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Original Research Paper

Radio-Diagnosis

OHVIRA SYNDROME – A RARE CASE REPORT

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ABSTRACT **Aims And Objectives Of Study:** 1. To evaluate the role of magnetic resonance imaging (MRI) in diagnosing uterine anomalies which helps in the early detection. 2. To aid the gynecologist/surgeon for prompt surgical intervention and hence preventing adverse outcomes. **Materials And Methods:** 19-year-old adolescent girl presented to the Gynecology Department with history of menarche 8 months back having complaints of pain in lower abdomen and irregular cycles. The patient was referred to the Department of Radio-diagnosis at SIMS & RC, Bangalore for further evaluation. The radiologist evaluated the case separately by MRI examination, using GE 1.5T MRI. Multiplanar multisequence MRI of female pelvis was performed and the imaging features were recorded. The imaging features and final diagnosis were documented in a structured case record form. **Result:** MRI revealed two uterine horns with two external os opening into two separate vaginal canals. Marked distention of right hemivagina with fluid levels due to transverse septum at lower third of right hemi vagina and right renal agenesis were noted. Per operative findings confirmed the imaging findings. **Conclusion:** In our study we found that the knowledge of the syndrome helped in early detection and the timely diagnosis by imaging which aided the gynaecologist for the prompt surgical intervention preventing adverse outcomes.

KEYWORDS : Didelphys, OHVIRA, HWWS, renal agenesis

BACKGROUND:

Obstructed hemi-vagina and ipsilateral renal agenesis (OHVIRA) syndrome is a rare congenital anomaly of paramesonephric and mesonephric ducts [1]. It is also called as Herlyn-Werner-Wunderlich syndrome (HWWS).

The patients are generally adolescent girls who present with cyclical pain abdomen and dysmenorrhea. This syndrome was earlier described by Herlyn and Werner [2]. Wunderlich described renal aplasia with bicornuate uterus and isolated hematocolpos [3].

Case Report:

19-year-old young female patient was referred to the Department of Radiology for MRI pelvis with history of menarche 8 months back having complaints of pain in lower abdomen and irregular cycles. On examination, no mass per abdomen was felt. On per speculum examination vulva appeared normal.

MRI pelvis showed two uterine horns with two cervices and the external Os opening into two separate vaginal canals.

There was marked distension of right hemivagina with fluid – fluid levels; the dependant fluid showed diffusion restriction with few foci of blooming on GRE sequences. Transverse band was noted in distal third of right hemivagina. Both ovaries were normal (Fig. 1a-c).

A diagnosis of uterine didelphys with right sided hemato/pyo-colpos secondary to transverse vaginal septum was made. Further MRI abdomen was performed in suspicion for renal anomalies which revealed, right renal agenesis with compensatory hypertrophy of the left kidney (Fig. 1d).

Above constellation of findings lead to the final diagnosis of OHVIRA syndrome.

Following which, patient was undertaken for diagnostic laparoscopy and the intra-operative findings were as follows (Fig. 2); transverse hemivaginal septum on right side with hematocolpos, for which incision and drainage was performed.

Left ureter was visualised by peristaltic movements. Right ureter was absent. Uterine didelphys with bilateral normal ovaries and patent fallopian tubes.

This was followed by vaginoscopy and right-side hysteroscopy which revealed normal cervical canal, endometrial cavity, internal os and right ostia.

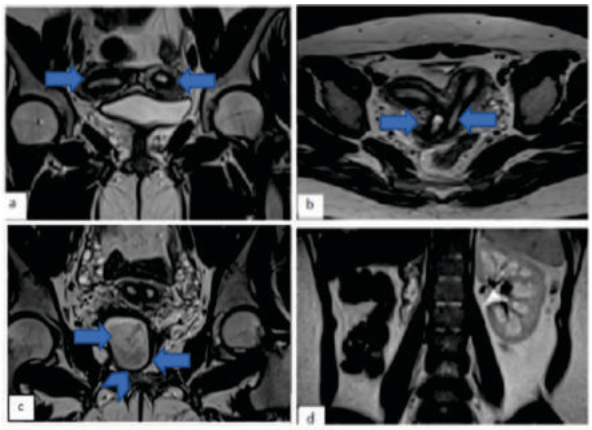

Fig. 1: MRI findings; **1a** - T2 coronal image showing two separate horns of the uterus depicted by two block arrows. **1b** - T2 axial image showing two separate cervical canals depicted by two block arrows.

