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The Herlyn-Werner-Wunderlich Syndrome: A Rare Cause of Urinary Retention, Pelvic Pain and Dysmenorrhea in an Adolescent Girl

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Abstract: *The Herlyn-Werner-Wunderlich Syndrome also referred as OHVIRA syndrome is a rare congenital mullerian anomaly characterised by uterus didelphys (or sometimes septate uterus), hemi-obstructed vagina with ipsilateral renal agenesis. Although usual presentation is after menarche but few reports of pre-pubertal presentation in infants and a delayed presentation until during pregnancy have also been reported. The key to diagnosis in such cases is high index of suspicion and detailed work up in order to avoid late diagnosis of such rare presentations leading to complications that might compromise the fertility and affect the quality of life of an individual and add to morbidity. We are describing here a case of an adolescent girl who presented with history of urinary retention, spasmodic dysmenorrhea and lower abdominal pain after an year of menarche who was diagnosed in time and managed appropriately with an uncomplicated post-operative period.*

Keywords*: Herlyn Werner Wunderlich Syndrome, OHVIRA Syndrome, Hemi- obstructed vagina, uterus didelphys, mullerian duct anomalies, urinary retention, pelvic pain*

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I. Introduction

Herlyn-Werner-Wunderlich syndrome (HWWS), first described [1,2] between 1971–1976, is most likely the result to an insult in early pregnancy leading to embryonic arrest at around 7- 8 weeks of gestation, at which point the adjacent müllerian and mesonephric ducts are simultaneously affected. HWWS is a combination of type III mullerian anomaly according to the American Fertility Society with mesonephric duct anomaly with vaginal septum. 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 [3].

HWW syndrome is usually discovered at puberty with non-specific symptoms, like increasing pelvic pain, muco-purulent discharge dysmenorrhea and palpable mass due to the associated haematocolpos or hematometra, which result from retained, longstanding menstrual flow in the obstructed hemi-vagina. It rarely occurs with primary infertility in early adulthood when the vaginal septum is incomplete [4]. The possible early presentation of this syndrome should be suspected in all neonates (females) with renal agenesis confirmed postnatally or with prenatal diagnosis [5]. It is common, in fact, an error of evaluation with planning of removal of mass, that can damage patients in term of chance for a successful reproductive outcome.

MRI is considered as a gold standard for diagnosis and pre-operative planning of the Herlyn-Werner-Wunderlich syndrome. It evaluates uterine morphology, detects communication between vaginal and uterine lumen, characterises fluid contents, has the capability of multiplanar imaging but with no radiation, can detect associated renal agenesis and diagnose complications like endometriosis [6].

The rarity of this condition may contribute to diagnostic delay. Early diagnosis with appropriate surgical intervention with vaginoplasty and septostomy decreases the long-term morbidity of these women. The surgical outcome is excellent, and it is associated with a successful reproductive performance in the future.

II. Case Report

16-year-old unmarried female, referred to our out-patient Department from periphery with complaints of retention of urine for 15 days, spasmodic dysmenorrhoea since menarche and lower abdominal pain for past 4 months which has been aggravated for the past 3 days. She had attained menarche 1 year back and her cycles were regular with average flow. She had associated dysmenorrhoea, which was persistent throughout the cycle and later persisted even beyond it. Later it culminated to persistent lower abdominal pain for the last 4 months that was gradually progressive and had increased in intensity for past 3 days. There was no history of altered bowel habits, haematuria, per vaginal discharge, or similar family history. For these complaints' patient consulted a local practitioner, who advised an USG that reported "A large cystic mass in right adnexa of about

150 × 60 mms extending up to lumbar region with agenesis of right kidney and compensatory hypertrophy of left kidney ". IVP was also performed on the patient which revealed absent right kidney and compensatory hypertrophy of the left kidney. Patient was operated in local hospital according to per op notes ??hematometra of about 250 ml was drained from rudimentary horn and then the uterus was closed. As stated by patient her post op period was uneventful and there was subsequent relief in dysmenorrhea**.**

On Examination the patient is of average built, weight-42 Kg, height-152 cm, BMI-18.7. Her general examination found no significant abnormalities. Secondary sex growth was normal for her age. On per abdominal examination a just palpable mass was felt arising from pelvis. External gynaecology examination was also within normal limits, however on catheterizing a bulge in vagina on right side was noted. Further internal genital examination through the rectum found a cystic mass with a smooth surface in the right lateral region of the midline with proximal border and its origin was difficult to be determined. The uterus was normal sized and there was no solid mass palpable around the cervix. The rest of the systemic examination was within normal limits. Her Hb was 14.2 g/dl and thyroid profile, LFT, RFT, RBS were within normal limits. The ultrasonography (whole abdomen and pelvis) done in our institute revealed normal upper abdomen scan and presence of a "large right tubo-ovarian mass of about size [172 × 90 × 89] mms, agenesis of right kidney, enlarged uterus, left ovary was normal and there was no associated pelvic ascites. MRI of the patient was planned but could not be done due to non-affordability. A differential diagnosis of bicornuate uterus, unicornuate uterus with hematosalpinx in the right non-communicating horn and a right sided tubo-ovarian mass with normal uterus was made. Patient was planned for exploratory laparotomy after proper counselling and on opening the abdomen following findings were made:

- There were two hemi-uteri,
- Left side tube and ovary were found to be normal, left hemi-uterus was bulky 6*7 cm.
- Right side hemi-uterus was enlarged due to hematometra and fallopian tube was adhered to the normal ovary.
- adhesions were maximum on the site of previous drainage.

Adhesiolysis was done. On restoring the anatomy, it was noted that hematometra in its lower part showed an indentation which aroused the suspicion of haematocolpos along with hematometra (HERLYN-WERNER-WUNDERLICH SYNDROME) the possible diagnosis of uterine didelphys with hemi-obstruction of vagina was thought. This led to the decision of changing the patient to lithotomy position. On placing the patient in lithotomy position the unobstructed cervix was found to be flushed to fornices. On right side incision at bulge close to cervical opening given transverse septal removal was done which led to drainage of tarry blood. Post-operative period was uneventful. Patient was discharged on post-op day 7. Unfortunately, patient has not returned for follow up after discharge.

III. Discussion

The combination of uterus didelphys-class III of American Fertility Society –AFS- classification [7] and obstructed hemivagina was described the first time in 1922 [8], then in 70's Herlyn, Werner and Wunderlich reported other similar cases associated with renal anomalies [1, 2]. The occurrence of the HWWS seems to be 0.1-3.8%, and it is probably underestimated [9]. HWWS was included in the class U3B uterine anomaly, class C2 cervix anomaly, and class V3 vaginal anomaly according to the classification of ESHRE/ESGE [10].

The estimated overall prevalence of MDA is 2–3% of women. Uterus didelphys constitutes 11% of MDAs. Hypoplasia or agenesis of the uterus and proximal vagina constitute 5–10% of MDAs. Associated renal anomalies are present in approximately 43% [11,12]. About 75% of patients with didelphys uterus have a partial or complete vaginal septum which is commonly longitudinal in the Herlyn-Werner-Wunderlich syndrome, reflecting a disorder of lateral fusion between the inferior portions of two Mullerian ducts [13]. Studies of uterine organogenesis have implicated the Hox and Wnt genes as regulators of uterine morphology. However, specific gene mutations have not been identified as associated with most MDAs [6]. The exact etiopathogenesis of HWW syndrome is still not known. Wolffian ducts play an important role in the development of internal genital organs and give rise to kidneys [8]. They are inductor elements for adequate fusion of the Mullerian ducts. If one of the Wolffian ducts is absent, the ipsilateral kidney and ureter will fail to fuse in the midline. This process can be complete or incomplete. Uterus didelphys is formed if failure of fusion is complete. The Mullerian duct on the side lacking the Wolffian duct displaces itself laterally and cannot come into direct contact with the urogenital sinus in the centre with the resultant formation of a blind sac, imperforate or obstructed vagina. The distal third of vagina developing from the urogenital sinus is not affected. A didelphic uterus results due to embryologic arrest during the 8th week of gestation. This ultimately affects the Mullerian and metanephric duct [14].

HWWS be classified according to the complete or incomplete obstruction of the as follows [15]

Classification 1, Completely obstructed hemi-vagina

Classification 1a, With blind hemi-vagina
Classification1b, Cervicovaginal atresia without communicating uteri
Classification 2, Incompletely obstructed hemi-vagina
Classification 2a, Partial reabsorption of the vaginal septum.
Classification 2b, with communicating uteri

This new classification of HWWS can help to provide clinicians with earlier diagnosis and treatments to prevent secondary pelvic endometriosis and pelvic inflammation. (6). According to this classification our patient belongs to classification 1a.

When clinical signs and symptoms are present, the ultrasonography is usually the initial image exam performed, but it is highly dependent on the expertise of the operator. Magnetic resonance imaging is considered the gold standard for diagnosis and preoperative planning for the treatment of HWWS [16-18]. Treatment should be individualized depending on complaints, and the main goal is to relieve the obstruction by remodelling the vagina. Some vaginal septa can be easily displaced to the side, and others may be thick enough to cause symptoms, and therefore require surgical excision. Currently, the preferred surgical approach for patients with HWWS is the full excision and marsupialization of the vaginal septum. Errors in the surgical management can occur when the diagnosis is not suspected and laparotomy to explore and resect the intraabdominal mass is performed. In general, hemi-hysterectomy with or without salpingo-oophorectomy is rarely indicated and should be avoided to provide the best chances for a successful reproductive outcome [19]. Some authors have advocated for the use of laparoscopy at the time of vaginal septectomy to assess the exact uterine anatomy, yet its impact on definitive treatment is still unclear. Vaginal adenosis in the previously obstructed vagina must be considered in the post-operative period. No definitive guidelines exist at present, although some authors recommend yearly Papanicolaou tests and colposcopy.

