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Case Report

Case Report: Diagnosing and Treating a 11 Year Old Girl with Herlyn-Werner-Wunderlich Syndrome

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Abstract

Introduction: The Herlyn- Werner-Wunderlich (HWW) syndrome is a very rare congenital anomaly characterized by uterus didelphys, obstructed hemivagina and mesonephric duct anomalies. Most often the later manifests as renal agenesis [1]. Incidence of this condition is 0.1-3% [2].

The patient is usually asymptomatic in early childhood, onset of symptoms occur after menarche and common complaints are progressive dysmenorrhea, recurrent pain abdomen and palpable lump over lower abdomen. Patient can also present with urinary problems like recurrent UTI, vesico- ureteric reflux, increased risk of hypertension, compensatory hypertrophy of existing kidney, in severe cases Chronic Kidney Disease CKD and End Stage Renal Disease ESRD that occur as complication of unilateral solitary kidney.

Case Report: Case report presented here is of an 11 year old girl who presented with pain lower abdomen and pelvic region, fever, palpable tender mass over right lower abdomen and urinary complaints. Further evaluation of the case with biochemical parameters and radiological modalities like Ultrasound (USG) and Magnetic Resonance Imaging (MRI) confirmed the diagnosis as HWW syndrome complicated with CKD after which surgical management of the case was followed.

Conclusion: Duplication of the uterus and cervix with obstructed hemivagina and iplsilateral renal agenesis (presence of both) should prompt the clinicians to think in the direction of HWW syndrome. Early diagnosis and prompt treatment of the condition can prevent possible serious complication in future.

Keywords: Herlyn Werner Wunderlich Syndrome; Müllerian Duct; Wolffian Duct; Uterus Didelphys

Introduction

Herlyn-Werner-Wunderlich (HWW) syndrome also known as OHVIRA syndrome (Obsructed hemivagina and ipsilateral renal agenesis) is a rare congenital abnormality involving abnormal development of Mullerianand Wolffian duct [3]. It is characterized by a triad of uterus didelphys, blind hemivagina and ipsilateral renal agenesis. Etiology of HWW syndrome involve embryological arrest in normal development of mesonephric or Wolffian duct which forms the renal system resulting in renal agenesisand paramesonephros or Mullerian duct which fail to fuse with its counterpart from opposite side. Thisprocess is affected by various genetic factors and signaling molecules like EMX 2, HOXA13, PAX 2 and Wnt which help in normal growth and differentiation of uro-genital system. Also recently LEm1 mutation is found in some cases with abnormal fallopian tubes, uterine aplasia and infertility [4].

Usually the symptoms appear after menarche depending upon the type of HWW syndrome. Patient usually present with progressive dysmenorrhea, constant dull aching pain lower abdomen which may beassociated with palpable mass. Symptoms are usually non specific and are mostly due to hematometra, hematosalpinx and hematocolpos [5]. Unusual presentation are renal or vaginal hemorrhage, endometriosis, infertility and benign or malignant tumors of genitor-urinary tract [6]. Sometimes urinarysymptoms like dysuria, frequency, burning micturition associated with recurrent UTI are the only presentation in pre pubertal female.

The final diagnosis is usually evaluation after radiological evalution with ultrasonography or MRI [7]. Earlier diagnosis and prompt treatment prevents future morbidities like endometriosis, infertility, recurrent UTI, chronic renal disease, benign and malignant tumours of genito-urinary tract.

Case Report

An 11 year old girl presented in ASIAN gynae OPD with severe dysmenorrhea with fever, dysuria and frequency of micturition since 4 days. Her menarche was 10 month prior to this episode. Previousmenstrual cycles were irregular, lasting for 2-3 days and associated with dysmenorrheal gradually increased in intensity with each successive period.

She is a known case of chronic kidney disease diagnosed since 4 year of age and was under survillenceand management for the same. Previous scans revealed absence of right kidney (right renal agenesis)with grade 4 vesico ureteric reflux in left kidney and bicornuate uterus. Patient had past history of dull aching pain lower abdomen and dysuria with elevated creatinine for which she was hospitalized 1 month back to this episode. Patient was born to a consanguineous couple via cesarean section and in- utero history of oligomenorrhea was there.