1c - T2 coronal image showing distended right vaginal canal with heterogenous collection and non-distended left hemivagina depicted by the block arrows.

Transverse vaginal septum depicted by the arrowhead. **1d** - T2 coronal image showing absent right kidney with hypertrophy of left kidney.

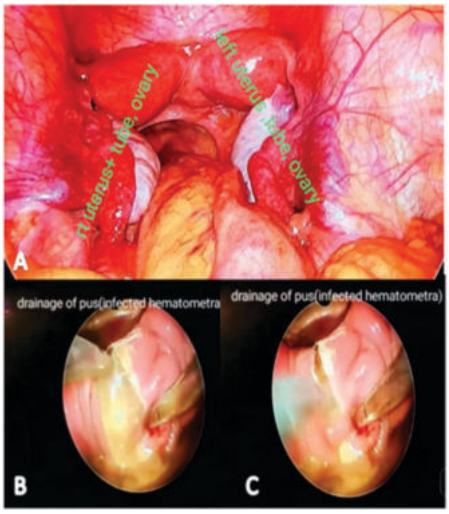

Fig. 2: Intraoperative findings;**2A** – shows two uterine horns (right and left) with two tubes and ovaries **2B & C** – shows drainage of pus from the pyometra and pyocolpos.

DISCUSSION:

The pathophysiology of OHVIRA syndrome is related to the abnormality in the development of mesonephric and paramesonephric ducts which are also called as Wolffian and Mullerian ducts respectively. The developmental abnormality of mesonephric ducts will result in renal agenesis. Obstructed hemivagina is due to lateral displacement of Mullerian duct as a result of ipsilateral Wolffian duct agenesis and thus, there is no communication between urogenital sinus and the Mullerian duct [4]. The failure to fusion will result in didelphys uterus.

OHVIRA syndrome has been classified based on complete and incomplete obstruction of hemivagina by Lan Zhu et. Al [5] which is as follows (Fig. 3);

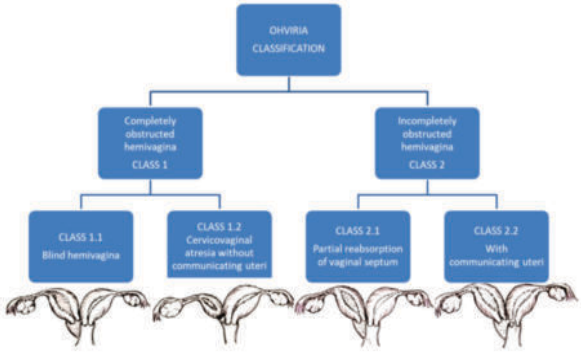

Fig. 3: Classification of OHVIRA syndrome by Lan Zhu et.al. Image courtesy - Zhu et al. New classification of Herlyn-Werner-Wunderlich syndrome. Chinese medical journal. 2015 Jan 20;128(02):222-5 [5].

DIAGNOSIS:

Imaging modalities of choice are USG and MRI with MRI being diagnostic as it clearly demonstrates the anatomy of uterus and vagina [6]. Patients present late in general as there is normal menstruation from one side and also because of the treatment with NSAIDs and OCPs for cyclical dysmenorrhea and lower abdominal pain. It is associated with other anomalies like renal dysplasia, ectopic ureter, high bifurcation of aorta, duplication of kidney, ureter and IVC [4].

Early detection is crucial for the patient in preventing the complications like pelvic endometriosis, pyocolpos and adverse outcomes in pregnancy which include miscarriages, preterm delivery and foetal growth restriction. In addition, associated renal anomalies can cause further morbidity to the patient. Rare complications like adenocarcinoma of obstructed side of cervix and clear cell carcinoma of obstructed side of vagina have also been reported [4].

CONCLUSION:

OHVIRA syndrome has a variable onset of presentation. While evaluating for uterine and vaginal anomalies, renal anomalies need to be looked into. USG is the primary modality in evaluation with MRI being the gold standard for diagnosis and preoperative planning. Thus, knowledge of the syndrome helps in early detection and preventing adverse outcomes.

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