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UTERINE DIDELPHYS WITH OBSTRUCTED HEMIVGINA AND IPSILATERAL RENAL ANOMALY (OHVIRA) SYNDROME: A RARE CASE STUDY

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ABSTRACT Uternie didelphys with obstructed Hemivagina and ipsilateral Renal anomaly (OHVIRA) syndrome is a rare congenital anomaly. These patients may present with varied symptoms. Hence may often get misdiagnosed or may go undiagnosed till a later age as the patient may be asymptomatic. We present here such an unusual case of a 36yrs old married female who came to our OPD with diffuse pain abdomen associated with dysmenorrhoea and was later diagnosed on MRI with OHVIRA syndrome who underwent septal resection and was relieved from the symptoms.

KEYWORDS : OHVIRA, MRI, septal resection

INTRODUCTION

Uterine didelphys with obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome is also known as Herlyn - Werner-Wunderlich syndrome. It is a rare congenital anomaly[1]. The incidence is around 2-3% of Müllerian abnormalities[2]. It is usually diagnosed at a young age where they can present with lower abdominal pain due to obstructed menstrual flow. But the diagnosis may get delayed because of its rarity and menstruation occurring through the other patent outflow tract[3]. The clinical presentation may vary. The most common symptoms are dysmenorrhea, abdominal pain, urinary complaints, abdominal or pelvic mass. Here is a case with the triad of uterus didelphys, obstructed hemivagina, and right renal agenesis which was diagnosed by magnetic resonance imaging (MRI) and managed.

CASE STUDY

36 yrs old married female P1L1A1 came with complaints of irregular menstrual cycles associated with dysmenorrhea since menarche. She presented to our OPD with diffuse pain abdomen on and off since two months which aggrevated during menses and subsided with analgesics. Obstetric history suggested she has a 11 yrs old female child born by LSCS which she conceived spontaneously with no obstetric complications.

PATHOLOGICAL FINDINGS

Examination revealed a bulge on the anterior vaginal wall on the right side with foul smelling greenish vaginal discharge which was sent for culture and sensitivity test which showed no growth of micro organisms. There was severe cervical motion tenderness and fornicial tenderness. USG suggested complete bicorporeal uterus with a right hematocolpos with tiny communication with left vagina. Right kidney was absent.

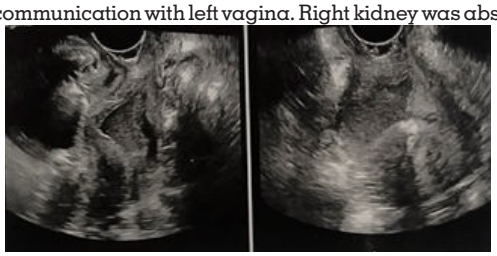

Fig 1: USG image of obstructed hemivagina with hematocolpos

MRI was used for further evaluation which revealed two separate horns of the uterus with each opening into a separate cervix suggestive of uterine didelphys. Right vaginal cavity appeared mildly distended and filled with fluid appearing hyperintense on T2W1 suggestive of haematocolpos secondary to vaginal obstruction. Right kidney was absent and based on these findings OHVIRA syndrome was suspected.

MANAGEMENT AND OUTCOME OF CASE:

Patient was treated with a course of antibiotics preoperatively. Subsequently she underwent cervicovaginal orifice reconstruction. Intraoperatively one cervical os was visualized, and a vaginal septum was noted with a 1 mm tiny communication through which grayish white discharge was seen.

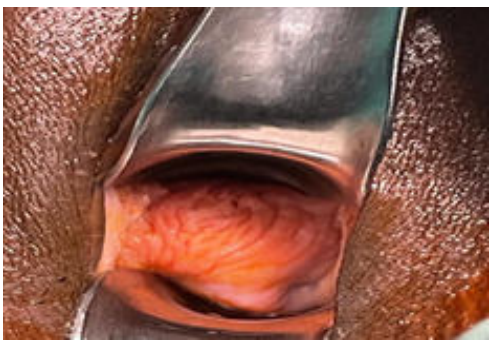

Fig 2: Bulge in right side of vagina

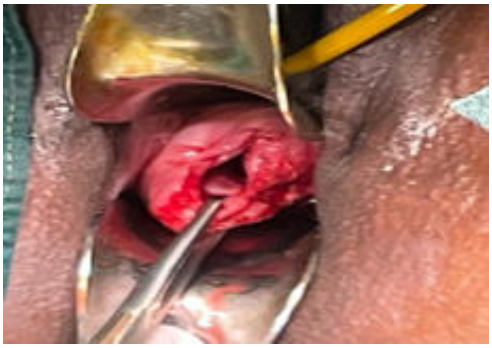

Fig 3: Septum resected

The septum was partially resected and hysteroscopy was done and the other cervix was visualized along with right tubal ostia.

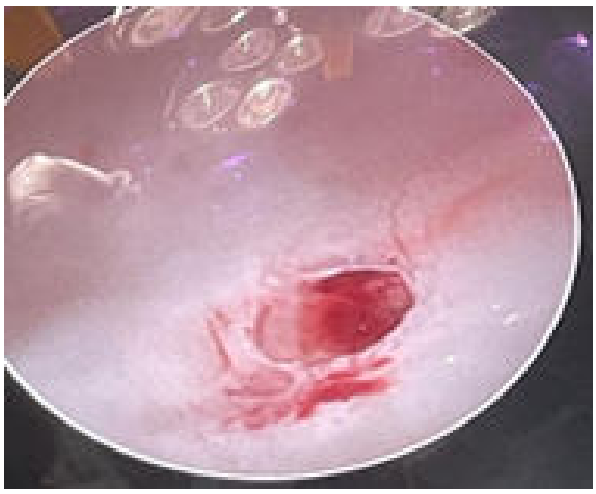

Fig 4: Post septal resection: right tubal ostia visualized through hysteroscopy

Vaginal packing was done. Patient was discharged 5 days following surgery after adequate postoperative care. When she came for subsequent followup patient had regular menstruation with no dysmenorrhea and no fresh complaints.

DISCUSSION

OHVIRA syndrome is a rare condition which includes a complex malformation of the female genital tract which often goes misdiagnosed due to nonspecific symptoms or undiagnosed due to patients being asymptomatic[4]. Female genital tract abnormalities are as a result of abnormal development of mesonephric and paramesonephric ducts and may be associated with renal abnormalities. Patients usually present with symptoms within few years of attaining menarche and the mean age of presentation is usually 15 yrs[5]. But in this case patient presented at a later age. Complications that may arise if untreated are endometriosis, infertility, pyocolpos or pyosalpinx and pelvic adhesions[6]. MRI is the gold standard technique for diagnosing any condition in the female reproductive anatomy[7]. In our patient MRI findings made the diagnosis easier. The primary aim is to relieve the symptoms by resection of as much of the obstructing vaginal septum and to ensure that the fertility of the patient is not compromised. We followed the same principle. The pregnancies rate with successful outcomes in patients with HWH syndrome is around 87%[8]. Preconceptional counseling is important and timely management can prevent adverse obstetric outcomes[9].

CONCLUSION

Thus an appropriate diagnosis of OHVIRA syndrome with timely management can relieve the symptoms and decrease the complications. A multidisciplinary approach with the help of gynecologist and radiologist can help in better diagnosis and management of this rare syndrome.

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