Evaluation of the case was started with multidisciplinary approach involving paediatrician, urologists, nephrologists, and paediatric surgeon. Lab tests revealed leukocytosis, moderate anemia, deranged renal profile (increased urea-86mg/dL, creatinine-3.8mg/dL) and hyperkalemia (5.4meq/L) While patient was under evaluation her creatinine rose to 9.2. USG lowerabdomen and pelvis revealed Mullerian duct anomaly- bicornuate bicollis uterus with loculated collection in right endometrial cavity and right lower cervical canal with possibility of right sided hematocolpos, hematometra and hematosalpinx. MRI pelvis confirmed the USG finding of bicornute bicollis uterus with loculated hematometra in the right uterine cavity and right hemicervix with nonvisualization of right kidney and left renal grade 4 VUR.

Microarray profile was normal however in whole exome sequencing heterozygous variant of uncertainsignificance was identified in GREB1L gene which is a possible candidate gene for Mayer-Rokitansky-Kuster-Hauser syndrome (MRKH).

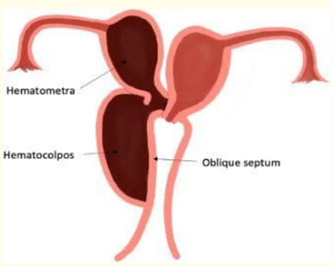

Figure 1: Mayer-Rokitansky-Kuster-Hauser syndrome.

On the basis of clinical, lab and radiological findings the patient was diagnosed with HWW syndrome complicated with CKD and left sided grade 4 VUR and right sided hematometra and hematocolpos. Withall experts opinion decision to examine under anaesthesia with hysteroscopy and laparoscopy or laparotomy was taken. Preoperatively she received antibiotics and repeated dialysis to correct the renalparameters.

Intra operative hysteroscopy revealed normal vaginal walls with a bulge towards right side. A small cervix was observed towards left end of the vagina. Hysteroscope was introduced through the cervical canal of left side to reach the left half of uterine cavity. Uterine cavity was lined with normal endometrium, uterine orifice of the fallopian tube noted in the left side. It was followed by laparotomy with urologist and paediatric surgeon. Right sided hematosalpinx and distended right horn with hematometra was noted. Dark brown chocolate color blood was aspirated from right side of the uterus.Hemihysterectomy with removal of right sided non- communicating rudimentary horn along with right fallopian tube done. Left sided uterine contour regular, left fallopian tube and ovary was seen normally.Left ureteric reimplantation with D-J stunting was done by urology team for vesicoureteric reflux.

Postop period remain uneventful with follow up dialysis and antibiotics. Patient discharged on post opday 5. Follow up biochemistry after 1 week suggested improving renal status with creatinine coming down to 3.2. The patient had pain free menstrual cycles after 1.5 months. Follow up with nephrology team continued.

Discussion

HWW syndrome or OHVIRA Syndrome is a rare congenital abnormality involving genito-urinary system. Incidence of this condition is 0.1-3% [2]. 1st reported by Purslow in 1922, the triad of uterus didelphys, obstructed hemivagina and ipsilateral renal agenesis was fully described by Herlyn and Werner in 1971. In 1976 Wunderlich described a case of bicornuate uterus with ipsilateral blind hemivagina with isolated hematoclpos and same side renal agenesis. The exact pathogenesis is not completely understood but it is associated with improper fusion of the Mullerian duct s during 8th weekof gestation and incomplete development of mesonephric duct [10]. Due to embryologic arrest of the growing mesonephric duct ; Mullerian duct of ipsilateral side is displaced laterally and can not fuse withits contralateral counterpart resulting in didelphic uterus. Also the displaced Mullerian duct can not come in contact with urogenital sinus centrally forming a blind sac which leads to an obstructed or imperforated hemivagina [11].

