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CASE REPORT

Herlyn–Werner–Wunderlich Syndrome: Premenarche

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ABSTRACT

Aim: To increase awareness about this rare condition in order to aid early diagnosis and effective treatment which can eventually prevent complications and infertility issues in the future.

Materials and methods: A case of a 10 year old pre-menarchal girl, who presented with this syndrome, was managed surgically. After this, a literature search was done and similar case reports were reviewed.

Results: The Herlyn-Werner-Wunderlich syndrome (HWW) is a rare Müllerian anomaly with an incidence of 0.1-3.8%. It is characterised by uterus didelphys with obstructed hemivagina and ipsilateral renal agenesis. This condition can be treated with surgery. Since delay in diagnosis may lead to complications, early diagnosis and treatment is beneficial. When Müllerian anomalies are encountered, a screening should also be made for congenital renal abnormalities and vice versa. This condition usually presents postmenarche with abdominal pain due to hematocolpos or dysmenorrhoea. However, there are rare occasions when this condition can present even before menarche.

Conclusion: The clinical presentation of HWW syndrome requires a high index of clinical suspicion. Though computed tomography or ultrasound can be used to confirm the diagnosis, magnetic resonance imaging is considered to be the preferred imaging modality. Early diagnosis and surgical management is important in order to prevent complications like retrograde tubal reflux, leading to endometriosis and infertility.

Keywords: Dysmenorrhea, Hematocolpos, Müllerian anomalies, Premenarchal pain abdomen, Renal agenesis, Uterine didelphys.

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INTRODUCTION

Herlyn–Werner–Wunderlich (HWW) syndrome is also known as obstructed hemivagina ipsilateral renal agenesis (OHVIRA) syndrome, which stands for the components of the syndrome – OHVIRA and uterine didelphys. This syndrome is most commonly seen in postmenarchal adolescent girls. To the best of our knowledge, there have been only seven case reports of this syndrome presenting in premenarche girls.[1] Hence, we present our case as the eighth case report of a girl who was treated for this syndrome at the age of 2 and again at the age of 11 before her menarche.

CASE REPORT

An 11-year-old girl presented to the outpatient department with purulent discharge per vaginum since a month, which did not subside with antibiotics. She had not yet attained menarche. She also had a similar episode at 2 years of age when she had been diagnosed with pyometra, hydroureter, and hydronephrosis with absent right kidney. She had undergone cystoscopy, vaginoscopy, retrograde pyelogram, and laparotomy with drainage of pyometra and dilatation of cervical canal. She remained asymptomatic until current presentation.

On examination, her general condition was good. Her breast development was Tanner II. However, she had no axillary or pubic hair. No abdominal mass or tenderness was noted. Her external genitalia were healthy and copious purulent vaginal discharge was noted.

Ultrasonography (USG) revealed a pyocolpos, pyometra, pyosalpinx, right renal agenesis, with compensatory hypertrophy of left kidney, uterus didelphys, and completely septate vagina up to lower one-third. She had been started on antibiotics; however, pus culture was reported as negative (Fig. 2).

She underwent examination under anesthesia, and purulent discharge was noted. A smooth bulge was seen on the medial wall of left hemivagina caused by right hematohemicolpos. An opening was created in the septum between the two hemivaginae, which revealed the right cervix. Resection of the lower part of the septum with drainage of the pus was done. Her condition improved and she was symptom free (Fig. 3).

A year later she attained menarche and currently has regular menstrual cycles with no dysmenorrhea. A follow-up ultrasound revealed two patent vaginae and two cervices with a didelphys uterus.

