Diagnosis Approach, Treatment, Evaluation and Fertility Preservation in patient with Herlyn-Werner-Wunderlich Syndrome

ABSTRACT

Introduction: Herlyn-Werner-Wunderlich (HWW) syndrome is a very rare female congenital anomaly of urogenital tract. The definite etiology of HWWS is still unknown, but it may be caused by the abnormal development of Mullerian and Wolffian ducts. The characteristic triad of this syndrome includes didelphys uterus, obstructed hemivagina, and ipsilateral renal agenesis, recently known as Obstructed Hemivagina and Ipsilateral Renal Anomaly (OHVIRA) syndrome. The most common presentation is pain and dysmenorrhea, and abdominal mass in the lower abdomen secondary to haematocolpos and/or haematometra.

Case presentation: Seventeen-year-old female, presented with intermittent lower abdominal pain. Physical examination revealed in normal limit. From the ultrasaonography examination, it was found uterus didelphys with left hemiuteri hematomethra, hematocolpos, with normal anatomy and function of the right kidney, but there was no left kidney. From magnetic resonance imaging (MRI) pelvic and laparoscopy showing a complete duplication of the uterus from the horn to the cervix with no connection between the two uterine cavities, both ovaries were normal, the right fallopian tube was normal, the left tube was enlarged, attached to the uterus and the left ovary, no left kidney was found. From the previous surgery, there was a misdiagnosed with brown cyst. The laparoscopy guiding diagnostic and operative management of the vaginal septectomy procedure was performed.

Conclusion: HWW Syndrome diagnosis can be established with history taking, physical examination and appropriate diagnostic tools. Physical examination such as rectal toucher often missed, regardless it can be used to determine some differential diagnosis. MRI is most accurate for providing details regarding the altered anatomy and for identifying associated hematocolpos, hematosalping or hematometra for HWW

syndrome cases. Surgical intervention by vaginal septectomy is performed to relieve symptoms and several examination in the future, provide better reproductive for fertility preservation.
Keywords: Herlyn-Werner-Wunderlich syndrome, Obstructed Hemivagina and Ipsilateral Renal Anomaly (OHVIRA) syndrome

ABSTRAK

Pendahuluan: Sindrom Herlyn-Werner-Wunderlich (HWW) merupakan kelainan kongenital saluran urogenital perempuan yang sangat jarang terjadi. Etiologi pasti dari sindrom HWW masih belum diketahui, tetapi mungkin disebabkan oleh perkembangan abnormal dari duktus Mullerian dan Wolffian. Tiga karakteristik sindrom ini termasuk uterus didelphys, obstruksi hemivagina, dan agenesis ginjal ipsilateral, yang dikenal sebagai sindrom *Obstructed Hemivagina and Ipsilateral Renal Anomaly* (OHVIRA). Keluhan yang paling umum disampaikan adalah nyeri dan dismenorea, serta massa di perut bagian bawah ssebagai akibat adanya hematokolpos dan/atau hematometra.

Presentasi kasus: Seorang perempuan usia 17 tahun, dengan keluhan nyeri perut bawah intermiten. Pemeriksaan fisik didapatkan dalam batas normal. Dari pemeriksaan ultrasonografi ditemukan uterus didelphys dengan hematometra hemiuterus kiri, hematokolpos, dengan anatomi dan fungsi ginjal kanan normal, tetapi tidak didapatkan ginjal kiri. Dari *Magnetic Resonance Imaging* (MRI) panggul dan laparoskopi menunjukkan duplikasi lengkap rahim dari tanduk ke leher rahim tanpa hubungan antara kedua rongga rahim. Kedua ovarium normal, tuba fallopi kanan normal, tuba kiri membesar, menempel pada rahim dan ovarium kiri, tidak ditemukan ginjal kiri. Dari operasi sebelumnya, terdapat misdiagnosis dengan diagnosis kista coklat. Prosedur septektomi dilakukan dengan panduan laparoskopi diagnostik sekaligus sebagai terapi pembedahan.

Kesimpulan: Diagnosis sindrom HWW dapat ditegakkan dengan anamnesis, pemeriksaan fisik dan dukungan alat diagnostik yang tepat. Pemeriksaan fisik seperti *rectal toucher* sering terlewatkan, padahal dapat digunakan untuk menentukan

beberapa diagnosis banding. MRI paling akurat untuk memberikan rincian mengenai anatomi yang berubah dan untuk mengidentifikasi adanya hematokolpos, hematosalping atau hematometra pada kasus sindrom HWW. Intervensi bedah dengan septektomi vagina dilakukan untuk meredakan gejala serta kemungkinan beberapa pemeriksaan yang harus dilakukan di masa depan, memberikan fungsi reproduksi yang lebih baik untuk mempertahankan fertilitas.
Kata kunci: Herlyn-Werner-Wunderlich syndrome, Obstructed Hemivagina and Ipsilateral Renal Anomaly (OHVIRA) syndrome

