CASE REPORT

Herlyn-Werner-Wunderlich Syndrome with Degenerating Fibroid– Uncommon Presentation of a Rare Anomaly

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Abstract:

Herlyn-Werner-Wunderlich Syndrome (HWWS) is a rare congenital anomaly of Mullerian and mesonephric ducts characterized by uterus didelphys with obstructed hemivagina and ipsilateral renal agenesis. It commonly presents after menarche with symptoms related to obstructive hemivagina. However in patients with significantly normal pubertal and menstrual history, the diagnosis of Mullerian Duct Anomaly (MDA) like HWWS is often missed. Here we report a case of 28 year old primigravida presenting with acute abdomen pain which was followed by spontaneous abortion. This is the first instance of HWWS coexisting with uterine fibroids with/without cystic degeneration based on our literature review. Early and accurate diagnosis for the correct treatment of this entity is also emphasized.

Keywords: Herlyn-Werner-Wunderlich syndrome, Mullerian Duct Anomaly, Spontaneous Abortion

Introduction:

HWWS is a rare congenital anomaly of Mullerian and mesonephric ducts characterised by a triad of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis. This entity is also known as Obstructed Hemivaginal and Ipsilateral Renal Anomaly (OHVIRA). The diagnosis is usually made after menarche by the symptoms related to the obstructed hemivagina such as pelvic pain, dysmenorrhea and palpable pelvic mass from associated hematocolpos or hematometra. The true incidence of this anomaly is unknown [1]. In this case report we wish to emphasize the need for early and accurate diagnosis for the correct treatment of this entity.

Case Report

A 28 year-old primigravida with gestational age of 13 weeks presented with complaints of severe lower abdomen pain for 10 days. There was no vaginal bleeding, discharge or fever. She attained menarche at the age of 13 years and had a significantly normal menstrual history. Abdominal ultrasound revealed bicornuate uterus, with healthy live fetus in the right horn and multiple fibroids in the left horn (Fig. 1). Largest of the fibroids measured about 9 x 4 cm. Ultrasound also revealed right renal agenesis with left renal compensatory hypertrophy (Fig. 1).

She was being managed conservatively for the pelvic pain. However, three days later she presented with spontaneous expulsion of dead fetus, placenta and membranes in-toto. Repeat USG revealed bicornuate uterus, hematocolpos and pelvic free fluid (Fig. 2).

MRI examination of the pelvis with screening of KUB region was performed for the detailed study of uterine anomaly. MRI revealed two separate uterine cavities, cervices and vaginas – suggestive

of uterus didelphys (Fig. 3). The right uterine horn cavity and myometrium were normal. The left uterine horn appeared enlarged and showed multiple fibroids of varying sizes, largest measuring 9.2 x 7 cm with cystic degeneration. Longitudinal septum was noted between the vaginal cavities with a small defect (~ 10 mm) in the superior portion. T2 hypointense content measuring 3 x 2.6 cm noted in the right vaginal cavity – possibly hematocolpos due to obstruction caused by transverse septum in right vaginal cavity. Left vaginal cavity appeared normal. Screening of upper abdomen showed right renal agenesis with compensatory left renal hypertrophy. Hence a diagnosis of uterus dideldphys with right hemivaginal transverse septum and right hematocolpos was made. As she also had absent right kidney, a final diagnosis of Herlyn-Werner-Wunderlich Syndrome, a rare MDA, was made. She was kept on short-term conservative treatment for the cystic degeneration of fibroid. Myomectomy and vaginal septectomy was advised prior to her next conception.