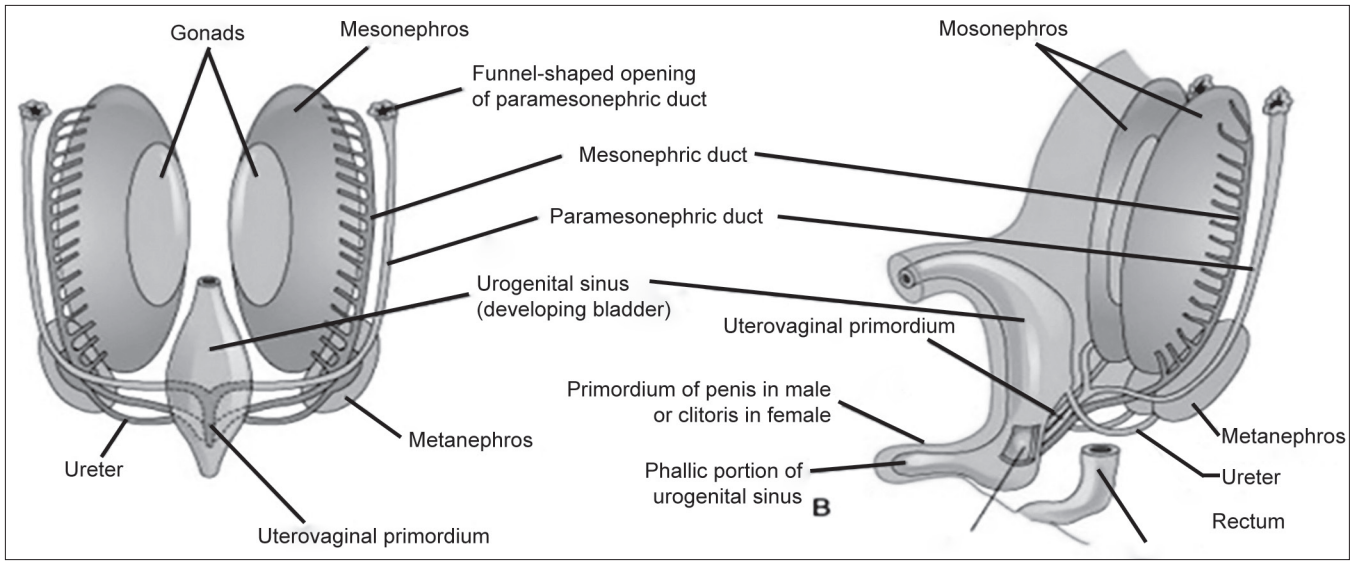

Fig. 1: Development of the urogenital tract

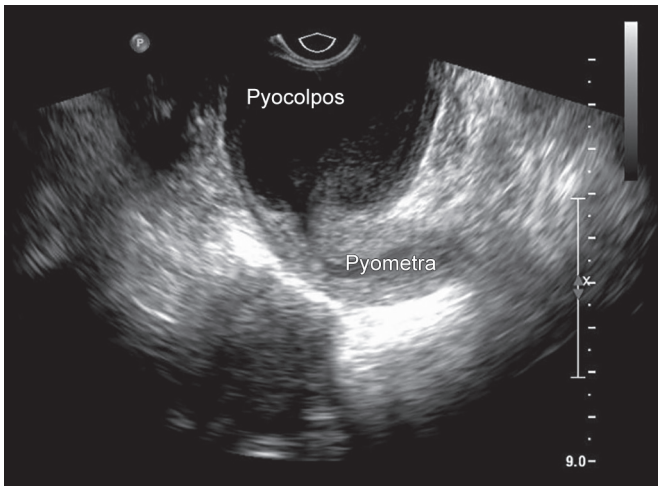

Fig. 2: Preoperative image showing pyocolpos and pyometra

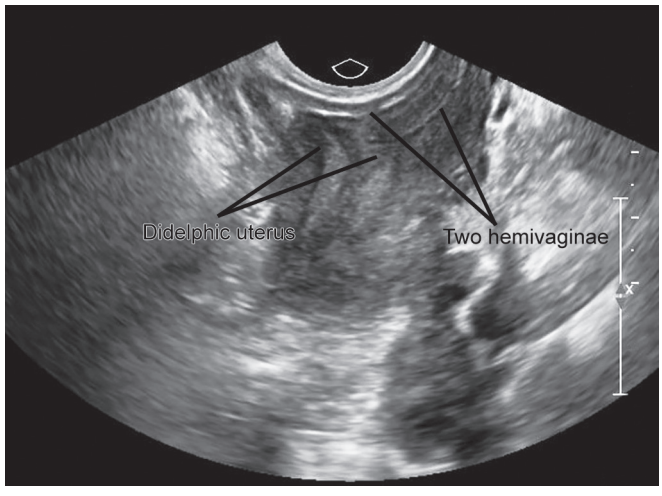

Fig. 3: Postoperative image showing didelphic uterus with two hemivaginae

DISCUSSION

The HWW syndrome is a rare complex of structural abnormalities of the mesonephric and Müllerian ducts. The diagnosis is difficult due to lack of specific findings on clinical examination. There are only a few hundred cases reported since its first description in 1922. The incidence of this syndrome is 0.1 to 3.5%.[2] Müllerian anomalies are found in 1 to 3% of the general population and 25% in recurrent pregnancy loss (RPL) and infertility patients.[3] Uterine didelphys is found in 11% of Müllerian anomalies and is associated with renal abnormalities in 81% of patients.[4] Herlyn–Werner syndrome was coined in 1971 when an association of renal agenesis with obstructed hemivagina was made. An association of a didelphys uterus was added by Wunderlich in 1976. There have been various classifications for Müllerian anomalies, namely Buttram and Gibbons in 1979, AFS in 1988, and the current ESHRE/ESGE in 2013. The HWW syndrome is described as class III – a failure of lateral and vertical fusion.

Mesonephros is responsible for the development and positioning of paired paramesonephric duct in close proximity (Fig. 1). At around 9 weeks gestation, the paramesonephric duct is positioned in such a manner that it is lateral to mesonephric duct in first part, crosses it anteriorly, and lies medial to it in converging portion. Due to failed positioning of paired paramesonephric duct, the two hemiuteri and hemicervices fail to unite, resulting in uterus didelphys. In OHVIRA syndrome, developmental arrest of ipsilateral mesonephric duct results in failure of distal hemivagina to develop, thereby resulting in obstructed hemivagina. The OHVIRA syndrome consists of uterus didelphys, unilateral low vaginal obstruction, and ipsilateral renal agenesis, all three components being secondary to mesonephric duct-induced Müllerian anomalies.[5]

This condition commonly presents postmenarche with abdominal pain due to hematocolpos or dysmenorrhea. There are only seven cases of this syndrome in prepubertal girls reported so far. These girls present with abdominal mass, urinary retention, or prenatal scan showing absence of one kidney.[6]

