Review
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Herlyn-Werner-Wunderlich syndrome with pyohematocolpos: a case report and review of literature

Herlyn-Werner-Wunderlichov sindrom s piohematokolposom: prikaz bolesnice i pregled literature

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Summary

Herlyn-Werner-Wunderlich syndrome (HWWS) is a rare malformation syndrome of the women reproductive tract characterized by uterus didelphys with obstructed hemivagina and ipsilateral renal agenesis. We report here a case of a 20 year-old patient presented to the emergency department with pain in the right lower quadrant. The clinical exam showed a vaginal fluctuant painful mass obliterating the right part of the vagina. Transvaginal ultrasound showed uterus didelphys, the right uterus was dilated with a dense fluid collection which corresponded to pyohematometra. It also revealed a dense vaginal collection which corresponded to pyohematocolpos. Abdominal ultrasound showed the absence of a right kidney. The laparoscopy showed uterus didelphys with a large right uterus. We performed the resection of the vaginal septum to reconstruct one vagina. The first follow-up visit revealed a healthy wound with no adhesion of the vaginal wall. Prompt and accurate diagnosis of female reproductive tract disorders, including HWWS, is necessary to prevent complications and preserve future fertility.

Key words: Herlyn-Werner-Wunderlich syndrome, pyohematocolpos, pyohematometra, ultrasound, laparoscopy

Sažetak

Herlyn-Werner-Wunderlichov sindrom (HWWS) je rijetka malformacija ženskog reproduktivnog sustava karakterizirana uterusom didelfisom, opstrukcijom hemivagine i istostranom agenezom bubrega. Prikazujemo slučaj 20-godišnje bolesnice koja se javila u hitnu službu s bolovima u donjem desnom kvadrantu. Klinički pregled ukazao je na bolnu fluktuirajuću masu koja je obliterirala desni dio rodnice. Transvaginalni ultrazvuk pokazao je prisutnost uterusa didelfisa, s time da je desni uterus bio dilatiran uslijed gustog tekućeg sadržaja koji je odgovarao piohematometri. Također je nađena vaginalna nakupina gustog tekućeg sadržaja koja je odgovarala piohematokolposu. Abdominalni ultrazvuk ukazao je na odsutnost desnog bubrega. Laparoskopija je pokazala uterus didelfis s uvećanim desnim uterusom. Resecirali smo vaginalni septum kako bi rekonstruirali jednu vaginu. Prvi kontrolni pregled je pokazao uredno cijeljenje rane bez priraslica vaginalne stijenke. Promptna i precizna dijagnoza malformacija ženskog genitalnog sustava, uključujući HWWS, potrebna je u svrhu prevencije komplikacija i očuvanja buduće plodnosti.

Ključne riječi: Herlyn-Werner-Wunderlichov sindrom, piohematokolpos, piohematometra, ultrazvuk, laparoskopija

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Introduction

Herlyn-Werner-Wunderlich syndrome (HWWS) is a rare congenital malformation syndrome characterized by a triad of uterus didelphys with obstructed hemivagina and ipsilateral renal agenesis, also known as OHVIRA.[1] In 1922., Purslow first described this syndrome in a young woman who presented with gradually increasing pelvic pain and a pelvic mass with regular menstruation.[2] The triad was reported in 1971. by Herlyn and Werner[3] and again in 1976. by Wunderlich.[4] The reported incidence for this anomaly is 0.1 to 3.8%.[5] The mean age of onset in patients with complete obstruction of the hemivagina is 13 years, with average time of four months from menarche to symptoms.[6] In cases of incompletely obstructed hemivagina the onset of symptoms may occur at a later time. Clinical manifestations are unspecific and most often include abdominal pain, painful menstruation and a palpably vaginal mass secondary to hematocolpos. Rarely, patients may develop pyohematocolpos, pyosalpinx and peritonitis as a result of an ascending infection due to retained discharge or menstrual blood in the obstructed hemivagina.[7] Ultrasound, magnetic resonance imaging and laparoscopy are used to establish the diagnosis.

