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CASE REPORT

Diagnosis and Treatment in Patient With Herlyn-Werner-Wunderlich Syndrome: A Case Causing Pelvic Pain

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ABSTRAK

Latar Belakang: Sindrom Herlyn-Werner-Wunderlich (HWW) adalah varian langka dari kelainan duktus Mullerian. Trias khas dari sindrom ini adalah uterus didelfis, septum hemivagina, dan agenesis ginjal ipsilateral atau sindrom *Obstructed Hemivagina and Ipsilateral Renal Agenesis* (OHVIRA). Gejala paling sering adalah nyeri perut maupun panggul, dismenorea, dan terdapatnya massa pada perut akibat dari hematokolpos, hematosalping atau hematometra.
Tujuan: Untuk mengetahui penegakan diagnosis dan penatalaksanaan operatif satu kasus sindrom HWW dengan keluhan utama nyeri panggul.
Metode: Pemeriksaan kasus dan penelusuran rekam medis di RSUP Dr Sardjito Yogyakarta.
Hasil dan Pembahasan: Perempuan berusia 14 tahun dengan nyeri panggul. Pemeriksaan fisik terdapat massa kistik di perut bagian inferior, teraba hingga 3 cm inferior umbilikus. Dari pemeriksaan *Intravenous Pyelografi* (IVP) didapatkan anatomi dan fungsi ginjal kanan yang normal namun tidak didapatkan adanya ginjal kiri. Pada *CT scan* abdomen dengan kontras, *Magnetic Resonance Imaging* (MRI) dan laparoskopi menunjukkan duplikasi lengkap uterus dari cornu ke serviks tanpa hubungan antara dua rongga uterus, kedua ovarium normal, tuba fallopi kanan normal, tuba kiri membesar, menempel pada uterus dan ovarium kiri. tidak didapatkan ginjal kiri. Selanjutnya dilakukan penatalaksanaan operatif prosedur septektomi vagina.
Kesimpulan: MRI paling akurat untuk memberikan rincian mengenai perubahan anatomi dan untuk mengidentifikasi hematocolpos, hematosalping maupun hematometra pada sindrom HWW. Intervensi bedah berupa septektomi vagina dilakukan untuk meredakan gejala serta memberikan harapan fungsi reproduksi dan sexual yang lebih baik.

Kata kunci: Sindrom Herlyn-Werner-Wunderlich; disgenesis Mullerian; nyeri panggul.

ABSTRACT

Background: Herlyn-Werner-Wunderlich (HWW) syndrome is a rare variant of Mullerian duct anomalies. 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 abdominal and pelvic pain, dysmenorrhea, and abdominal mass secondary to hematocolpos, hematosalping or hematometra.
Objective: to determine the diagnosis and operative management of HWW syndrome case with pelvic pain as the chief complaint.
Method: case examination and tracing medical records at Dr Sardjito Hospital Yogyakarta.
Results and Discussion: Fourteen-year-old female, presented with pelvic pain. Physical examination revealed a cystic mass in the abdomen inferior, palpable up to 3 cm inferior to the umbilicus. From the Intravenous Pyelography (IVP) examination, it was found normal anatomy and function of the right kidney, but there was no left kidney. On a contrast-enhanced abdominal CT scan, magnetic resonance imaging (MRI) 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. Operative management of the vaginal septectomy procedure was performed.
Conclusion: 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 and sexual functions.

Keywords: Herlyn-Werner-Wunderlich syndrome; Mullerian disgenesis; pelvic pain.

