CASE REPORT:

Herlyn-Werner-Wunderlich Syndrome in pregnancy

Novy Ngan, Erry Gumilar Dahlan, Eighty Mardiyan Kurniawati*
Department of Obstetric and Gynecology, Faculty of Medicine, Universitas Airlangga, Dr Soetomo Hospital, Surabaya, Indonesia

ABSTRACT

Objectives: To prevent Herlyn-Werner-Wunderlich syndrome's complication in pregnancy
Case Report: Presenting 2 cases pregnant with Herlyn-Werner Wunderlich syndrome, which is both of the case diagnosed at teenage and becoming pregnant then delivered by cesarean due to severe preeclampsia.
Conclusion: High suspectious is needed to diagnose Herlyn-Werner-Wunderlich syndrome and early diagnosis is better prognosis. The gold standard diagnostic is MRI. The treatment is resection of the vaginal septum, drainage, and marsupialization. Preconception Counseling is important to prevent or to reduce the pregnancy complications.

Keywords: Herlyn-Werner Wunderlich syndrome; Mullerian ducts; Wolffian ducts; uterus didelphys; haematocolpos; haematometra

ABSTRAK

Tujuan: Untuk mencegah komplikasi kehamilan dengan sindroma Herlyn-Werner Wunderlich
Laporan Kasus: Dua kasus hamil dengan sindroma Herlyn-Werner Wunderlich, dimana kedua pasien telah terdiagnosis pada saat remaja dan hamil kemudian melahirkan per abdominam oleh karena preeklampsia berat.
Simpulan: Kecurigaan yang tinggi diperlukan dalam mendiagnosis SHHW dan prognosis sindroma ini baik bila diagnosis dini. MRI merupakan gold standard diagnostik. Terapi sindroma ini adalah reseksi septum vagina, drainase dan marsupialisasi. Penting dilakukan Preconception Counseling (PCC) sebelum hamil, untuk mengurangi atau mencegah terjadinya komplikasi kehamilan dengan sindroma Herlyn-Werner-Wunderlich.

Kata kunci: Sindroma Herlyn-Werner Wunderlich; duktus Mullerian; duktus Wolfian; uterus didelfis; hematokolpos; hematometra

***Correspondence:** Eighty Mardiyan Kurniawati, Department of Obstetric and Gynecology, Faculty of Medicine, Universitas Airlangga, Dr Soetomo Hospital, Jl Prof dr Moestopo 6-8, Surabaya 60286, Indonesia. Email: ety_emka@yahoo.com

pISSN:0854-0381 ● eISSN: 2598-1013 ● doi: http://dx.doi.org/10.20473/mog.V27I22019.76-78
● Maj Obs Gin. 2019;27:76-78 ● Received 20 Feb 2018 ● Accepted 11 Jul 2018
 Available at https://e-journal.unair.ac.id/MOG/

INTRODUCTION

Herlyn-Werner Wunderlich syndrome (HWWS) is a rare congenital anomaly of Mullerian ducts and Wolffian ducts, with incidence of 1 : 2.000 to 1 : 28.000. It is most commonly diagnosed in puberty due to abdominal mass secondary to haematocolpos or haematometra, pain and dysmenorrhea.[1-5] The gold standart to diagnosed HWWS is with Magnetic Resonance Imaging (MRI), it can classification this sindrome also for treatment.[6] The long term complication is affected the reproductive system, but about 80% of it, can spontaneous pregnant. The complication of pregnancy with HWWS are miscarriage, premature labour, premature rupture of membranes, malpresentation, IUGR, preeclampsia and placenta accreta.[7-9]

Based on data from outpatient departement, devision of urogynecology, Dr. Soetomo Hospital, Surabaya, Indonesia from 2008 to 2017, we had 6 cases patient with Herlyn-Werner Wunderlich syndrome, which is 2 patient were pregnant.