Introduction

Herlyn-Werner-Wunderlich (HWW) syndrome is a rare female congenital anomaly with estimated occurrence is about 0.1%–3.8%. The exact etiology is still unexplained, but it may be caused by abnormality development of Mullerian and Wolffian ducts. It is defined as uterus didelphys and blind hemivagina associated with ipsilateral renal agenesis, which also known as obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome.[1] The estimated overall prevalence of Mullerian Duct Anomaly (MDA) is 2–3% of women. Uterus didelphys create 11% of MDAs in which associated renal anomalies are presented in approximately 43%.[2-5]

The syndrome is classified by complete or incomplete obstruction of the vagina as classification 1 (completely obstructed hemivagina) and classification 2 (incompletely obstructed hemivagina). The clinical findings in these two types are distinctly different.[1]

From the classification 1, the uterus behind the septum is completely isolated from the contralateral uterus, and no communication is presented between the

duplicated uterus and vagina. Hematocolpos may occur only a few months after menarche. Hematometra and hematosalpinx occurred in some more severely affected patients, as well as bleeding in the periadnexal and peritoneal space. Patients with this classification have an earlier age of onset, with a short time from menarche to present acute symptom of abdominal pain, fever, and vomiting. Classification1 is divided into classification 1.1-with blind hemivagina, there is no communication between the duplicated uterus and vagina; and classification 1.2-cervicovaginal atresia without communicating uteri, cervix behind the septum is atrectic or maldeveloped.[1]

From the classification 2, there is a small communication exists between the two vaginas, which makes the vaginal cavity behind the septum incompletely obstructed. The uterus behind the septum is completely isolated from the contralateral uterus. The menses can outflow through the small communication, but the drainage is impeded. These patients have a later age onset with chief complaint of purulent or bloody vaginal discharge that often comes years after menarche. Classification 2 is divided into classification 2.1-partial reabsorption of the vaginal septum, there is a small communication exists between two vaginas with resultant incomplete obstruction; and classification 2.2-with communicating uteri, there is a small communication exists between two duplicated cervices with a completely obstructed hemivagina.[1]

Case Presentation

A female in her teens, presented with intermittent left lower abdominal pain. She denied nausea, vomiting, fever, abdominal distension or abdominal mass. The menarche had occurred 3 years before, her cycles were regular and the menstrual bleeding usually lasted about 4–5 days with dysmenorrhoea. There was history of urinary disorders including micturition difficulty. From previously ultrasonography examination, she was suspected brown cyst then the doctor did laparotomy histerotomy drainage with vaginal cross insicion almost 1 year ago. After 5 months later she felt left lower abdominal pain again, bloody vaginal discharge with odor smell. Yet, she denied recent weight loss, change in bowel habits, and sexual activity. She also had no history of diseases.

From physical examination including rectal and external genitalia examination, the results were within normal limits. Actually on usual cases it can be found abnormal rectal examination that is an extraluminary mass in the anterior rectum, originating from the hematocolpos in the obstructed hemivagina.

A laboratory examination of complete blood and urine sample were in normal limits. There were no signs of urinary tract infection. From the trans-abdominal ultrasound that has been done, the results are as shown in Figure 1. There was uterus didelphys, the left hemiuteri size 9.5x4x1x4.6 cm with hematometra (+), while the right one size 4.1x3.1 cm and no hematometra. Hematocolpos was also found with the size 6.3x5.9x4.9cm, septum thickness was 5.8 mm, distance between introitus and interseptum was 4.7 cm, right kidney (+), left kidney was not visualized.

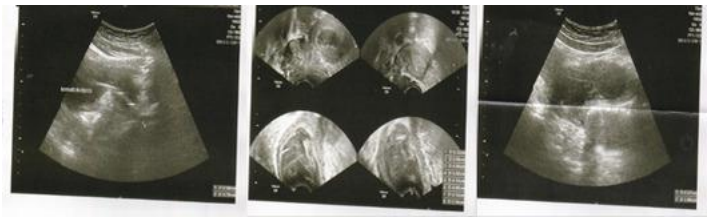

Figure 1. Trans abdominal ultrasonography

Pelvic MRI with contrast as shown in Figure 2 was carried out uterus didelphys, two vagina, left obstructed hemivagina, bilateral hidrocoplos with minimal bilateral hidrometra because of chronic hemorrhage. Other abnormality was agenesis left kidney which line to Herlyn Werner Wunderlich Syndrome (HWWS).

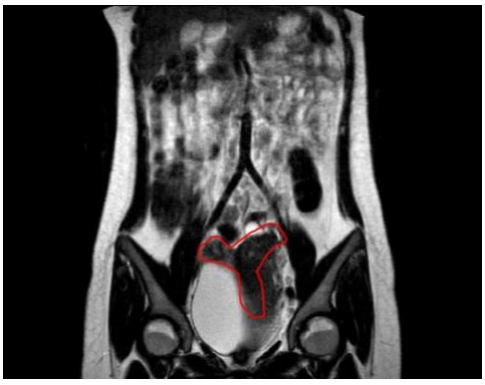

Figure 2. Pelvic MRI with contras

From the results of laparoscopy diagnostic, there were adhesion between omentum and anterior abdomen, didelphys uterus, left hematosalping, paratuba cyst, and left hematometra. The left hemivagina was identified, then it was appeared hematopyocolpos in the left vaginal septum. From Figure 3 and Figure 4 it can be seen that the patient has a non-communicating didelphys uterus, with hematometra on the

left hemiuterus, left hydrosalping, and hematocolpos in the left hemivagina which is totally obstructed. The patient also lacked of overall structure of the left kidney. A diagnosis of uterus didelphys with left hematometra, hematocolpos and hematosalping with left renal agenesis suggested of the Herlyn-Werner-Wunderlich syndrome (classification 1.2) was established.