INTRODUCTION

Herlyn-Werner-Wunderlich (HWW) syndrome is a rare form of Mullerian Duct Anomalies (MDA). The characteristic triad of this syndrome includes didelphys uterus, obstructed hemivagina and ipsilateral renal agenesis (hence, also known as OHVIRA syndrome).[1] The incidence of the didelphys uterus is approximately 1 in 2000 to 1 in 28.000, and it is accompanied by unilateral renal agenesis in 43% of cases. The incidence of unilateral renal agenesis is 1 in 1100 and between 25% until 50% of affected women have associated genital anomalies.[2-4]

HWW syndrome is classified based on the existing vaginal obstruction. This classification can be generalized by gynecologists around the world. Based on complete or incomplete hemivaginal obstruction, HWW syndrome can be classified as follows: Classification 1, completely obstructed hemivagina and Classification 2, imperfectly obstructed hemivagina. The clinical features associated with these two classifications are very different.[5]

Classification 1 is more susceptible to hematometra, hematosalping and hemoperitoneum in more severe conditions. Acute abdominal pain, fever, and vomiting are common symptoms a few months after menarche. Endometriosis is a common complication, which can develop into secondary endometriosis, pelvic adhesions, pyosalping or pyocolpos, if not managed properly. Classification 2 is usually accompanied by complaints of vaginal discharge mixed with pus or blood, and even genital system infections can occur. Most patients with menstrual cycles are generally normal, but with longer menstrual periods, followed by chronic pain.[5]

METHODS

Starting with a case search, then an examination is carried out according to the procedure. Completeness of data obtained from medical record of Sardjito hospital Yogyakarta.

RESULTS AND DISCUSSION

Results

Fourteen-year-old female, presented with pelvic pain. She denied nausea, vomiting, abdominal distension, hematochezia, fever, chills, diarrhea, or constipation. The menarche had occurred 1 year before, her cycles were irregular and the menstrual bleeding usually lasted about 4–5 days with severe dysmenorrhoea. She had not been sexually active yet. There is no history of urinary disorders including burning micturition. She also denied recent weight loss, change in bowel habits and has no history of diseases or surgical interventions.

Physical examination revealed a lower abdominal mass below 3 cm of the umbilicus, with mild lower abdominal pain. There is no abdominal distension. Rectal examination revealed an extraluminary mass in the anterior rectum, originating from the hematocolpos in the obstructed left hemivagina. From the examination of external genitalia, the results were within normal limits.

A laboratory examination of a urine sample is performed with a negative pregnancy test result. There were no signs of urinary tract infection. Complete blood count results are within normal limits. A CT scan with contrast was carried out with no left kidney, no visible abnormalities in the bladder, rectum, pancreas, liver, bladder, and spleen.

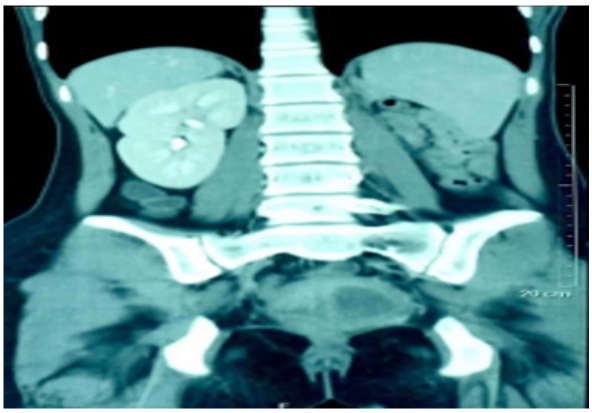
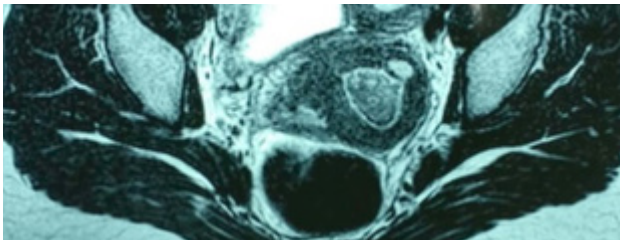


Image 1. Coronal View of the CT Abdomen and Pelvis Shows Absent Left Kidney



Images 2. Axial Image of The Pelvis Shows The Uterine Horns Duplication and Endometrial Cavities Consistent with Didelphys