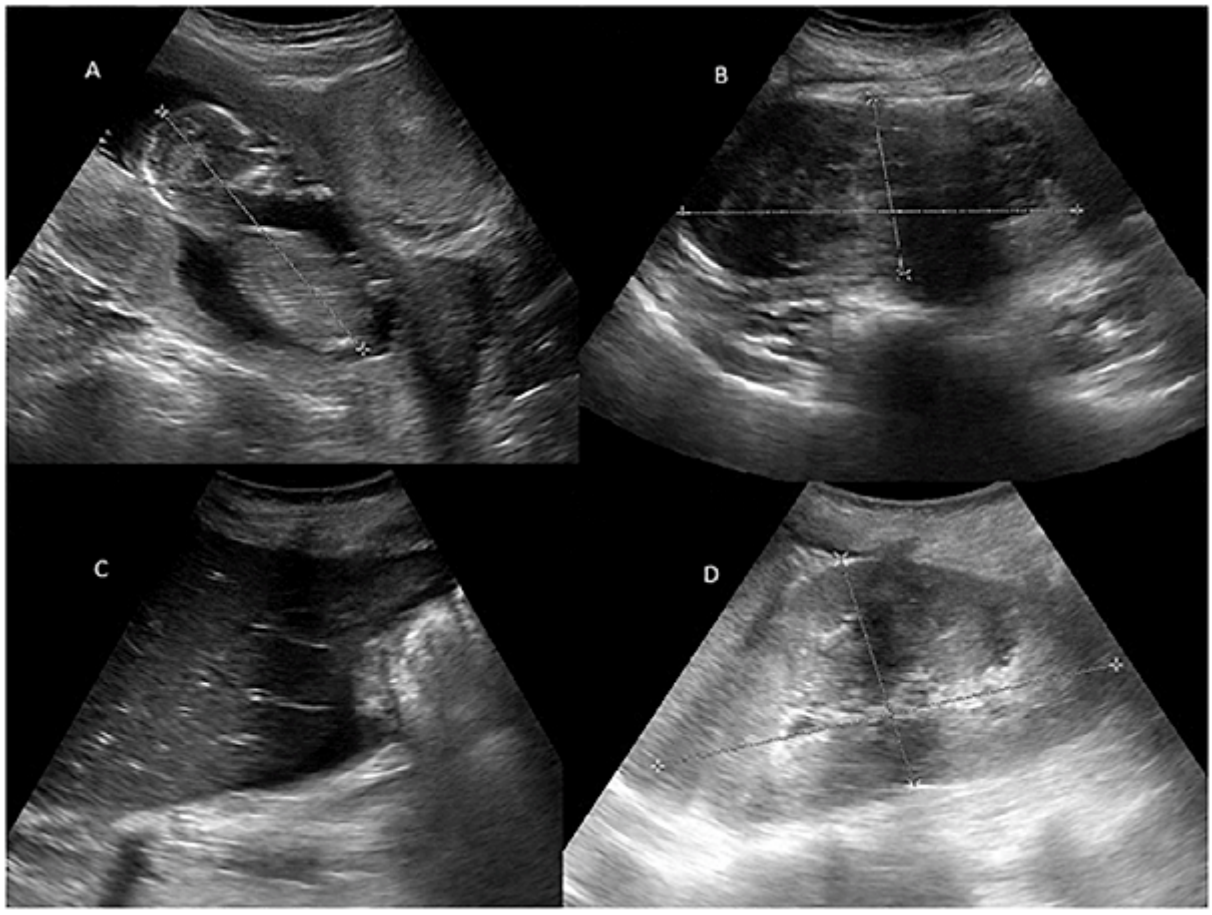

Fig. 1: Grey Scale Ultrasound Images shows (A) Live Fetus in the Right Horn of Uterus (B) Large Fibroid in the Left Horn of Uterus (C) Right Renal Agenesis (D) Compensatory Left Renal Hypertrophy

