International Journal of Clinical Obstetrics and Gynaecology

ISSN (P): 2522-6614
ISSN (E): 2522-6622

www.gynaecologyjournal.com
2024; 8(4): 169-172
Received: 24-06-2024
Accepted: 23-07-2024

Dr. Subarna Debnath
Senior Resident, Department of Obstetrics & Gynecology. Agartala Government Medical College, Agartala, Tripura, India

Dr. JL Baidya
Professor, Department of Obstetrics & Gynecology. Agartala Government Medical College, Agartala, Tripura, India

Dr. Dhruba Prasad Paul
Assistant Professor, Department of Obstetrics & Gynecology. Agartala Government Medical College, Agartala, Tripura, India

Corresponding Author:
Dr. Subarna Debnath
Senior Resident, Department of Obstetrics & Gynecology. Agartala Government Medical College, Agartala, Tripura, India

Delayed diagnosis of a rare Mullerian anomaly in an adolescent girl: A case report

Dr. Subarna Debnath, Dr. JL Baidya and Dr. Dhruba Prasad Paul

DOI: https://doi.org/10.33545/gynae.2024.v8.i4c.1491

Abstract
Introduction: The syndrome of obstructed hemi-vagina and ipsilateral renal anomaly (OHVIRA), also known as Herlyn-Werner-Wonderlich syndrome is a rare congenital anomaly of the Mullerian ducts (paramesonephric).
Presentation of case: In this report we present a rare case of HWWS diagnosed in a 16-year-old female presenting with cyclical abdominal pain and lump in lower after the onset of menarche, describe the various diagnostic modalities and treatment options available, along with a current review of the literature.
Discussion: Patients with HWWS usually present within 1 year after menarche with hematocolpos on the side of the obstructed hemi-vagina, producing abdominal pain, dysmenorrhea, and abdominal mass. An early and correct diagnosis is necessary to relieve the symptoms and prevent complications caused by retrograde menstruation which may lead to endometriosis and subsequently infertility.
Conclusion: The purpose of this case report is to offer a better under-standing of the pathophysiology of HWWS so that when a young female presents with common, nonspecific symptoms, treating physicians may include HWWS on their list of differential diagnoses. Greater awareness of HWWS will lead to earlier detection and, consequently, reduced complications caused by delayed diagnosis.

Keywords: Mullerian duct, hemi-vagina, haematocolpos, delayed diagnosis

Introduction

- The syndrome of obstructed hemi-vagina and ipsilateral renal anomaly (OHVIRA), also known as Herlyn-Werner-Wonderlich syndrome is a rare congenital anomaly of the Mullerian ducts (paramesonephric).
- The prevalence of congenital Mullerian duct anomalies is reported to be 7% of all young women [1].
- Anatomically, HWWS is characterized by three key anomalies of the female reproductive tract: Uterus didelphys, unilateral blind hemivagina, and ipsilateral renal agenesis. With normal external genitalia, diagnosis is often delayed until after menarche [2] with most affected patients presenting with progressive dysmenorrhea and a suprapubic mass on abdominal exam.
- Other common presenting symptoms include intermenstrual bleeding, mucopurulent vaginal discharge, and fever [2, 3].
- Early diagnosis is key to preventing as endometriosis, infertility, and spontaneous abortion.
- The menstrual flow that comes from the patent unobstructed Hemi-vagina gives the appearance of normal menses. Consequently, accurate diagnosis and surgical treatment can be delayed for several months or even years [1].
- A small subset of individuals will have a micro-perforation of the hemivaginal septum.
- Ultrasonography may reveal uterus didelphys, associated hematocolpos or hematometrocolpos and renal agenesis [4].
- MRI is the gold standard for diagnosis and preoperative planning and provides more detailed evaluation of the anomaly such as cervical agenesis or aplasia or presence of communication between two cervices or vaginas [5].
- In this report we present a rare case of HWWS diagnosed in a 16-year-old female presenting with cyclical abdominal pain and lump in lower after the onset of menarche.
- We consider the patient's clinical history, illustrate the utility of various diagnostic modalities, describe common treatments, and review recent literature.