CASE REPORT

Case 1 : Mrs. RH/ 31 yo

Patient was diagnosed with Herlyn-Werner Wunderlich syndrome at age 21 by ultrasound, BNO-IVP and laparo-vaginoscopy. USG revealed Didelphys uterus with hematometra, left and right uterus normal, Cystic mass at pouch of doughlas, endometrioma, left adnexa meas 42 x 98 mm, and right adnexa cannot be visualized. BNO-IVP result is Suspect of renal agenesis, left and the funtion of other renal was normal. Laparo-vaginoscopy revealed didelphys uterus, hydrosalping, left fallopian tube, with Non-paten fallopian tube bilateral, cystic mass at pouch of doughlas area, and cyst at anterior vaginal wall. She was treated with laaprotomy followed by adhesiolisis and drainage of hematosalping and chomotubation with patent right fallopian tube and resection of vaginal septum also done. Her menstrual history was regular accompany by dysmenorrhea.

She was pregnant after 7 month post resection of vaginal septum. She had done routine antenatal care, multivitamins given, no aspirin. Her BP range was 130/90 to 160/100 mmHg, she noticed her Blood Pressure (BP) increase for the first time at 29 weeks Age of Gestation (AOG), urinalysis result was +1 proteinuria. Serial ultrasound was done within normal limits. Cesarean section was done att 34 weeks AOG due to Preterm Premature Rupture of Membrane (PPROM) and severe preeclampsia, delivered baby boy, birth weight of 1700 grams with Apgar Score (AS) of 7-8. Her second baby also delivered by cesarean section at 33/34 weeks AOG due to PPROM, delivered baby girl, birth weight of 1800 grams with AS 7-8. At this time she didn't have any complaints with regular menses, no contraception used.

Case 2 : Mrs. AL/27 yo

She was diagnosed with HWWS at age 18 yo by ultrasound and MRI. USG result was didelphys uterus, ovarian cyst measuring 5,4 x 4,5 cm with suspect of renal agenesis. MRI revealed multiple congenital anomalies, which is double ureter at right renal, left renal agenesis, didelphys uterus, fusiform vagina measuring 5,3 x 2,1 cm with pus, she didn't did any treatment at that time. She had regular menstrual cycle with dysmenorrhea.

After 2 years of married (age 25) without contraception, patient was pregnant but uneventually she had miscarriage. After 4 month of her last pregnancy, she was pregnant. At 5/6 weeks AOG she had done resection of vaginal septum, drainage of pus and marsupialization due to hematometra. Patient had routine prenatal check up and had serial USG, which reveal normal limit. She underwent cesarean section at 38/39 weeks AOG due to severe preeclampsia, delivered baby boy, birth weight of 3350 grams with AS of 7-8. Patient had regular menses without dysmenorrhea noted. She used Combination Oral Contraceptive (COC).

DISCUSSION

Herlyn-werner-wunderlich syndrome (HWWS) is a rare case, the incidence of uterus didelphys in singleton pregnancy is 1 : 3.000.[1-5] Pregnancy with HWWS is a high risk pregnancy, because its correlated with Recurrent Pregnancy Lost (RPL), malpresentation, Intrauterine Growth Restriction (IUGR), preterm, PPROM, preeclampsia and placenta accreta.[9-12]

Its difficult to diagnosis HWWS because of the rarity of this case, high suspectious is needed. In case 1, the diagnosis was by USG, because of uterus didelphys, we suspect there is others anomaly, patient underwent laparo-vaginoscopy and to confirm diagnosis of HWWS BNO-IVP was done which revealed renal agenesis. Its same as case 2, where is USG and MRI was done and confirm the diagnosis of HWWS. MRI had accuracy of 96% to diagnosis HWWS.[3-6,13]

The treatment of choice patient with HWWS is resection of vaginal septum, drainage and marsupialization. It

had been done to both of cases. In case 1, after 7 months of procedure she was pregnant. In second case after pregnancy at 5/6 weeks AOG patient underwent the resection of the vaginal septum and drainage of pus, it maybe due to ascending infection from the vagina.[12-15]

The complication of pregnancy with HWWS can be prevent by Preconception Counseling (PCC) before get pregnant. In both cases, the patient had severe pre-eclampsia, which can be prevent by given aspirin low dose as early 13 weeks AOG. PPROM in case 1 can be predicted if patient had PCC before pregnancy, it can be screen for infection. Preterm labor also can be because of incompetent cervix, for the next pregnancy we can advised the patient to measured the cervical length. Abortus in case 2, it can be because of the abnormality of the implantation and decreased of vascularization at the implantation area.[16,17]

Mode of delivery patient with HWWS without obstructed hemivagina is per vaginam, except there is obstetric indication. HWWS patient with complete family should advised for sterilization because of chance to have placenta accreta due to abnormal of the uterus, which is the endometrium and myometrium are thin and the vascularization also decreased at the implantation area.[18]

CONCLUSION

High suspectious is needed to diagnose Herlyn-Werner-Wunderlich syndrome. The complications of pregnancy with Herlyn-Werner-Wunderlich syndrome can be prevent by Preconception Counseling (PCC).

