VOLUME-6 | ISSUE-4 | APRIL - 2017 • ISSN No 2277 - 8179 | IF : 4.176 | IC Value : 78.46

Original Research Paper

OHVIRA syndrome: Imaging in diagnosis in an asymptomatic newborn – case report

Radiology

KEYWORDS: OHVIRA syndrome, Herlyn-Werner-Wunderlich syndrome, Mullerian anomaly, obstructed vagina, renal agenesis

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ABSTRACT

OHVIRA (Obstructed hemivagina and ipsilateral renal agenesis) syndrome also known as Herlyn-Werner-Wunderlich syndrome is a rare congenital anomaly of the female urogenital tract involving both Mullerian and Wolffian ducts, characterized by the triad of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis. It usually presents and is diagnosed at puberty due to non-specific symptoms of pelvic pain with or without a pelvic or vaginal mass shortly following menarche. We present a case of this rare anomaly diagnosed in the first week of post-natal life by using imaging modalities like USG and MRI in a female neonate with no clinical symptoms.

INTRODUCTION

OHVIRA or Herlyn-Werner-Wunderlich syndrome is a triad of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis. The prevalence of uterine anomalies diagnosed is 5.5% in an unselected population, 8% in infertile women, and higher in those with miscarriage (13.3%) and in infertile women who also had a history of miscarriage (24.5%) (1). The mean incidence of uterus didelphys in Mullerian duct anomalies is 11.1%. (2). Renal agenesis is more commonly seen in uterus didelphys than in other types of Mullerian duct anomalies (3). The incidence of OHVIRA syndrome is small and very few isolated case reports have been published. Most of these cases have been diagnosed post menarche as the patient presents with symptoms of acute pelvic pain, severe dysmenorrhea and vaginal or pelvic mass that occur secondary to hematocolpos or hematometra. We present an unusual case diagnosed antenatally as left renal agenesis and diagnosed as OHVIRA syndrome in the first week of postnatal life using USG and MRI, in a female neonate with no clinical symptoms.

CASE REPORT

The patient was a female newborn, gravida 1, para 1, born of normal vaginal delivery at 39 weeks 2 days of gestation. Her birth weight was 3100 grams. She was the first child of healthy and non-consanguineous parents with the mother 24 years old and father 25 years old. Antenatal sonography done outside had revealed absent left kidney suggestive of left renal agenesis. She was sent to us for postnatal sonography at day 1 of life for confirmation of left renal agenesis.

USG findings:

Trans-abdominal sonography was performed with low and high frequency transducers.

Abdominal sonography revealed empty left renal fossa with left adrenal gland oriented parallel to the spine. There was no evidence of an ectopic left kidney. The right kidney was normal measuring 4.6 x 2.5 cms in size and situated in the right renal fossa.

Trans-abdominal pelvis sonography revealed presence of 2 uterine horns with normal endometrial thickness, 2 cervices and 2 hemivaginas suggestive of uterus didelphys. The left hemivagina was dilated with fluid with internal echoes within. The distal part of left hemivagina was not canalized. The right hemivagina was collapsed.

These findings were suggestive of OHVIRA syndrome and a MRI was done for confirmation of the findings at day 4 of life.

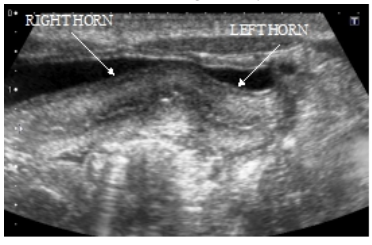

Figure 1: Trans-abdominal Ultrasound grey scale image in transverse section shows two uterine horns in transverse section with normal endometrial thickness

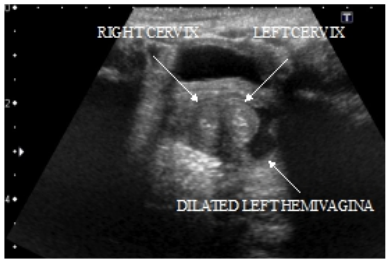

Figure 2: Trans-abdominal Ultrasound grey scale image in transverse section shows two cervices with dilated left hemivagina