- The purpose of this case report is to offer a better understanding of the pathophysiology of HWWS so that when a young female presents with common, nonspecific symptoms, treating physicians may include HWWS on their list of differential diagnoses. Greater awareness of HWWS will lead to earlier detection and, consequently, reduced complications caused by delayed diagnosis.

Case report

- 16 years old unmarried girl presented with a history of cyclical pain in lower abdomen during her menstruation since her menarche and lump in lower abdomen felt during menstruation for last 1 year.
- The patient attained menarche at the age of 13 years and had regular menstrual cycles, once every 28 to 30 days with 3–5 days' duration of menstrual flow, which was associated with dysmenorrhoea.
- On physical examination, the patient was conscious, oriented and alert.
- Her vitals were within normal limits.
- Respiratory, cardiovascular, and nervous system examinations were within normal limits.
- Per abdomen examination revealed soft and tender 16 weeks gravid uterine sized lump arising from pelvis and occupying suprapubic and right iliac fossa, without guarding or rigidity.
- Patient underwent per speculum examination under anaesthesia which revealed a large cystic mass in the anterior and right vaginal wall. Cervical os could not be seen.
- All routine blood investigations were within normal limits.
- Ultrasonography of abdomen and pelvis revealed non-visualization of right kidney, two separate uterine horns,
- and a cystic collection posterior to uterine horns with low-level echoes within suggestive of haematometra and haematocolpos.
- MRI pelvis was done for further evaluation and revealed absent right kidney, uterine didelphys, haematocolpos, right-sided haematosalpinx; suggestive of Herlyn-Werner-Wunderlich syndrome. The distance between the introitus and longitudinal vaginal septum was 3.14 cm.
- CT IVP was done to evaluate the course of ureter and it showed absent right kidney and ureter with compression of the left ureter at the level of the crossing of the left common iliac artery with mild left sided hydronephrosis.
- Patient was planned for resection of the hemivaginal septum.
- Resection of the haemivaginal septum was done per-vaginally followed by drainage of haematocolpos and after resection of the septum, two separate cervices were visualised which confirmed the diagnosis of didelphic uterus.
- Margins of the resected septum was sutured to prevent stenosis.
- The post-operative period was uneventful.
- On follow-up, 1 month postoperatively, she had no complaints. One year after surgery, she remains asymptomatic.

Discussion

- The incidence of HWWS is not known.
- It is estimated that 6% of patients with uterine duplication have an obstructed hemivagina, and that renal agenesis is found in 63%-81% of uterine duplications and in 92%-100% of obstructed hemivaginas [6].
- Obstructed hemivagina and renal agenesis in patients with uterus didelphys seems more likely to occur on the right side [6].
- The obstruction usually occurs on the same side as the renal anomaly because the female reproductive tract develops at the same time and is close to the urinary system from the intermediate mesoderm.
- Patients with HWWS usually present within 1 year after menarche with hematometrocolpos on the side of the obstructed hemivagina, producing abdominal pain, dysmenorrhea, and abdominal mass [7, 8].
- Ultrasound may be sufficient to make a correct diagnosis but magnetic resonance imaging remains the preferred imaging method for investigation of mullerian duct anomalies particularly in pediatric patients [9].
- The syndrome falls under Type III Mullerian duct anomaly classification system of the American Society for Reproductive Medicine (ASRM).
- An early and correct diagnosis is necessary to relieve the symptoms and prevent complications caused by retrograde menstruation which may lead to endometriosis and subsequently infertility.
- Imaging particularly MRI, as in this case plays a major role in accurate diagnosis which could be missed clinically. It evaluates uterine morphology, detects communication between uterine and vaginal lumen, characterizes fluid contents and diagnose complications like endometriosis. [10].
- In our case, MRI pelvis revealed absent right kidney, uterine didelphys, haematocolpos, right-sided haematosalpinx; suggestive of Herlyn-Werner-Wunderlich syndrome. The distance between the introitus and longitudinal vaginal septum was 3.14 cm.
- It should be noted that in the majority of cases, the surgery of choice is the resection of the vaginal septum via colpohysteroscopy with preservation of the hemiuterus [11].
- Treatment of choice for OHVIRA syndrome is resection of the vaginal septum in order to achieve the continuity of the vagina.
- This may be performed as single-stage vaginoplasty with resection of the vaginal septum or a two-stage procedure with drainage of the hematocolpos first and then resection of the septum in the second procedure [12, 13].
- Single-stage vaginoplasty is the preferred method of treatment; two-stage procedure may be preferred in cases of infection or anatomic distortion [14].
- Single-stage vaginoplasty is rarely complicated and recurrent stenosis usually does not develop.
- In our case single stage vaginoplasty was done.
- Kapczuk, *et al*. reported a case of spontaneous perforation of the septum that developed bladder injury and vesicovaginal fistula after single-stage vaginoplasty [15].
- Vaginoplasty and resection of the vaginal septum may not be possible when the vaginal septum is proximally located and thick and hematocolpos is either small or non-existent; hemi hysterectomy may be needed in complicated cases [15-17].