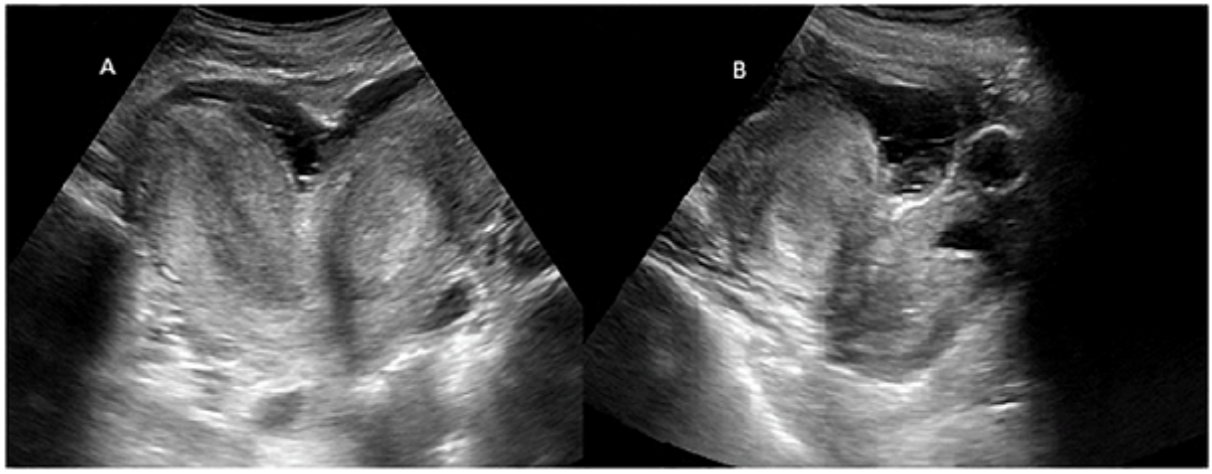

Fig. 2: Grey Scale Ultrasound shows (A) Right and Left Uterine Horns without Any Fetus; Minimal Fluid Anterior to the Uterine Horns (B) Right Sided Hematocolpos

