EUROPEAN JOURNAL OF BIOMEDICAL AND PHARMACEUTICAL SCIENCES
http://www.ejbps.com

ISSN 2349-8870
Volume: 8
Issue: 4
199-203
Year: 2021

HERLYN-WERNER-WUNDERLICH SYNDROME CLASSIFICATION 2.2, LATE PRESENTATION IN PATIENT WITH ONE SUCCESSFUL PREGNANCY OUTCOME: SONOGRAPHIC AND MAGNETIC RESONANCE (MR) IMAGING FINDINGS OF THIS UNCOMMON UROGENITAL ANOMALY

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Article Received on 29/01/2020 **Article Revised on 19/02/2021** **Article Accepted on 09/03/2021**

ABSTRACT
Herlyn-Werner-Wunderlich syndrome is an uncommon anomaly affecting the genitourinary system which is typified by duplicated uterus and vagina along with homolateral agenetic kidney. Ultrasound and MRI are useful in delineating uterine morphology, obstructed hemivagina and absent kidney. Very rare case of this syndrome with late manifestation in a 26-year-old woman, mother of 5 year old child who presented with bleeding per vaginum, pelvic pain and pelvic mass is presented in this report.

KEYWORDS: Herlyn-Werner-Wunderlich syndrome (HWWS), Magnetic resonance imaging (MRI), Ultrasonography (USG), Uterus didelphys, Blind hemivagina.

BACKGROUND
Herlyn-Werner-Wunderlich syndrome (HWWS) is characterized by uterus didelphys, blind hemivagina and ipsilateral renal agenesis[1] therefore it is also known as Obstructed hemivagina and Ipsilateral Renal Anomaly (OHVIRA).[2] Here is case of Herlyn-Werner-Wunderlich syndrome in a 26-year-old married woman, mother of 5 year old child who presented with bleeding per vaginum, pelvic pain and pelvic mass.

CASE PRESENTATION
26 year old married women presented with lower abdominal pain, intermittent spotting with passage of clots in between menstrual cycles for the past 2 years. There was history of emergency lower segment caesarean section in view of oligohydramnios in first pregnancy. On PV examination whole of vagina was filled by polypoidal mass, shaggy surface, cervix could not be felt, uterus can't be made out. The patient was sent for ultrasonography which revealed large cystic lesion with internal echoes posterior to vaginal wall, double uterine cavity, double cervix with absent left kidney. MRI was done for better characterization of pelvic anatomy. MRI revealed two separate uteri each with a separate cervix and vagina with e/o small communication between the two at the level of cervix. The left hemivagina was overdistended, showing high signal intensity contents on T1W as well as T2W s/o haemorrhagic contents along with dependent blood clots. The haemorrhagic contents(mildly hyperintense on T1) were also seen in both the uterine cavities (left >right). The right vagina was stretched and pushed laterally by the over distended left vagina. The obstructed left hemivagina showed septum distal to the site of obstruction. Left kidney was not visualized in left renal fossa. B/l ovaries were normal. These MRI findings of uterus didelphys with two cervix and two hemivagina with obstructed left hemivagina leading to left hematocolpos with b/l heamatometra (left >right) with e/o communication at the level of cervix with agenesis of left kidney were suggestive of OHVIRA (Obstructed hemivagina with ipsilateral renal agenesis) syndrome[Classification 2.2]. The patient underwent surgery. The septum was excised and hematocolpos and hematometra was drained.