Legends

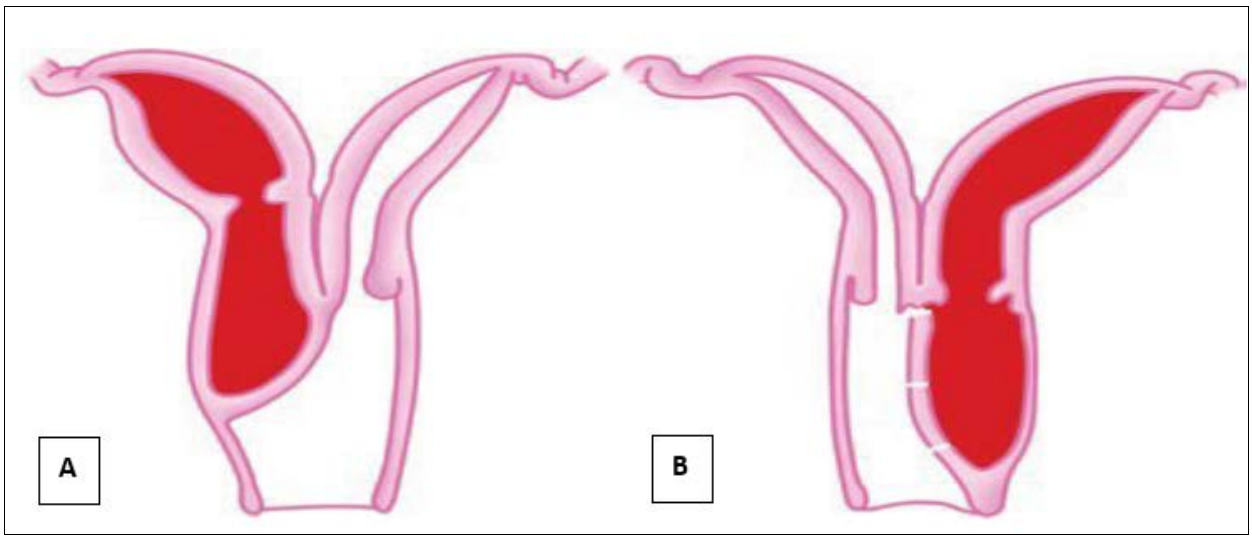