Case report

A 20 year-old girl presented to the emergency department with pain in the right lower quadrant and fever 37.7°C. The laboratory test showed leukocytosis (leukocits were 23700×10^9/L). C reactive protein was 10.6 mg/L. She was on the fourth day of menstrual bleeding. Her medical history was uneventful. She had menarche at 13 years and a normal menstrual cycle of 28/5 days. The clinical exam showed normal external genitalia and a vaginal fluctuant painful mass obliterating the right part of the vagina. In the right vaginal wall, there was a defect through which bad-smelling purulent discharge mixed with the blood flowed. Normal cervix was identified on the speculum exam. The pelvic ultrasound showed uterus didelphys, the right uterus was dilated with a dense fluid collection which corresponded to pyohematometra. It also showed a dense vaginal collection of 10×4 cm which corresponded to pyohematocolpos (Figure 1). Abdominal ultrasound showed the absence of the right kidney. The patient was scheduled for surgical treatment. The laparoscopy showed uterus didelphys with a large right uterus (Figure 2). The ovaries were normal. No endometriosis implants were identified. The fluid collection in the blocked vagina was drained, evacuating a bloody and purulent content. We performed resection of the vaginal septum to reconstruct one vagina (Figure 3). After that treatment, the right cervix became visible. A final vaginoscopy showed a unique vaginal cavity with double cervix. The patient tolerated the procedure well and was discharged from hospital on the 3rd postoperative day. The first follow-up visit was four weeks after the operation. She had no symptoms and no complications were observed and the right hemivagina had collapsed. Vaginal examination revealed a healthy wound with no adhesion of the vaginal wall. Thus, her recovery was uneventful.

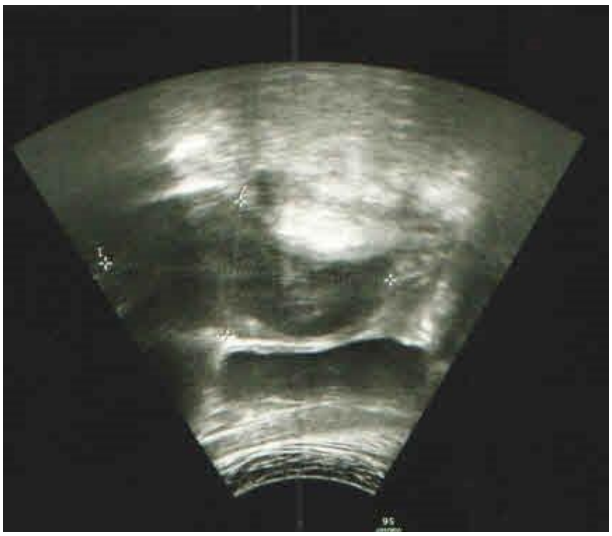

Figure 1 Ultrasound finding of the dilated, hemiobstructed vagina with fluid collection 10×4 cm (pyohematocolpos)
Slika 1. Ultrazvučni prikaz dilatirane, djelomično opstruirane vagine s nakupinom tekućine 10×4 cm (piohematokolpos)

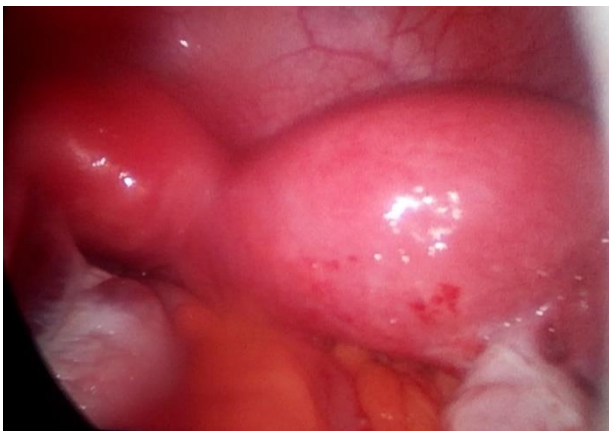

Figure 2. Intraoperativelly laparoendoscopic finding: left normal uterus, right large uterus (pyohematometra)
Slika 2 Intraoperativni laparoendoskopski nalaz: lijevo normalan uterus, a desno povećan uterus (piohematometra)

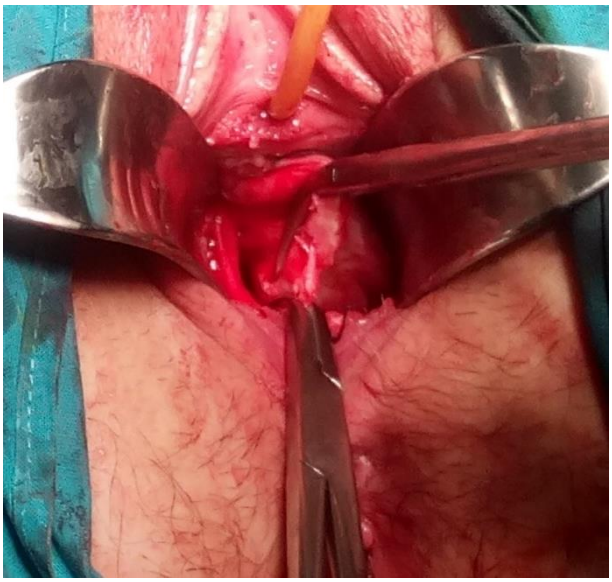

Figure 3 Intraoperative finding of the incised and resected vaginal septum
Slika 3. Intraoperativni prikaz incidiranog i reseciranog vaginalnog septuma

