterus and double cervix is suggestive of an embryologic arrest at 8 weeks of pregnancy that simultaneously affects mullerian and metanephric ducts. The etiology of the syndrome is unknown. It is thought to be multifactorial and associated with fusion anomalies of Mullerian ducts[11]. Pelvic pain is the most common presenting symptom (90%) followed by an abdominal mass (40%) and pressure symptoms. 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 and vomiting.

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.

Early detection of Mullerian anomalies is important for counseling and planning the proper management and helps in preventing complications and preserving future fertility[12] . Ultrasonography is the usual initial imaging modality which is widely available. However MRI gives better characterization of the contents of the endometrial and cervical canal. Also any other associated adnexal pathology is better demonstrated. In case of septate uterus, septa is demonstrated unequivocally by MRI[13-16] .Two stage vaginoplasty in the form of drainage of the hematocolpos in one operation followed by another operation to resect the septum is the classic treatment option [17]. Single stage vaginoplasty, advocated in our case, in the form of drainage of the collected blood, complete septum resection followed by suturing of the lateral vaginal wall was proposed to be a suitable alternative to the two stage procedures without any complications. However, postoperative stenosis, recurrence of hematometra[18] and infection are significant possibilities necessitating a second operation. To overcome this possibility, different treatment modalities have been tried to reduce the need of a second operation and minimize the risk of postoperative reobstruction such as the use of vaginal molds, dilators[19,20] and coated tracheobronchial stent[21] The integrity of the hymen represent a major cultural issues in

our community, the hysteroscopic excision of vaginal septum in uterus didelphys has been recommended for the management of those patients with good outcome[22] .

CONCLUSION

OHVIRA syndrome is a rare complex congenital anomaly of female genital system with potential short and long term complications. The diagnosis is likely to be missed because of the normal menstruation and nonspecific abdominal pain. Hence greater awareness and early diagnosis with timely intervention can prevent future complications. In conclusion, an accurate diagnosis can typically be made on the basis of history and examination, in combination with ultrasound and MRI followed by excision of vaginal septum and drainage of hematocolpos which is an ideal approach to manage cases with obstructed hemivagina and ipsilateral renal anomaly (OHVIRA syndrome) with uterus didelphys.

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