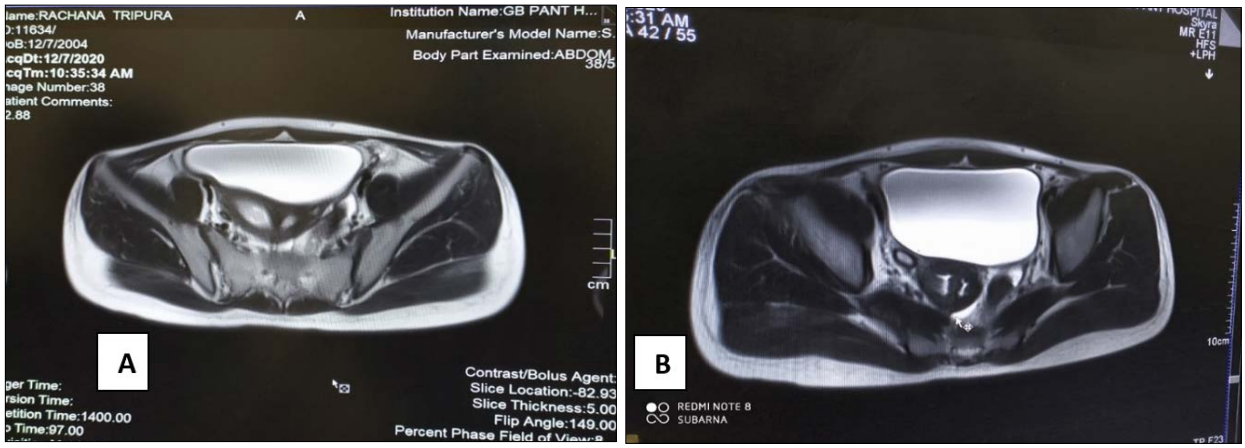
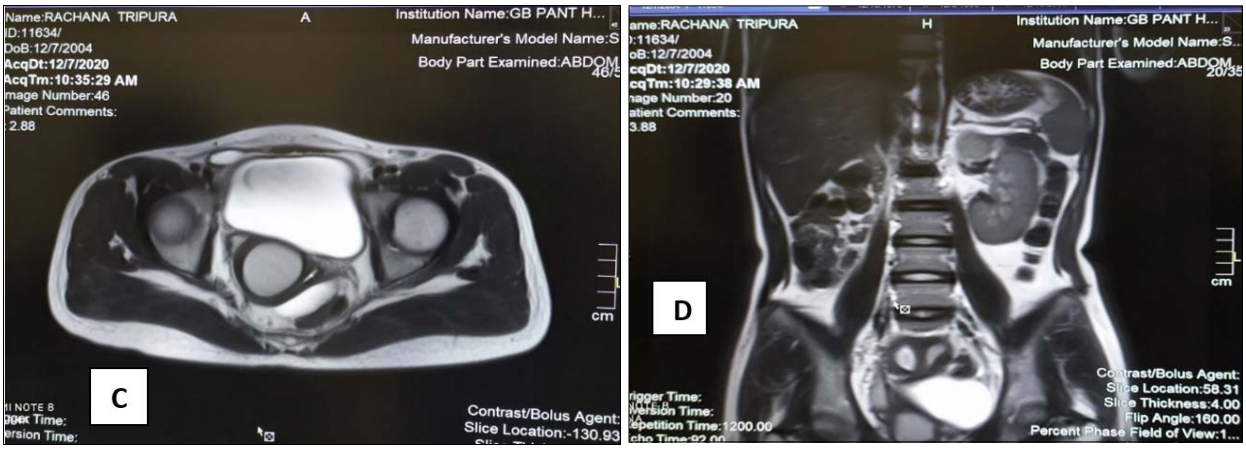