The diagnosis is usually made with clinical examination and confirmed with imaging. The investigations that are useful are two-dimensional or three-dimensional USG, sonohysterography and hysterosalpingography.[7] Magnetic resonance imaging (MRI) is a good tool for evaluation.[8] Hysterolaparoscopy has been considered the gold standard in the management of HWW syndrome.[9]

Delay in diagnosis can lead to pelvic adhesions, endometriosis, pyometra, pyocolpos, pyosalpinx, chronic pelvic pain, and future fertility issues. Adenocarcinoma of the obstructed uterus and clear cell carcinoma of the vagina have also been observed.[10]

SURGICAL INTERVENTION

Incision of the longitudinal vaginal septum is indicated for an obstructed hemivagina with hematocolpos or dyspareunia.[11] Metroplasty is controversial.[12] Strassman metroplasty is considered in those with RPL or preterm delivery. Studies do not support repair of uterus didelphys to improve pregnancy outcome. Successful pregnancy has been reported in 57% cases without metroplasty. Heinonen[13] and others improved fetal survival rate from 57 to 92% by cervical cerclage. Laparoscopic hemihysterectomy and trachelectomy is a minimally invasive surgical option for patients with HWW syndrome.[14]

There has been a difference in opinion about this option. Altchek and Paciuc[15] have reported pregnancy occurring twice in a previously obstructed didelphys uterus after surgical correction where the obstructed uterus was preserved. Lee and Weber[16] have reported a case of successfully achieving vaginal patency with the tracheobronchial stent in a patient with OHIVRA who had previously reobstructed twice.

FERTILITY OUTCOMES

Of all the Müllerian anomalies, arcuate and didelphys uterus have the best obstetric outcome with a term birth of 56%. Literature mentions a successful pregnancy in 87% women after surgical treatment.[17] However, there is an increased risk of miscarriage (32%), preterm birth (28%), breech and cesarean delivery. Pregnancies occur in both the affected and unaffected uterus.[18]

CONCLUSION

The clinical presentation of HWW syndrome requires a high index of clinical suspicion. Unilateral renal agenesis in prenatal scan should prompt imaging to rule out Müllerian anomalies.[19] Though computed tomography or ultrasound can be used to confirm the diagnosis, MRI is considered pivotal in diagnosis. Surgical excision or complete division of the septum is the treatment of choice. Since delay in diagnosis may lead to complications, early diagnosis and treatment is beneficial.