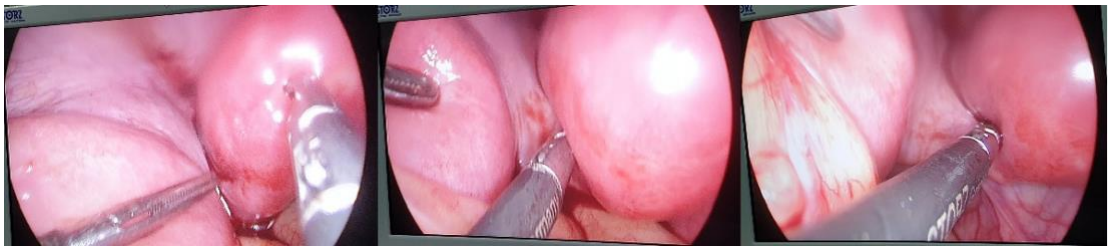

Figure 3. Laparoscopic shows didelphys uterus

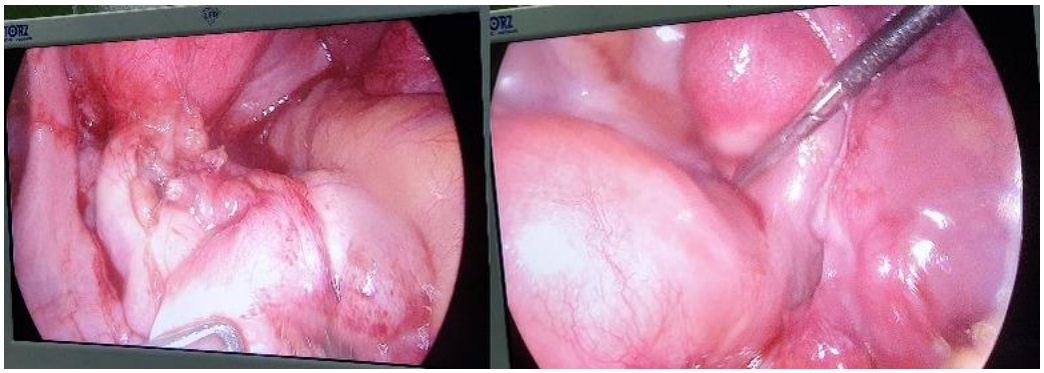

Figure 4. Laparoscopic shows left hematometra and left hydrosalphing

Discussion

In the reported cases, the misdiagnosed from the previous ultrasonography may be caused by appearance left hematometra which pushed the other hemivagina looked

like brown cyst. For the diagnosis of this syndrome, both ultrasound and MRI are very useful imaging techniques.[6] MRI is more accurate than hysterosalpingography (HSG) and ultrasound in the detection of acute and chronic complications.[7]

Vaginal septectomy procedure was performed over the entire length of the longitudinal vaginal septum, from the vaginal introitus to between the two uterine cervixes, about 7cm long. Sutures were made to the base of the septectomized vaginal tissue, to ensure that future adhesions would not occur. The condition of the uterus didelphys with each uterine cavity has a depth of 8 cm, and both uterine cervix that appear normal, do not need to be corrected at the time of vaginal septectomy. Long-term follow-up is required, although it is said that up to 87% of pregnancies can occur, even 62% of them can reach term gestation and undergo uncomplicated delivery.[2]

HWW syndrome or Obstructed Hemivagina and Ipsilateral Renal Anomaly (OHVIRA) syndrome is a rare female congenital anomaly due to development of Mullerian and Wolffian ducts. HWW Syndrome diagnosis can be established with history taking, physical examination and appropriate diagnostic tools. Physical examination such as rectal toucher often missed, regardless it can be used to determine some differential diagnosis. MRI is most accurate for providing details regarding the altered anatomy and for identifying associated hematocolpos, hematosalping or hematometra for HWW syndrome cases. Surgical intervention by vaginal septectomy is performed to relieve symptoms, provide better reproductive for fertility preservation. For further evaluation can be conducted by vaginal patency examination periodically, hysterosalpingography (HSG), and hysteroscopy.

Conclusion

1. HWW Syndrome diagnosis can be established with history taking, physical examination and appropriate diagnostic tools. Physical examination such as rectal toucher often missed, regardless it can be used to determine some differential diagnosis.
2. MRI is most accurate for providing details regarding the altered anatomy and for identifying associated hematocolpos, hematosalping or hematometra for HWW syndrome cases.
3. The use of laparoscopy can be done with the aim of establishing a diagnosis or to assist in surgical management.

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