IV. Conclusion

Herlyn-Werner-Wunderlich syndrome (HWWS) is a rare, combined Mullerian and Mesonephric duct anomaly characterised by the triad of uterus didelphys, obstructed hemi-vagina and ipsilateral renal agenesis. A high suspicion index is warranted for early recognition and prompt treatment in order to prevent future serious complications such as endometriosis, pelvic adhesions and infertility.

CONFLICT OF INTERESTS
None
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None

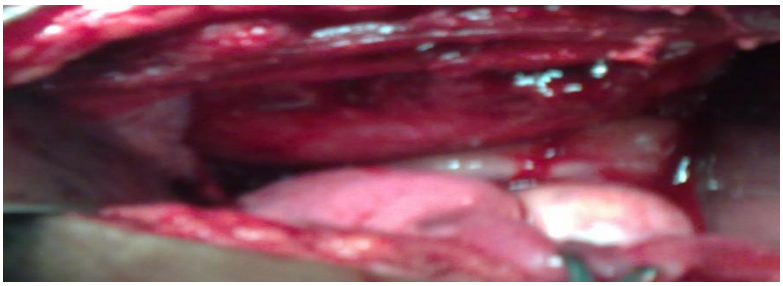

FIGURE 1: Figure shows the two hemi-uteri with normal left sided tube and ovary and adhesions aroud right tube.

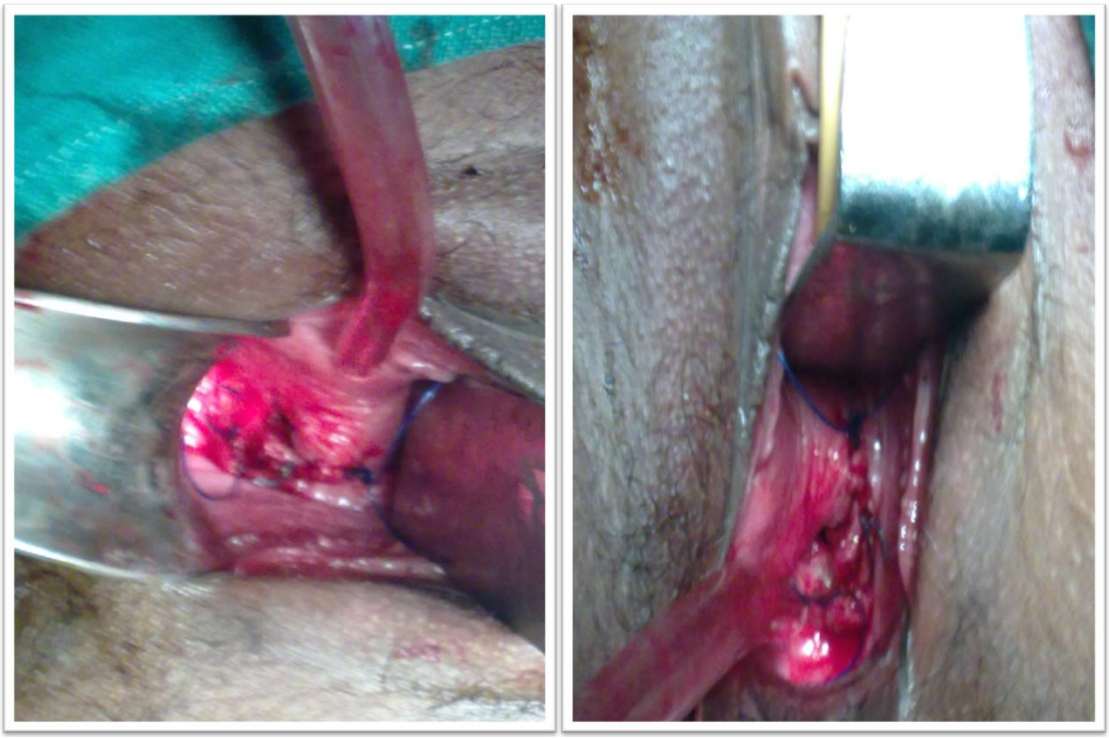

FIGURE 2: This figure shows the suction cannula pointing the right sided obstructed hemi-vagina after resection of the transverse septum and marsupilisation.

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