CLINICAL SIGNIFICANCE

The aim of this article is to focus on the importance of a high level of suspicion in order to diagnose HWW syndrome. A search for Müllerian anomalies is required when renal agenesis is detected. This is vital for early diagnosis and treatment. The ultimate goal is prevention of complications like endometriosis and preservation of coital and reproductive abilities.

REFERENCES

1. Angotti R, Molinaro F, Bulotta AL, Bindi E, Cerchia E, Sica M, Messina M. Herlyn–Werner–Wunderlich syndrome: an "early" onset case report and review of literature. Int J Surg Case Rep 2015 Apr;11:59-63.
2. Resetkova N, Christianson M, Kolp, L. Uterine didelphys with obstructed hemivagina and ipsilateral renal agenesis with hydronephrosis. Fertil Steril 2012 Mar;97(3):S30-S31.
3. Acien P, Incidence of Müllerian defects in fertile and infertile women. Hum Reprod 1997 Jul;12(7):1372-1376.
4. Li S, Qayyum A, Coakley FV, Hricak H. Association of renal agenesis and mullerian duct anomalies. J Comput Assist Tomogr 2000 Nov-Dec;24(6):829-834.
5. Bajaj SK, Misra R, Thukral BB, Gupta R. OHVIRA: uterus didelphys, blind hemivagina and ipsilateral renal agenesis: advantage MRI. J Hum Reprod Sci 2012 Jan-Apr;5(1): 67-70.
6. Vilhena C, Metello JL, Casal E, Romão F. Herlyn-Werner-Wunderlich syndrome: 3D ultrasonographic diagnosis in premenarche. Middle East Fertil Soc J 2014 Sep;19(3):229-232.
7. Valenzano MM, Mistrangelo E, Lijoi D, Fortunato T, Lantieri PB, Risso D, Costantini S, Ragni N. Transvaginal sonohysterographic evaluation of uterine malformations. Eur J Obstet Gynecol Reprod Biol 2006 Feb;124(2):246-249.
8. Sahu M, Naik SS. Pregnancy with uterine didelphys with obstructed hemivagina and pyocolpus. J Clin Diagn Res 2015 Feb;9(2):QJ01-QJ02.
9. Philbois O, Guye E, Richard O. Role of laparoscopy in vaginal malformation. An experience in 22 children. Surg Endosc 2004 Feb;18(1):87-91.
10. Watanabe Y, Etoh T, Nakai H. Adenocarcinoma of the lower female genital tract in patients with Herlyn-Werner-Wunderlich syndrome. Am J Obstet Gynecol 2012 Dec;207(6): e5-e6.
11. Propst AM, Hill JA 3rd. Anatomic factors associated with recurrent pregnancy loss. Semin Reprod Med 2000 Feb;18(4):341-350.
12. Iverson RE, DeCherney AH, Laufer MR. Surgical management of congenital uterine anomalies. UpToDate 2012.
13. Heinonen PK. Clinical implications of the didelphic uterus: long-term follow-up of 49 Cases. Eur J Obstet Gynecol Reprod Biol 2000 Aug;91(2):183-190.

14. Al Sawah E, Plosker SM, Mikhail E. Laparoscopic hemi-hysterectomy and trachelectomy in a case of Herlyn-Werner-Wunderlich syndrome. Surg Technol Int 2016 Oct;XXIX: 181-184.
15. Altchek A, Paciuc J. Successful pregnancy following surgery in the obstructed uterus in a uterus didelphys with unilateral distal vaginal agenesis and ipsilateral renal agenesis: case report and literature review. J Pediatr Adolesc Gynecol 2009 Oct;22(5):e159-e162.
16. Lee YL, Weber AF. Using the tracheobronchial stent to achieve vaginal patency after repeated failed attempts in a patient with OHVIRA. J Pediatr Adolesc Gynecol 2013 Apr;26(2):e64.
17. Beck MM, Yohen N. Herlyn-Werner-Wunderlich syndrome: a case series and review of the literature Int J Gynecol Obstet 2015;17(1):1-4.
18. Wang J, Zhu L, Lang J, Liu Z, Sun D, Leng J, Fan Q. Clinical characteristics and treatment of Herlyn-Werner-Wunderlich syndrome. Arch Gynecol Obstet 2014 Nov;290(5):947-950.
19. Dias JL, Jogo R. Herlyn-Werner-Wunderlich syndrome: pre- and post-surgical MRI and US findings. Abdom Imaging 2015 Oct;40(7):2667-2682.