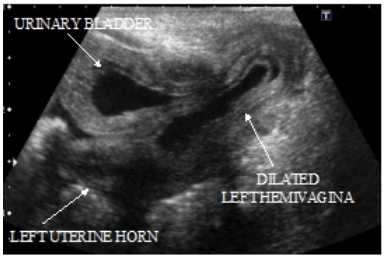

Figure 3: Trans-abdominal Ultrasound grey scale image in longitudinal section shows left uterine horn with dilated left hemivagina with non-canalization of its distal part

VOLUME-6 | ISSUE-4 | APRIL - 2017 • ISSN No 2277 - 8179 | IF : 4.176 | IC Value : 78.46

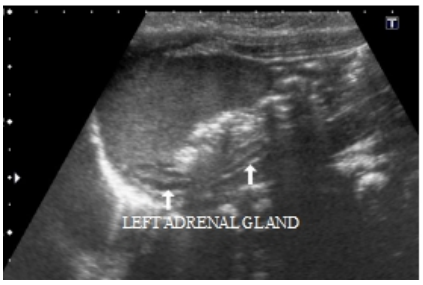

Figure 4: Trans-abdominal Ultrasound grey scale image in longitudinal section shows empty left renal fossa with lying down left adrenal gland

MRI findings:
MRI was performed on 3T MRI scanner with informed consent obtained from the mother of the female child. It confirmed the findings of 2 separate endometrial cavities, 2 separate cervices and 2 separate vaginas suggestive of uterus didelphys. There was presence of T1 hypointense and T2 hyperintense dilated left hemivagina measuring 1.3 x 1.2 x 2.7 cms not showing diffusion restriction or blooming on GRE suggestive of hydrocolpos. The left hemivagina was falling short of 9 mm from the skin surface. The right hemivagina appeared normal.

Both uterine horns and cervices were similar in size measuring 1.4 x 0.8 x 0.6 cms. Both the endometrial cavities were normal with endometrial thickness measuring 1.1 mm and 1.3 mm in right horn and left horn respectively.

Bilateral adnexae appear normal.

Left renal fossa was empty with no ectopic kidney. This was suggestive of left renal agenesis. The right kidney was normal situated in right renal fossa.

Final diagnosis of Uterus didelphys and Obstructed left hemivagina with ipsilateral renal agenesis (OHVIRA) with resultant left hydrocolpos was given.

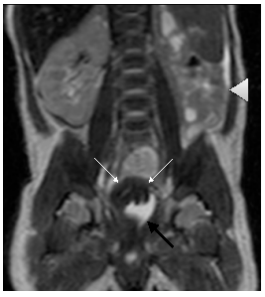

Figure 5: MRI - T2 coronal image shows two uterine horns with normal endometrial thickness (white arrows) with dilated left hemivagina (black arrow) and normal right hemivagina. Also empty left renal fossa (white arrow head) is seen.

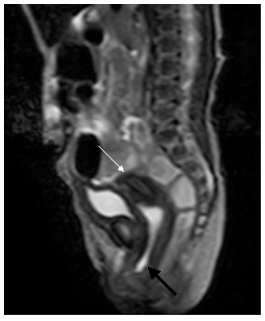

Figure 6: MRI - T2 sagittal image shows left uterine horn (white arrow) with dilated and obstructed left hemivagina (black arrow)

DISCUSSION
OHVIRA syndrome is a combination of type III Mullerian duct anomaly (uterus didelphys), as per the Mullerian duct anomaly classification proposed by Buttram and Gibbons (4), and Wolffian (mesonephric) duct anomaly with vaginal septum.

