Case 13438

Eurorad

Herlyn-Werner-Wunderlich syndrome - OHVIRA syndrome: a rare congenital anomaly

Published on 19.06.2016

DOI: 10.1594/EURORAD/CASE.13438
ISSN: 1563-4086
Section: Genital (female) imaging
Area of Interest: Genital / Reproductive system female
Procedure: Imaging sequences
Imaging Technique: MR
Special Focus: Congenital Case Type: Clinical Cases
Authors: Shaista Shoukat, Warda Sattar, Kausar Illahi Bux.
Patient: 22 years, female

Clinical History:

22-year-old unmarried woman presented to the outpatient department complaining of irregular menstrual cycle and lower abdominal pain for 3 months. She attained menarche at the age of 12 years. Physical examination showed well-developed breasts and normal axillary and pubic hair distribution. Baseline laboratory analysis and hormonal profile were normal.
Imaging Findings:

MRI examination of the pelvis was done. Multiplanar and multisequential imaging including T1WI, T2WI, T2-FATSAT sequences were acquired.
The examination showed two uteri and cervices separated by a longitudinal vaginal septum. (Fig. 1) The right uterine cavity and cervix were distended and obliterated by fluid with blood MR signal returning heterogenously high signals on T1 and T2 weighted image. (Fig. 1a, 2, 3)
Right kidney and ureter were not visualized. Left kidney was seen at its normal anatomical location.
Discussion:

OHVIRA (obstructed hemi-vagina and ipsilateral renal anomaly)/Herlyn-Werner-Wunderlich (HWW) syndrome is a rare complex congenital anomaly of the female genital system. It is a triad of obstructed hemi-vagina, uterine 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 aetiology of the syndrome is unknown. However, it is multifactorial and associated with Mullerian ducts fusion anomalies in utero.

The classical presentation is of a young girl presenting with severe dysmenorrhea, few months to years after attaining menarche. Pelvic pain is the most common presenting symptom (90%) followed by an abdominal mass (40%) and pressure symptoms. Patients can also present at a later age with foul-smelling vaginal discharge due to pyocolpos. [2] Because of regular menstruation in these patients (at menarche), diagnosis is often delayed. Patients can present with complications like endometriosis, hydrosalpinx, secondary infection and pelvic adhesions causing chronic pelvic pain. Thus if not diagnosed and treated in time, it can lead to long-term problems like infertility.

The diagnostic modalities commonly used are ultrasonography and MRI. Ultrasound can diagnose the collection

inside the uterus or vagina (secondary to obstruction, and may raise the differential of adenexal masses i.e. endometriomas and cystadenoma), but cannot identify the type of Mullerian anomaly, while MRI due to its multiplanar and tissue characterization abilities can give precise information about uterine morphology and about the continuity with each vaginal (obstructed and non-obstructed) lumen. MRI has more sensitivity in detecting the uterine contour, the shape of the cavity, the presence of a septum, as well as the presence of the associated pathology such as endometriosis, pelvic inflammation, and adhesions. Thus MRI is the gold standard method for accurate diagnosis and allows the most appropriate treatment option to be used. [3]

The standard management of these patients is excision of vaginal septum and drainage of haematometrocolpos. A simple excision of the vaginal septum can relieve the patient of her symptoms and avoid infertility. [4] Altchek and Paciuc have reported pregnancy to occur twice in a previously-obstructed didelphys uterus after surgical correction. [5] The surgeon must, therefore, make every effort to preserve the obstructed uterus.

In conclusion, greater awareness of rare entity and early diagnosis with timely intervention can prevent patient future complications. MRI has proved to be of great help in correct diagnosis of this rare anomaly, avoiding laparoscopy.
Differential Diagnosis List: Herlyn-Werner-Wunderlich syndrome / OHVIRA syndrome, Ovarian cystadenoma, Endometrioma, Mullerian duct anomaly (Bicornuate uterus)