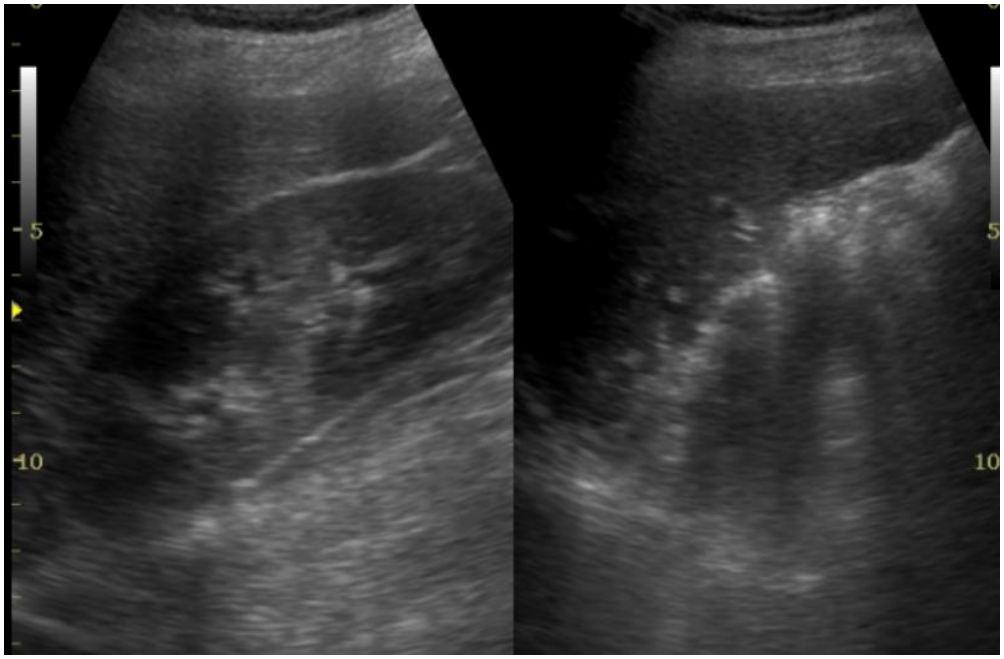


Figure1. Ultrasound images showing Right kidney in right renal fossa and empty left renal fossa.

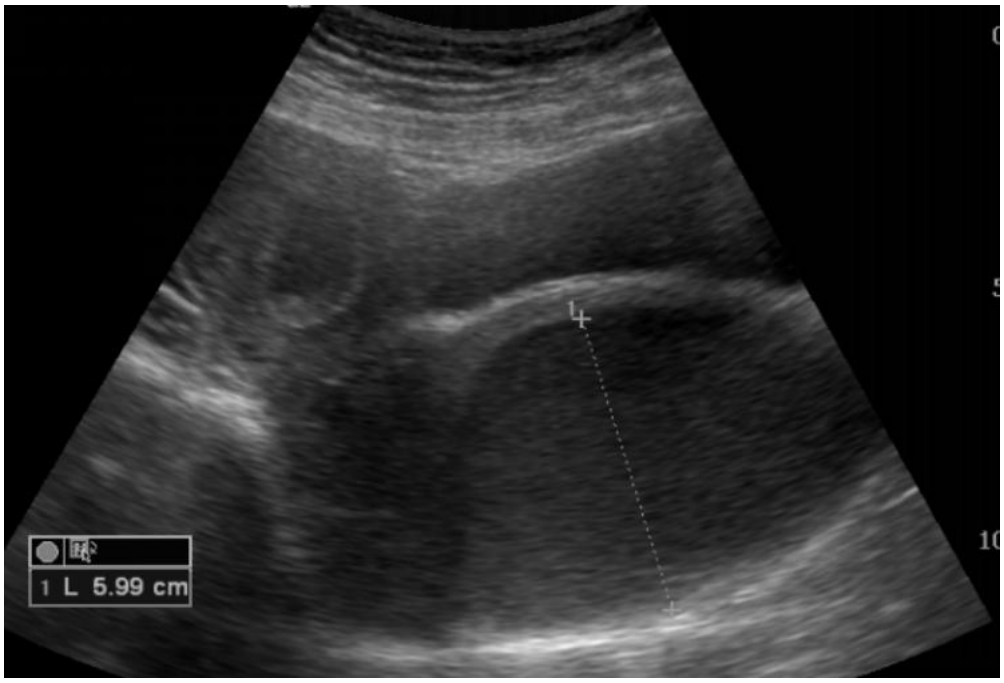
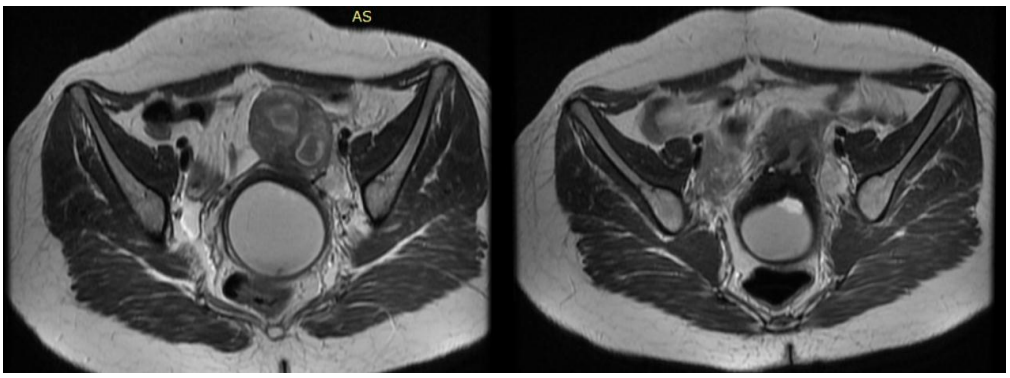


Figure2. Ultrasound image showing large cystic lesion with internal echoes posterior to vaginal wall.