The uterus and cervix develop from the fusion of paired Mullerian ducts. The classic theory of vaginal development says that upper part of vagina develops from Mullerian ducts and the lower part from sinovaginal bulbs derived from urogenital sinus. The fusion of the 2 sinovaginal bulbs form the vaginal plate, which canalize later to form vaginal lumen. However, this theory was found inadequate in explaining the complex congenital Mullerian anomalies like OHVIRA syndrome. Therefore, Acien proposed a new theory of vaginal origin (5) according to which the uterus and cervix were derived from Mullerian ducts while the vagina was completely of mesonephric (Wolffian) origin although its lining was derived from Mullerian tubercle. Mesonephros is responsible for development and positioning of paired Mullerian duct in close proximity. At around 9 weeks gestation, the Mullerian duct is positioned in such a manner that it is lateral to mesonephric duct in first part, crosses it anteriorly and lies medial to it in converging portion. Due to failed positioning of paired Mullerian duct, the two hemiuteri and hemicervices fail to unite, resulting in uterus didelphys. In OHVIRA syndrome, developmental arrest of ipsilateral mesonephric duct results in failure of distal hemivagina to develop, thereby resulting in obstructed hemivagina. Further an early failure of metanephric diverticulum to develop (around 5 weeks) from mesonephric duct results in agenesis of ureteric bud, which leads to agenesis of ipsilateral ureter and kidney. Thus, all the 3 components of OHVIRA syndrome, namely uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis is secondary to mesonephric duct-induced Mullerian anomalies.

The classic renal manifestation of OHVIRA syndrome is ipsilateral renal agenesis, but reports of duplicated kidneys, dysplastic kidneys, rectovesical bands, or crossed fused ectopia have also been described (6).

Most of the patients suffering from this syndrome are diagnosed late due to its rarity and the nonspecific clinical presentation. Typically, the most common finding of a patient with OHVIRA syndrome is pelvic pain shortly following menarche in association with vaginal or pelvic mass and normal menstrual periods. The normal menstrual flow comes from the patent unobstructed hemivagina and is responsible for further delay in diagnosis. The obstructed hemivagina results in hematometra and hematocolpos in the ipsilateral horn. This gets often mis-diagnosed as endometriosis and accurate diagnosis and surgical treatment may be further delayed for several months or even years unless a high index of suspicion is maintained. Also there are increased chances of development of pelvic endometriosis due to retrograde menstrual seeding in these patients further complicating the diagnosis.

The potential complications of this syndrome are distinct in acute complications, such as pyohematometrocolpos, pyosalpinx, or pelviperitonitis, and long-term complications, such as endometriosis, pelvic adhesions and increased risk of abortion or infertility.

It is difficult to diagnose OHVIRA syndrome in infancy, as they are asymptomatic. In a newborn, the maternal hormones are responsible for secretions, which result in hydrocolpos on the side of obstructed hemivagina, which can be diagnosed by imaging modalities besides uterus didelphys and ipsilateral renal agenesis. Early diagnosis like in our case can not only reduce the morbidity and complications but also increase the pregnancy rate in future.

Imaging modalities used to diagnose this condition include ultrasonography and MRI. Computed tomography (CT) has a limited role in evaluation of the female pelvis because of the poor soft tissue

resolution and further needs to be avoided in newborn due to the radiation exposure. MRI is the most accurate diagnostic method and has the advantages of multiplanar capabilities, better soft resolution and lack of radiation exposure. Laparoscopy is not needed for the diagnosis of most of the cases.

The treatment requires surgical intervention in the form of excision of vaginal septum to relieve obstruction and letting the collection in obstructed hemivagina drain into the patent hemivagina. In fact, hemihystrectomy, done in earlier times, is no more preferred now as the reported incidence of pregnancy in both horns is almost equal. (7) If remained undiagnosed in early life, the hydrocolpos may get infected resulting in pyocolpos or post menarche the patient can develop hematometra and hematocolpos with other complications like endometriosis, which may require a more aggressive surgery.

So, it is important to investigate for urinary tract anomalies in all patients with Mullerian ducts anomaly. But it is also important to investigate for Mullerian duct anomalies in all female patients with urinary tract anomalies like renal agenesis, hypoplastic or dysplastic kidneys and crossed fused ectopic kidneys, particularly early in life before the onset of menses to reduce the morbidity and complications arising from obstruction, and increase the pregnancy rate in future.

In conclusion, OHVIRA syndrome can be diagnosed in a female newborn even in absence of clinical symptoms, using USG and MRI, if a high index of suspicion is maintained following antenatal or postnatal diagnosis of urinary tract anomaly like renal agenesis or dysplastic kidney. MRI helps to give a more accurate diagnosis. Early diagnosis can result in early treatment and reduction of morbidities and complications associated with a delayed diagnosis.

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