University Journal of Surgery and Surgical Specialities

ISSN 2455-2860 **2020, Vol. 6(1)**

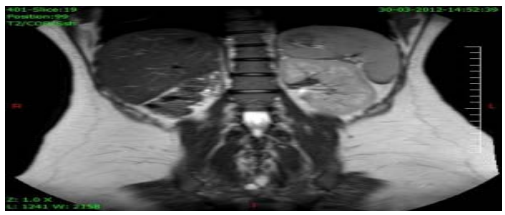
Herlyn-Werner-Wunderlich syndrome

JEYANTHI S
Department of Obstetrics and Gynaecology, CHRISTIAN MEDICAL COLLEGE

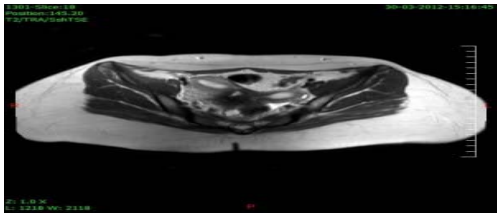
Abstract :
Herlyn-Werner-Wunderlich (HWW) syndrome is a rare congenital anomaly of the urogenital tract involving Mllerian ducts and Wolffian structures. It is characterized by triad of didelphys uterus, obstructed hemivagina and ipsilateral renal agenesis. The correct diagnosis can be difficult due to presence of non specific symptoms and absence of specific findings upon physical examination which can delay the diagnosis. The treatment consists of vaginal septum excision and marsupialisation to maintain patency of vagina. Herein we report a case of Herlyn-Werner-Wunderlich syndrome with pyocolpos.

Keyword :
Herlyn-Werner-Wunderlich syndrome, HWW syndrome, didelphys uterus, ipsilateral renal agenesis, obstructed hemivagina

Mrs. X, 27 Years old, married for 6 months presented with complaints of dyspareunia since her marriage and foul smelling vaginal discharge for 12 days duration. She attained menarche at the age of 14 years. She had regular cycles with spasmodic dysmenorrhoea. On examination her height was 160 cm, weight was 58 kgs and Body mass index was 22.7. Breast was Tanners stage 4. General examination was unremarkable. Abdomen examination revealed tenderness on deep palpation in the lower abdomen .There was no mass palpable per abdomen .External genitalia appeared normal. Speculum examination revealed a bulge on the right side of vagina, and cervix was seen on the left side. Per Vaginal examination, uterus was anteverted, normal size, mobile, a bulge was felt on the right hemivagina, and cervix was felt through the left side of vagina. **MRI of Abdomen and Pelvis** ; showed didelphys uterus with two cervices, obstructed right hemivagina probably due to transverse vaginal septum at upper ¾ of vagina. Left hemi vagina was normal and separately seen. There was associated Right renal agenesis with compensatory hypertrophy of the left kidney. These features were suggestive of **Herlyn-Werner-Wunderlich syndrome.**



Picture 1 ; MRI imaging of T2 coronal view showed right renal agenesis with compensatory left renalhypertrophy



Picture 2;MRI imaging of T2 transverse view showed didelphys uterus with obstructed right hemivagina.

She underwent Excision **and marsupialisation of the of vaginal septum** following which the cervix on the right side could be felt. **Intra-operatively** she was found to have right hemi vagina almost coming upto the introitus which had about 20 ml of thick yellowish discharge. Left cervix was felt separately. Post operative period was uneventful. She was relieved of the symptoms after surgical excision of the vaginal septum and marsupialisation. There after six months later she underwent Diagnostic **hysterolaparoscopy** as a part of infertility workup .On Laparoscopy she had didelphys Uterus, both tubes and ovaries were normal with minimal endometriotic deposits seen in Pouch of Douglas. There was bilateral free tubal spill seen on chromopertubation. Hysteroscopy showed two separate cervix and two uterine cavities with two ostia. She conceived spontaneously a month after the procedure and underwent **Elective Lower segment caesarean section** at 39 weeks for

Extended Breech presentation on 14.8.2013 .Both mother and baby are doing well.