In 2015 Zhu teal. Proposed a detailed classification of HWW syndrome on the basis of observation of patients in Peking union Medical College Hospital [7]. In type 1, with blind hemivagina ; hemivagina is completely obstructed, uterus behind the septum is completely isolated from the contralateral uterus, and no communication is present between the duplicated uterus and vagina. In Type 2 there is communication between 2 vaginas due to partial resorption of the vaginal septum, also the uterus behind the septum is completely isolated from the contralateral uterus. Menstrual blood can outflow through the small communication but the drainage is impeded. Type 3 is with communication between2 uteri: in this classification hemivagina is completely obstructed, and a small communication exist between 2 duplicated cervices, menstrual blood can outflow through the small perforation in uterus but again the flow is impended.

A literature review in Milan University showed a two fold prevalence of right sided anomaly (91 out of 138 cases) [8]. In our case, the anomaly was also confirmed on right side. As per literature, the

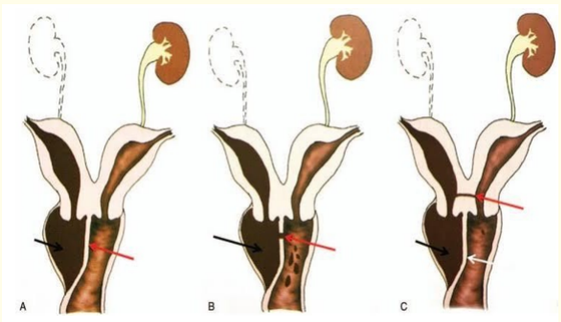

Figure 2

onset symptoms can start at 5 year of age [9]. Reports of patients aged between 5 months and 4 years having urinary retention or masses in the lower abdomen that need laparoscopic treatment are present [9].

Additionally, prenatal diagnosis of the absence of one kidney and then post delivery confirmation of genital anomalies have been reported [9]. In our case patient has in-utero history of oligohydramnioswhich could be as a result of renal anomaly.

Successful surgical intervention following early diagnosis and treatment improves quality of life andprevent development of endometriosis.

Conclusion

Presence of unilateral renal agenesis and uterus didelphys should prompt the gynaecologist to use appropriate imaging techniques for correct interpretation, faster diagnosis and treatment of rare HWWsyndrome in order to prevent future complications. However considering unilateral renal agenesis multidisciplinary approach is best for ensuring more insight in evaluation and treatment of this condition.

Bibliography

1. Mandava Anitha., *et al*. "OHVIRA syndrome". *Journal of Paediatrics and Adolescent Gynaecology* (2012).

2. Aveiro Ana Cristina., *et al*. BMJ Case report (2011).

3. Burgis J. "Obstructive mullerian anomalies: case report, diagnosis and management". *American Journal of Obstetrics and Gynecology* (2001).

4. Wilson D and Bordoni B. Embriology, Mullerian ducts (Para mesonephric ducts): Stat Pearls: Orlando, FL, USA, (2021).

5. Zhu L. "New classification of HWWS". *Chinese Medical Journal (Engl)* (2015).

6. Jindal G., *et al*. "Uterus didelphys with unilateral obstructed hemivagina, hematocolpos, hematosalpinx with unilateral renal agenesis". *Journal of Reproduction Science* (2009).

7. Zhu L., *et al*. "New classification of Herlyn -Werner-Wunderlich syndrome". *Chinese Medical Journal (Engl)* (2015).

8. Vercellini P., *et al*. "Asymmetrical lateral distribution of obstructed hemivagina and renal agenesis in women with uterus didelphys: institutional case series and a systematic literature review". *Fertility and Sterility* 87 (2007).

9. Angotti R., *et al*. "Herlyn- Werner- Wunderlich syndrome: an early onset case report and review of literature". *International Journal of Surgery Case Reports* (2015).

10. Rechberger T., *et al*. "Congenital anomalies of the female reproductive tract- diagnosis and management". *Cureus* (2024).

11. Guifeng Jia., *et al*. "A case report on Herlyn- Werner- Wunderlich syndrome with spontaneous abortion". *Medicine (Baltimore)* 97.36 (2018): e12004.