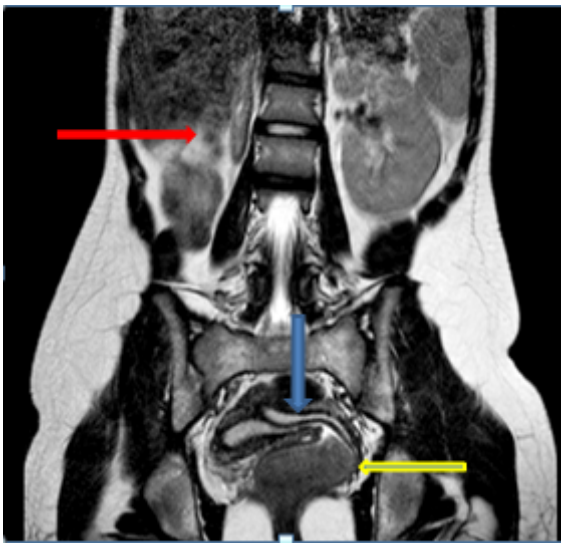
Final Diagnosis: Herlyn-Werner-Wunderlich syndrome / OHVIRA syndrome

References:

Resetkova N, Christianson M, Kolp L. (2012) Uterinedidelphys with obstructed hemivagina and ipsilateral renal agenesis with hydronephrosis. Fertil Steril 97:s30-1.
Dhar H, Yasser AR, Illham H. (2011) Uterusdidelphys with obstructed hemi vagina, ipsilateral renal agenesis and right pyocolpos: a case report. Oman Med J 26:447-50. (PMID: 22253958)
Saxena N, Channawar S, Khadkikar R, Chauhan AR (2014) OHVIRA Syndrome. JPGO Volume 1 Issue 12
Bhimarao Patil, Rashmi Mysore Nagaraju (2015) Uterine didelphys with obstructed hemivagina and ipsilateral renal anomaly - OHVIRA syndrome: a rare congenital anomaly. ijrcog (3):889-892
Altchek A, Paciuc J (2009) Successful pregnancy following surgery in the obstructed uterus in a uterus didelphys with unilateral distal vaginal agenesis and ipsilateral renal agenesis:Case report and literature review. J Pediatr Adolesc Gynecol 22:e159–62. (PMID: 19576808)

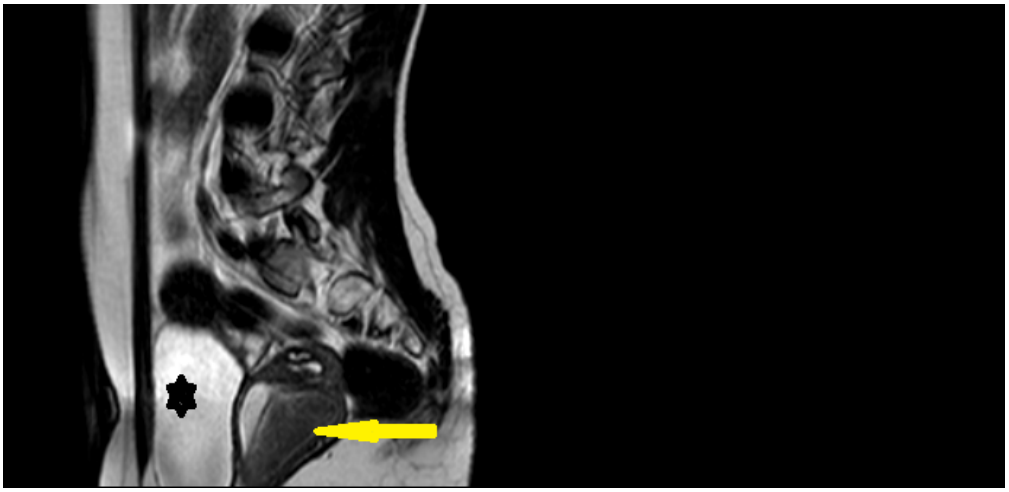
Figure 1

a



Description: MRI, T2WI coronal image: didelphys uterus as two separate uterus and cervices (blue arrow) hematocolpos in one of the cervical cavities (yellow arrow)
non visulaization of right kidney (red arrow) **Origin:** Daignostic radiology department. JPMC.