On MRI there was a complete duplication of the uterus from the horn to the cervix without any communication between the two uterine cavities. The right uterine cavity appears to be dilated by fluid-intensity lesions (T1 hypointense and T2 hyperintense), while the left uterine cavity is dilated by mixed fluid-blood lesions (T1 hypo-hyperintense and T2 hyperintense). The right vagina is duplicated with the distal aspect of the left vagina. There is no prominent dilatation of the right side of the vagina. Right and left ovaries within normal limits. The left salping was dilated with fluid filled lesions. Abdominal ultrasound was performed for further evaluation showing bilateral normal ovaries, dilated uterus and left fallopian tube with internal echo consistent with hematometra and left hydrosalping. There is no left kidney.

On the Intravenous Pyelogram (IVP) examination at the 15th and 30th minutes, it was seen that the contrast material filled the right pelvicocalyx system, right ureter and bladder. The caliber and pelvic system of the kidneys are within normal limits, the nephrogram in the left kidney and left ureter is not visualized. On laparoscopic examination shows uterus didelphys, both ovaries and right tube are within normal limits, the left tubes appear to be enlarged and adhere to the uterus and ovaries.

From the results of these investigations, it can be seen that the patient has a non-comunicating didelphys uterus, with hematometra on the left hemiuterus, left hydrosalping, and hematocolpos in the left hemivagina which is totally obstructed. The patient also lacked the overall structure of the left kidney and left ureter. All of these results support the diagnosis of HWW syndrome.

DISCUSSION

The exact etiology and pathogenesis of HWW syndrome are not well known. It is generally said that these congenital abnormalities include anomalous development of the Mullerian (paramesonephric) and Wolffian (mesonephric) ducts. The internal genital organs and lower urinary tract are derived from two paired urogenital structures that develop in both sexes namely the Wolffian duct and the Mullerian duct.

In women, the Mullerian duct is located just beside the Wolffian duct, which acts as an inductor element, grows downward towards the midline, crosses the Wolffian duct, comes into contact with each other, and joins to form the uterovaginal canal, forming the fallopian tubes, uterus and two-thirds the upper part of the vagina. Not developing or failing to fuse the distal segments of the Mullerian duct can result in uterine hypoplasia or agenesis, unicornuate, didelphys, bicornuate, septate and arcuate. The wolffian duct is the origin of the kidney, and is an inductor element for Mullerian duct fusion. Anomalous development of the tail section of one of the Wolffian ducts is the cause of unilateral renal agenesis associated with imperforate hemivagina.[3]

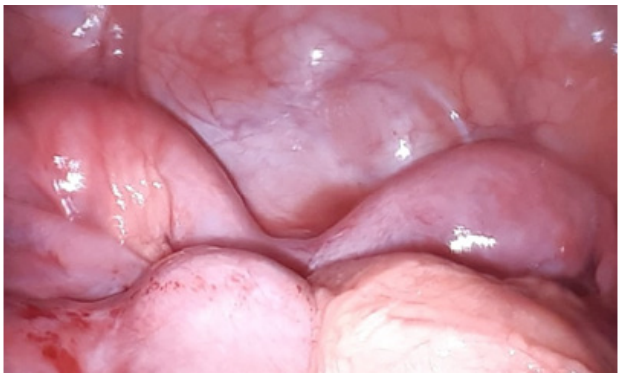
In the reported cases there were no abnormalities in the appearance of the external genitalia. The abnormality is found in the internal genitalia in the form of a didelphys uterus, with left-sided hematosalping and vaginal duplication. The left side of the hemivagina is obstructed so that there is a hematocolpos. The presence of obstruction results in pain with each period or dysmenorrhoea. In more chronic conditions, complaints of pain can be felt continuously. Long term, the risk of endometriosis cannot be ruled out.[6]