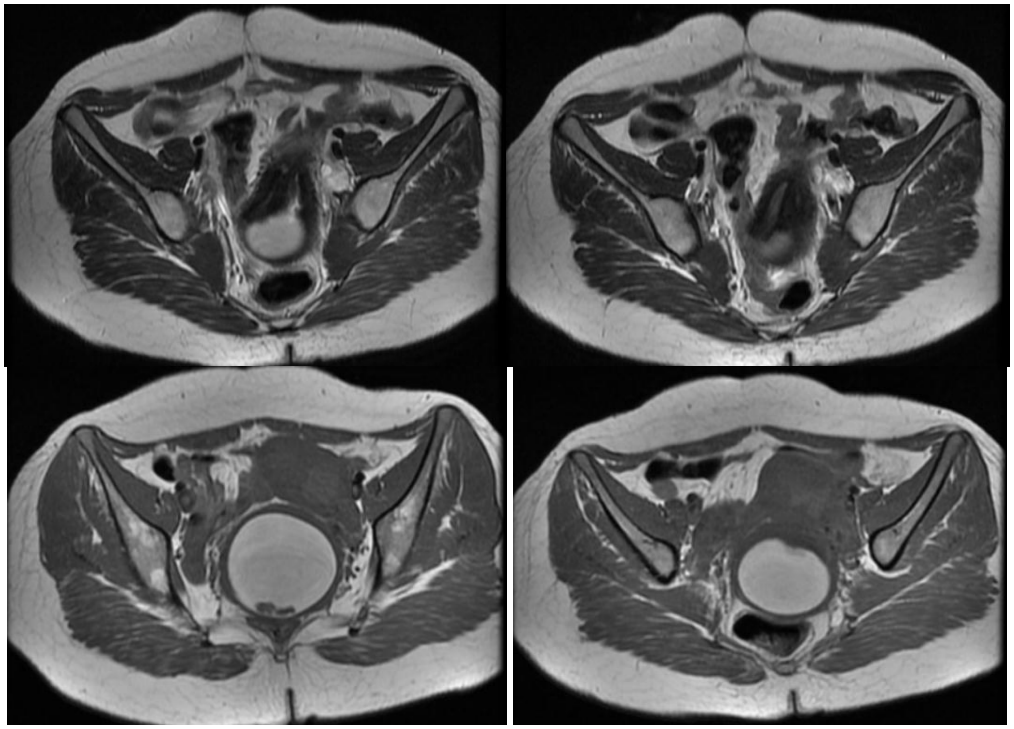


Figure 3: Axial MRI images(cranial to caudal) showing two separate uteri (top right image) each with separate cervix(top left image) and vagina(middle right image) with e/o small communication between the two at the level of cervix(top left image). The left hemivagina was overdistended, showing high signal intensity contents on T1W(bottom row images) as well as T2W(top and middle row images) s/o haemorrhagic contents along with dependent blood clots. The haemorrhagic contents(mildly hyperintense on T1) were also seen in both the uterine cavities (left >right). The right vagina was stretched and pushed laterally by the over distended left vagina.

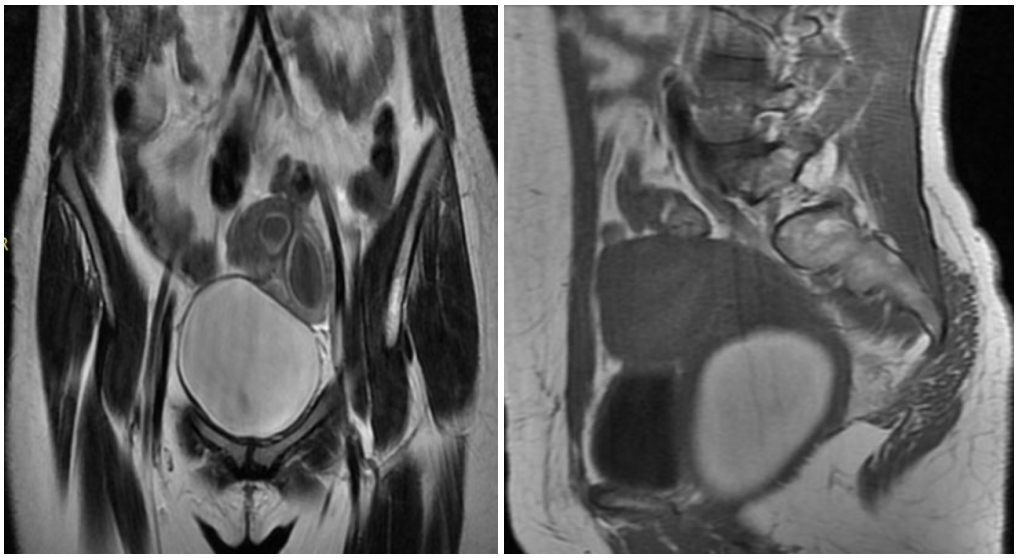


Figure 4: T2W coronal and T1W sagittal MRI images showing distended left hemivagina showing T2,T1 hyperintense signal suggestive of hematocolpos. Two distended uterine cavities with mildly T1 hyperintense signal suggestive haematometra are also seen.