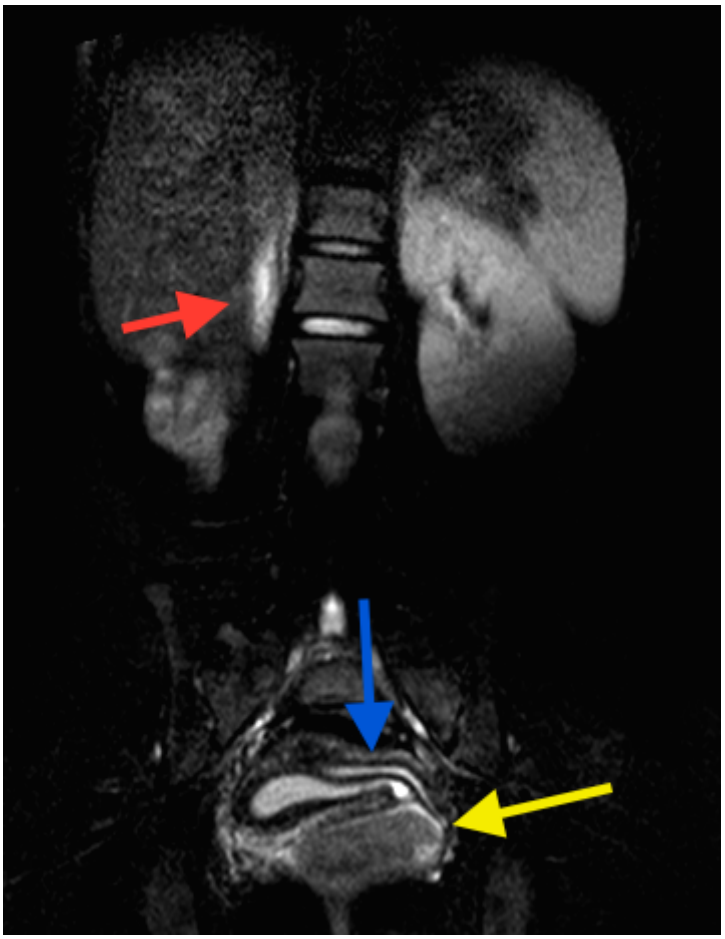
b



Description: MRI, T2WI sagital image: hematocolpos in one of the cervical cavities (yellow arrow), urinary bladder (asterisk). **Origin:** Daignostic radiology department. JPMC.

Figure 2

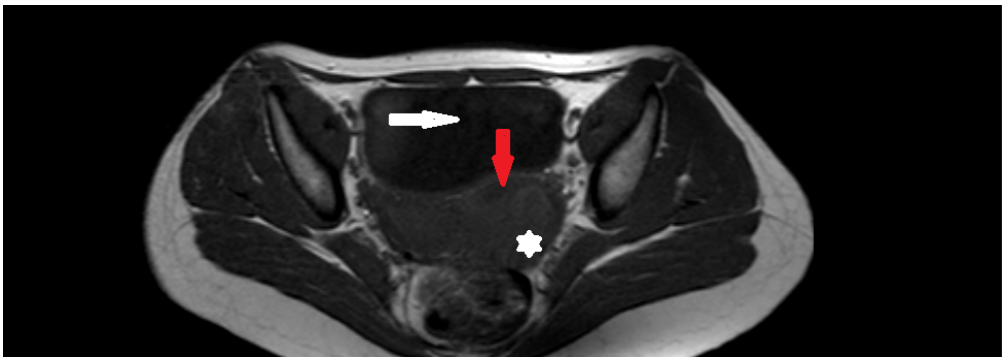
a



Description: MRI T2-FS coronal: non-fatty high T2 signal within the obstructed horns (blue arrow), haemtocolpos (yellow arrow), non-visualization of right kidney (red arrow). **Origin:** daignostic radiology department.JPMC

Figure 3

a



Description: MRI, T1WI axial image:
Bicornuate uterus (red arrow). Blood showing relatively heterogenously high signal in one of the cervical cavities (asterisk) in comparison to low fluid signal in urinary bladder (white arrow). **Origin:** Department of daignostic radiology. JPMC