For the diagnosis of this syndrome, both ultrasound and MRI are very useful imaging techniques.[2,7] Three-D ultrasound can be used both abdominally and vaginally and allows to see images of the three planes orthogonally. This technique seems to provide high quality images similar to those obtained by MRI and high precision in the diagnosis of congenital anomalies, as well as a lower cost and better tolerance. MRI is more accurate than hysterosalpingography (HSG) and ultrasound in the detection of acute and chronic complications.[7] Magnetic Resonance Imaging (MRI) is the modality of choice for the diagnosis of HWW syndrome and other congenital disorders because of its ability to display better anatomical images of the internal genital organs and high sensitivity to indicate the presence of blood.[6]

In cases of genitourinary anomalies, initial ultrasound examination is preferable to MRI rather than CT. This option is because the CT examination involves more radiation exposure as well as limited

soft tissue resolution. In the case of uterovaginal congenital abnormalities, proper anatomical delineation of the uterus, tubes, cervix, and vagina is essential. The presence or absence of and the severity of endometriosis, pelvic inflammation, and adhesions can be evaluated properly with MRI.[6]

Laparoscopy remains the gold standard for diagnosis of female genital tract abnormalities but is only used when MRI is unavailable or fails to establish a diagnosis.[3] Laparoscopy is also a gold standard diagnostic modality that can provide an added advantage as it allows for a hematocolpos or hematometra therapeutic drainage procedure, vaginal septectomy, or marsupialization. Laparoscopic excision of the vaginal septum is the management of choice for HWW syndrome. Pregnancy was reported in about 87% of patients, although 23% had a risk of abortion.[6] IVP can be used to visualize urinary tracts and provide an overview of kidney function, in this case the right kidney has a normal function and there are no abnormalities in the right ureter.



Images 3. Laparoscopic Finding Shows Uterus Didelphys and Normal Finding of Both Ovaries

The surgical management of cases of HWW syndrome accompanied by hemivaginal obstruction resulting in secondary hematocolpos is a septectomy procedure. The main purpose of this procedure is to relieve pain symptoms by making menstrual blood flow, no more hematocolpos. It is hoped that in the future the reproductive process can occur properly.[1,7,8] As far as possible, delay in diagnosis should be avoided because it will cause or even worsen existing endometriosis.[6]

In this patient, she developed pelvic pain due to vaginal obstruction with secondary hematocolpos, which in turn this process continues into a hematometra in the left hemiuterus. There was no hematometra in the right hemiuterus of the uterus didelphys or hematocolpos in the right hemivagina because there was no obstruction in the channel on the right side. In addition, there is a non-communicating condition in the didelphys uterus, so that menstrual blood from the left hemiuterus cannot flow or mix with menstrual blood in the right hemiuterus.

Based on the physical abnormalities obtained, it was decided to perform surgical management of vaginal septectomy on the obstructed vaginal septum. The vaginal septum in this patient was a complete longitudinal vaginal septum which closed access to all sides of the left hemivagina. Septectomy is performed on the entire septum along the vagina, so that there are no more obstacles to the passage of menstrual blood from the left hemiuterus. Currently, only vaginal septectomy is performed for correction, with the aim of relieving the obstruction and associated pain. The patient's non-communicating didelphys uterus has not been corrected yet.

Further evaluation in the long term is needed to determine whether the patient requires surgical correction of the uterus or not. This may depend on the patient's future reproductive function. Although uterine didelphys has not shown to affect fertility, the rates of spontaneous abortions have been reported ranging from 23%-40%.[1,2] A study by Wang *et al* reported 52.9% ipsilateral pregnancies following excision of the vaginal septum. Cesarean section may be required for successful delivery.[9,10]

CONCLUSIONS

To summarize, HWW syndrome is a rare disorder of MDA characterized by didelphys uterus and OHVIRA. The early detection is important due to the possible associated complications. MRI is most accurate for providing details regarding the altered anatomy and for identifying hematocolpos, hematosalping, or hematometra. Surgical interventions are to relieve symptoms and ensure successful reproductive and sexual function.

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