DISCUSSION

Herlyn-Werner-Wunderlich syndrome is a uncommon type of uterovaginal duplication, with three types of aberrations which are didelphys uterus, unilateral obstructed hemivagina and ipsilateral renal agenesis.[3] Müllerian duct anomalies have an incidence of 1-5% in the general population and 13-25% in women with recurrent pregnancy loss, with OHVIRA constituting 0.1610% of these anomalies.[4] OHVIRA syndrome results from an embryologic anomaly occurring during the 8th week of gestation that involves both the metanephric ducts and paramesonephric ducts. Failure of complete lateral fusion of the paramesonephric ducts results in uterus didelphys and lack of development of

the metanephric duct results in ipsilateral renal agenesis.[5]

During embryogenesis, absence of Müllerian Inhibiting Factor after 6 weeks stimulate bidirectional growth of paired Müllerian (Paramesonephric) ducts in females. B/L paramesonephric ducts cross the Wolffian ducts (which act as inductors for promoting the former's growth), migrate towards each other and fuse leading to formation of uterovaginal primordium, with regression of the Wolffian ducts at the same time. By 9th week, there occur lateral fusion of the caudal part of medially migrating paramesonephric ducts and intervening septa get resorbed, leading to formation of a single uterovaginal canal which inturn forms uterus and upper two third of vagina. The upper portion of the Müllerian ducts which do not fuse form the fallopian tubes. Urogenital sinus forms the lower one third of the vagina. Vertical fusion takes place between the cranially moving sinovaginal bulb, which forms the vaginal plate and joins the canal.[6]

In HWWS embryonic arrest of the caudal portion of one of the Wolffian duct from an insult as early as 4th gestation week causes its maldevelopment, preventing the cross over and subsequent fusion of the Mullerian ducts.[7] The side on which wolffian duct is absent , the paramesonephric duct of that side gets laterally displaced, due to which it cannot come in the centre to fuse with the contralateral duct and each duct forms a separate cervix and hemivagina, leading to uterus didelphys. Complete non union between the paramesonephric ducts and the cranially migrating uterovaginal bulb results in a blind sac or vaginal atresia; while partial non fusion produces a remnant transverse vaginal septum which may be incomplete or complete with obstruction and hematometra or hematometrocolpos. The location of the septum may be in the upper vagina in 46%, in mid vagina in 40% and in lower vagina in only 16%.[5] The ureteric bud develops from the Wolffian duct which induces the metanephric blastema to form the kidney and if Wolffian duct is absent it leads to homolateral renal agenesis. Renal anomalies are known to be associated in 35% of mullerian duct anomalies in the form of agenesis, duplication, malrotation or ectopia.[7]

Classification of OHVIRA syndrome

OHVIRA syndrome has classification 1 and 2 according to the type of obstruction of hemivagina- complete or partial obstruction, because clinical details associated with each type are different.[1] Each of these classifications are further divided into two types: In classification 1.1, the affected hemivagina is completely obstructed and the uterus is completely isolated from the contralateral one. In classification 1.2, the hemivagina is completely obstructed and the cervix on same side is maldeveloped due to which menses from the uterus cannot drain through the atretic cervix. In classification 2.1, there is communication between the two hemivaginas, which makes the vaginal cavity behind the septum incompletely obstructed. In classification 2.2 the hemivagina is completely obstructed, and there is a small communication between the duplicated cervices (as in this case) and the menses from the uterus on the side of obstructed hemivagina outflow through this communication to the opposite side cervix and get drained. As communication between the two sides is small, the menses cannot be drained completely.

The clinical presentation of patient varies in all the above mentioned types. In classification I, the common presentation can be pelvic pain started after menarche, with findings of pelvic mass.[8] In classification 2, the presentation can be delayed (as in this case), because the obstructed hemivagina side can be drained through the communication between the obstructed and non obstructed side. Acute onset of abdominal pain, fever and vomiting can be the presenting symptoms due to bleeding from the fallopian tube.[9]

The imaging work up includes ultrasonography and Magnetic Resonance (MR) imaging. Ultrasonography can show uterovaginal duplication, hematocolpos or hematometrocolpos along with the absence of ipsilateral kidney[3] and as MR has multiplanar image acquisition capabilities, it can provide comprehensive data regarding the pelvic anatomy. MR can delineate the anomaly meticulously including the communication between the two cervices or vagina.

CONCLUSION

As the conclusion; HWWS is a uncommon urogenital anomaly, nonetheless it can be suspected in female patients who have absent kidney on one side and pelvic mass. Ultrasonography and MR imaging can well characterise the disease entity and vaginal septoplasty is the treatment of choice for obstructed hemivagina which lead to satisfactory long-term results.

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