REFERENCES

1. Bhoil B, Ahluwalia A, Chauhan N. 2016. Herlyn-Werner-Wunderlich syndrome with hematocolpos: an unsual case report of full diagnostic approach and treatment. Int J Fertil Steril. 2016;10(1):136-40
2. Cox D, Ching BH. 2012. Herlyn-Werner-Wunderlich syndrome: a rare presentation with pyocolpos. Journal of Radiology. 2012;6(3):9-15.
3. Del Vescovo et al. Herlyn-werner-wunderlich syndrome: MRI findings, radiological guide (two cases and literature review), and differential diagnosis. BMC Medical Imaging. 2012;12:4
4. Khaladkar SM, Kamal V, Kamal A, Kondapavuluri SK. 2016. The Herlyn-Werner-Wunderlich syndrome – A case report with radiological review. Polish Journal of Radiology. 2016;81:395-400.
5. Sucayan-Sta Ana MM, Gorgonio NM. Pregnancy in Herlyn-Werner-Wunderlich syndrome: A case report and review of literature. PJOG. 2015;39:35-42
6. Zhu L et al. New classification of Herlyn-Werner-Wunderlich syndrome. Chinese Medical Journal. 2015;128:222-5
7. Haberal et al. Placenta percreta with concomitant uterine didelphys at 18 weeks of pregnancy : a case report and review of the literature. J Matern Fetal Neonatal Med. 2016;29 (21):3445-8
8. Han BH et al. 2012. Uterus didelphys with blind hemivagina and ipsilateral renal agenesis (Herlyn-Werner-Wunderlich syndrome) suspected on the presence of hydrocolpos on prenatal sonography. Journal of Clinical Ultrasound. 2012;41(6):380-382
9. Britton RD, Lockwood CJ. 2014. Creasy and Resnik's Maternal-Fetal Medicine: Recurrent Pregnancy Loss. Elsevier. 2014. p. 707-17.
10. Chifan M, Tirnovanu M, Grigore M, Zanoschi C. 2012. Cervical incompetence associated with congenital uterine malformations. Rev Med Chir Soc Med Nat Lasi. 2012;116(4):1063-8.
11. Dietrich JE, Millar DM. 2014. Obstructive reproductive tract anomalies. J Pediatric Adolesc Gynecol. 2014;27:396-402.
12. Kendrick J, Holmen J, You Z, Smits G, Chonchol M. Association of unilateral renal agenesis with adverse outcome in pregnancy: A matched cohort study. Am J Kidney Dis. 2017;70(4):506-11.
13. Wang ZJ, Daldrup H, Coakley FV, Yeh BM. Ectopic ureter associated with uterine didelphys and obstructed hemivagina: preoperative diagnosis by MRI. Pediatric Radiol. 2010;40:358-60.
14. Heinonen PK Pregnancies in woman with uterine malformations, treated obstruction of hemivagina and ipsilateral renal agenesis. Arch Gynecol Obstet. 2013;287:975-8.
15. Tzialidou-Palermo et al. Diagnostic challenges of hemihematocolpos and dysmenorrhea in adolescents: obstructed hemivagina, didelphys or bicornuate uterus and renal aplasia is a rare female genitalia malformation. Arch Gynecol Obstet. 2012;286:785-91.
16. Weisberg E, Fraser IS. Contraception and endometriosis: challenges, efficacy, and therapeutic importance. Open Access Journal of Contraception. 2015;6:105-15.
17. Westland R, Schreuder MF, Ket JC, Van Wijk JA. 2013. Unilateral renal agenesis: a systemic review on associated anomalies and renal injury. Nephrol Dial Transplan. 2013;28:1844-55.
18. Rao SS et al. 2016. Pregnancy in uterus didelphys delivered by caesarean delivery: case report. International Journal of Reproduction, Contraception, Obstetrics and Gynecology. 2016;5(7):2434-7.