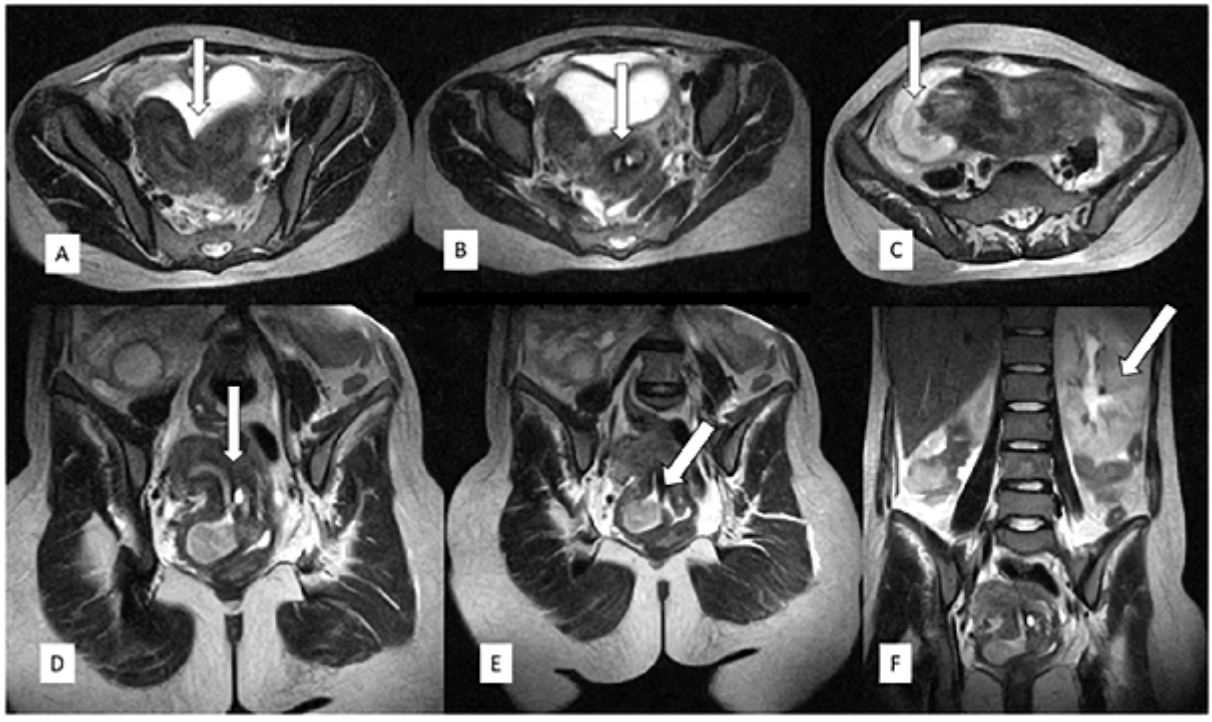

Fig. 3: T2 Weighted MRI (A and B) Axial image shows Two Separate Uterine Cavities and Cervical Canals with Ascites (C) Coronal Image shows Large Fibroid with Cystic Degeneration (D) Coronal Image shows Separate Cervix and Vagina with Obstructed Hemivagina and Hematocolpos on the Right Side (E) Coronal Image shows Small Defect Noted in the Superior Aspect of Longitudinal Vaginal Septum (F) Coronal Image shows Right Renal Agenesis with Compensatory Left Renal Hypertrophy (White Arrow)

Discussion:

The Wolffian (mesonephric) ducts and the Mullerian (paramesonephric) ducts are the two paired urogenital structures from which the internal genital organs and the lower urinary tract derive. The fallopian tubes, uterus and the upper 2/3rd of the vagina develop from the bilateral mullerian ducts. As a result of any interruption in the development of this embryological event different types of uterine anomalies can result in MDA such as uterine agenesis, hypoplasia, unicornuate, didelphys, bicornuate, arcuate and septate uterus. Embryological arrest at 8 weeks of gestation is the cause of uterine didelphys and renal agenesis [1, 2].

The American Society for Reproductive Medicine (ASRM) [3] has classified MDA under seven groups and vaginal anomalies into 3 groups. HWWS includes type III of MDA, type II vaginal anomaly and renal agenesis. The most common renal tract anomaly associated with MDA is renal agenesis with right sided prevalence [2, 4, 5].

The syndrome usually presents with acute or chronic pelvic pain following menarche caused by hematocolpos due to obstructive hemivagina. There may be delay in presentation due to spontaneous decompression of hematocolpos from a congenital defect in the vaginal septum in rare cases as reported here. Right sided obstructive hemivagina is more common as mentioned in the literature [7]. The potential complications of this syndrome are hematocolpos, hematometra, pyosalpinx or pelviperitonitis [7, 8]. Long-term complications include endometriosis from retrograde menstruation, pelvic adhesions, infertility or increased risk of abortions.

Our patient presented during pregnancy with significantly normal menstrual history probably due to the congenital defect in the vaginal septum facilitating spontaneous decompression of hematocolpos masking typical obstructive symptoms. As mentioned in previous case reports, successful pregnancy is achieved in 87% patients with HWWS, while 23% have risk of abortion [6]. Spontaneous expulsion of dead foetus at 14 weeks of gestation in this case could be likely due to multiple large fibroids with onset of cystic degeneration, hindering the pregnancy. HWWS with super added fibroids has never been reported and this report becomes the first instance of HWWS with fibroid uterus.

Vaginal septectomy or marsupialisation is the treatment of choice of obstructed hemivagina5. Metroplasty and elective caesarean section has to be considered in the patients with prior history of spontaneous abortion or premature delivery. Laparoscopy remains the gold standard diagnostic modality with additional advantages of therapeutic drainage of hematocolpos/ hematometra, vaginal septectomy and marsupialisation [7, 8].

Genitourinary anomalies have to be initially screened by ultrasound followed by MRI as it gives detailed elaboration of uterovaginal anatomy and there is no radiation exposure. Prompt antenatal detection of renal agenesis, neonatal screening for HWWS and preconceptional counseling with ultrasound screening in young females can reduce the morbidity resulting from HWWS.

References

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