Fig 1: Uterine didelphys with obstructed hemi-vagina. A. Complete obstruction. B. Partial Obstruction



A. Uterine Didelphys with distended right horn B. 2 Cervices (Bicolis)



C. Right side obstructed hemi-vagina with haematocolpos D. Absent Right Kidney

Fig 2: MRI pelvis, T_2 WI axial view at different level

Conclusion

This case highlights the importance of recognizing Herlyn-Werner-Wunderlich syndrome (HWWS) in young women presenting with cyclical abdominal pain and lower abdominal masses after menarche. HWWS is a rare congenital anomaly involving obstructed hemi-vagina and ipsilateral renal anomaly, often leading to significant symptoms like dysmenorrhea and abdominal distention due to hematocolpos. Early and accurate diagnosis, facilitated by imaging modalities such as MRI, is crucial to prevent complications such as endometriosis and potential infertility. This case underscores the need for heightened awareness among clinicians to consider HWWS in differential diagnoses for adolescent females with nonspecific gynecological symptoms. Timely intervention, including

surgical resection of the vaginal septum, can significantly alleviate symptoms and prevent long-term complications. Enhanced understanding and early detection of HWWS can improve patient outcomes and reduce the risks associated with delayed diagnosis.

Conflict of Interest
Not available

Financial Support
Not available

References

1. Ashton D, Amin HK, Richart RM, Neuwirth RS. The incidence of asymptomatic uterine anomalies in women undergoing transcervical tubal sterilization. Obstet Gynecol. 1988;72:28-30.
2. Wang S, Lang JH, Zhu L, Zhou HM. Duplicated uterus and hemivaginal or hemicervical atresia with ipsilateral renal agenesis: An institutional clinical series of 52 cases. Eur J Obstet Gynecol Reprod Biol. 2013;170(2):507-511. DOI: 10.1016/j.ejogrb.2013.07.015.
3. Wang J, Zhu L, Lang J, *et al*. Clinical characteristics and treatment of Herlyn-Werner-Wunderlich syndrome. Arch Gynecol Obstet. 2014;290(5):947-950. DOI: 10.1007/s00404-014-3286-5.
4. Rastogi A, Khamesra A. Herlyn-Werner-Wunderlich syndrome: A rare urogenital anomaly masquerading as acute abdomen. Indian J Pediatr. 2010;77(8):917.
5. Hamidi H, Haidary N. Late presentation, MR imaging features and surgical treatment of Herlyn-Werner-Wunderlich syndrome (classification 2.2): A case report. BMC Womens Health. 2018;18:161.
6. Santos XM, Dietrich JE. Obstructed hemivagina with ipsilateral renal anomaly. J Pediatr Adolesc Gynecol. 2016;29:7-10.
7. Wu TH, Wu TT, Ng YY, Ng SC, Su PH, Chen JY, *et al*. Herlyn-Werner-Wunderlich syndrome consisting of uterine didelphys, obstructed hemivagina, and ipsilateral renal agenesis in a newborn. Pediatr Neonatol. 2012;53:68-71.
8. Angotti R, Molinaro F, Bulotta AL, Bindi E, Cerchia E, Sica M, *et al*. Herlyn-Werner-Wunderlich syndrome: An "Early" onset case report and review of literature. Int J Surg Case Rep. 2015;11:59-63.
9. Zhang H, Qu H, Ning G, Cheng B, Jia F, Li X, *et al*. MRI in the evaluation of obstructive reproductive tract anomalies in pediatric patients. Clin Radiol. 2017;7:612.e7-612.e15.
10. Piccinini PS, Doski J. Herlyn-Werner-Wunderlich syndrome: A case report. Rev Bras Ginecol Obstet. 2015;37:192-196. doi:10.1590/S0100-720320150005077.
11. Zhu L, Chen N, Tong JL, Wang W, Zhang L, Lang JH. New classification of Herlyn-Werner-Wunderlich syndrome. Chin Med J (Engl). 2015;128(2):222-225. DOI: 10.4103/0366-6999.149208.
12. Gungor Ugurlucan F, Bastu E, Gulsen G, Kurek Eken M, Akhan SE. OHVIRA syndrome presenting with acute abdomen: A case report and review of the literature. Clin Imaging. 2014;38:357-359.
13. Tong J, Zhu L, Lang J. Clinical characteristics of 70 patients with Herlyn-Werner-Wunderlich syndrome. Int J Gynaecol Obstet. 2013;121:173-175.
14. Smith NA, Laufer MR. Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome: management and follow-up. Fertil Steril. 2007;87(4):918-922.
15. Kapczuk K, Friebe Z, Iwaniec K, Kedzia W. Obstructive Mullerian anomalies in menstruating adolescent girls: A report of 22 cases. J Pediatr Adolesc Gynecol. 2018;31:252-257.
16. Donnez O, Jadoul P, Squifflet J, Donnez J. Didelphic uterus and obstructed hemivagina: Recurrent hematometra in spite of appropriate classic surgical treatment. Gynecol Obstet Invest. 2007;63:98.
17. Güdücü N, Gönenç G, İşçi H, Yiğiter AB, Dünder İ. Herlyn-Werner-Wunderlich syndrome: Timely diagnosis is important to preserve fertility. J Pediatr Adolesc Gynecol. 2012;25

How to Cite This Article
Debnath S, Baidya JL, Paul DP. Delayed diagnosis of a rare Mullerian anomaly in an adolescent girl: A case report. International Journal of Clinical Obstetrics and Gynaecology 2024;8(4):169-172.