DISCUSSION

Müllerian duct anomalies (MDAs) are congenital anatomic abnormalities of the female genital tract that arise from nondevelopment or nonfusion of the müllerian ducts or failed resorption of the uterine septum .The first stage of müllerian duct **development** begins at approximately 6 weeks of gestational age and mullerian duct grow caudally cross over the wolffian ducts to meet at the midline. The three phases of fusion, resorption, and vaginal induction proceed from the 9th to the 22nd gestational week. Frequent association of urinary anomalies are due to close association with Müllerian duct development .The uterus, fallopian tubes, and upper two-thirds of the vagina originate from paired müllerian ducts, whereas the lower third of the vagina arises separately from the urogenital sinus. Uterus didelphys with obstructed hemivagina and ipsilateral renal agenesis is a rare congenital anomaly referred to as **Herlyn-Werner-Wunderlich syndrome (HWW).** This syndrome was described for the first time in 1922.The incidence of an obstructive Mullerian anomaly is 0.1~3.8%. (1) .The incidence of didelphys uterus related to HWW, is approximately 1/2,000 to 1/28,000, and it is accompanied by unilateral renal agenesis in 43% of cases. (2)The exact cause, pathogenesis, and embryologic origin of HWW syndrome are uncertain but the diagnosis and treatment of early stage can relieve acute symptoms and preserve normal fertility. (3,4) Mullerian duct anomalies are clinically important because they are associated with infertility, menstrual disorders, and obstetric complications. They are also associated with an increased incidence of endometriosis due to obstructed uterine drainage. In **Uterus didelphys**, individual horns are fully developed, normal in size with two cervices. Each hemiuteri is associated with one fallopian tube. Ovarian malposition may also be present. A longitudinal or transverse vaginal septum may be noted. The vagina may be single or double. Duplicated vagina is more likely in a didelphic uterus. Ipsilateral renal agenesis with partial vaginal septum on the same side and didelphys uterus is explained by embryologic arrest at the 8th week of gestation affecting simultaneously the mullerian and metanephric ducts. Incidence of renal anomalies in Didelphys uterus was found to be 20%. (5). Vaginal obstruction develops because the paramesonephric ducts cannot meet the urogenital sinus centrally. Without a mesonephric duct, the ipsilateral kidney and ureter cannot develop, consequently HWW syndrome occurs.

The routine clinical presentation of this syndrome is abdominal pain that starts right after menarche and is caused by hematocolpos. Patient may present with Primary infertility and dyspareunia. The complications of this syndrome are pyohematocolpos, pyosalpinx, or pelvic peritonitis, long-term complications are endometriosis, pelvic adhesions and increased risk of abortion or infertility . The **role of imaging** is to help detect, diagnose and distinguish surgically correctable forms of mullerian malformations . Three-dimensional Ultrasound (3D) has higher sensitivity and specificity to evaluate malformations. MRI is more sensitive in detecting the uterine contour, the shape of the intrauterine cavity, the character of the septum and the continuity with each vaginal (obstructed and non-obstructed) lumen, and fluid content nature .It also detects endometriosis ,pelvic inflammation and adhesions (6,7). Complete excision and marsupialisation of the vaginal septum is the preferred approach because there may be recurrence of the obstruction and pyometra after a simple incision alone . After the septum is excised, laparoscopy can be performed for potential treatment of associated endometriosis, adhesions, or both. Successful pregnancy with the corrective surgery is seen in 87% of cases, however 23% of cases have abortion,15% of cases have preterm birth ,62% of cases have full term pregnancy and uncomplicated deliveries (8).

Conclusion

Mullerian duct abnormalities should be considered as a differential diagnosis in young girls presenting with acute abdominal symptoms and severe dysmenorrhoea shortly after menarche. When a high suspicion of mullerian duct abnormalities exists , MRI is the best imaging modality to diagnose. With a diagnosis of HWW syndrome the imaging should include upper abdomen to check for the presence of ipsilateral kidney. If renal agenesis is diagnosed renal function evaluation should be done. Complete excision and marsupialisation of vaginal septum will improve the overall success rate.

References

1.Burgis J. Obstructive Müllerian anomalies: case report, diagnosis, and management. Am J Obstet Gynecol 2001;185:338-44.

2. Alan JW, Louis RK: Campbell-Walsh Urology. Philadelphia: Saunders;93270-6.5.Stampe Sorensen S. *Estimated prevalence of mullerian duct anomalies. Acta Obstet Gynecol Scand 1988*; *67*:*441*- 445.

3. Orazi C, Lucchetti MC, Schingo PM, Marchetti P, Ferro F.Herlyn-Werner-Wunderlich syndrome: uterus didelphys, blind hemivagina and ipsilateral renal agenesis. Sonographic and MR findings in 11 cases. Pediatr Radiol 2007; 37:657-65.

4. Zurawin RK, Dietrich JE, Heard MJ, Edwards CL. Didelphic uterus and obstructed hemivagina with renal agenesis: case report and review of the literature. J Pediatr Adolesc Gynecol 2004; 17:137-41.

5. Vercellini P, Daguati R, Somigliana E, Viganò P, Lanzani A, Fedele L. Asymmetric lateral distribution of obstructed hemivagina and renal agenesis in women with uterus didelphys: institutional case series and a systematic literature review. Fertil Steril 2007;87:719-24.

6. Park Noh Hyuck, Park Hee Jin, Park Chan Sup, Park Sung Il: Herlyn-Werner- Wunderlich Syndrome with Unilateral Hemivaginal Obstruction, Ipsilateral Renal Agenesis and Contralateral Renal Thin GBM Disease: A Case Report with Radiological Follow Up. J Korean Soc Radiol 2010, 62:383-388.

7. Mintz MC, Thickman DI, Gussman D et al (1987) MR evaluation of uterine anomalies. AJR Am J Roentgenol 148:287–290

8. Candiani GB, Fedele L, Candiani M. Double uterus, blind hemivagina, and ipsilateral renal agenesis: 36 cases and longterm follow-up. Obstet Gynecol 1997;90:26-32.

An Initiative of The Tamil Nadu Dr. M.G.R. Medical University
University Journal of Surgery and Surgical Specialities

An Initiative of The Tamil Nadu Dr. M.G.R. Medical University
University Journal of Surgery and Surgical Specialities