Discussion

HWWS is a rare complex congenital anomaly of the urogenital tract that involves abnormal development of the Mullerian and mesonephric ducts in female embryos. It is also known as the OHVIRA syndrome (Obstructed hemivagina and ipsilateral renal agenesis). The syndrome falls under type III Mullerian duct anomaly classification system of the American Society for Reproductive Medicine. In HWWS there is an insult to the paramesonephric system and metanephros. The uterus, fallopian tube, cervix, and upper two-thirds of the vagina develop from the paired paramesonepric ducts. The duct arises from the urogenital ridge. Then, caudally, it runs lateral to the mesonephric duct, and finally, in the midline, it comes in close contact with the upper part of the vagina.[8] When they fail to fuse, they produce two hemiuteri and hemicervicis, resulting in mulerian anomalies associated with HWWS.[9] An insult to the metanephric diverticulum results in ipsilateral agenesis of the ureter and kidney.[10] The simultaneous insult to the paramesonephric system and metanephros could suggest a multifactorial origin. Renal agenesis related genes such as CHD1L, TRIM32, RET and WNT 4 may be associated with HWWS.[11] Other anomalies include horseshoe kidney, pelvic kidney, renal dysplasia, duplication of the kidneys and ureters, ectopic ureter, high-riding aortic bifurcation, IVC duplication, intestinal malrotation and ovarian malposition.[12] Rarely, adenocarcinoma of the obstructed side of the uterine cervix and clear cell carcinoma of the obstructed portion of the vagina are also noted.[13,14] According to the proposed classification system based on a review of 79 patients[15], HWWS is categorized as: Classification 1 – a completely obstructed hemivagina (1.1-with blind hemivagina; 1.2-cervicovaginal atresia without communicating uteri) and Classification 2-an incompletely obstructed hemivagina (2.1-partial resorption of the vaginal septum; 2.2-with communicating uteri). A right-sided prevalence has been described.[16] Our case falls under classification 2.1.(Figure 4).

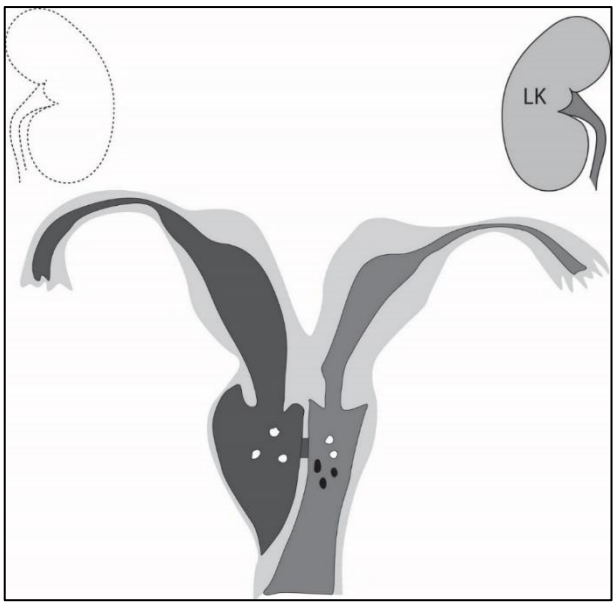

Figure 4 Diagrammatic representation of OHVIRA. On the right side pyohematometra, pyohematocolpos, incompletely obstructed hemivagina and renal agenesis are clearly visible.
Slika 4. Shematski prikaz OHVIRA-e. Na desnoj strani se jasno vide piohematometra, piohematokolpos, djelomično opstruirana hemivagina i renalna ageneza.

Classically, a patient with HWWS can present with severe dysmenorrhea a few months to 1 year after attaining menarche.[17] Other patients can present with a pelvic or vaginal mass secondary to hematocolpos, abnormal vaginal discharge, acute retention of urine, infertility, complicated pregnancy and labor or endometriosis. Usually, HWWS remains undiagnosed and asymptomatic during early childhood with normal external genitalia. Compared to patients with incomplete obstruction, patients with completely obstructed hemivagina are diagnosed earlier (often soon after menarche) and more likely present with symptoms of abdominal pain, fever and emesis than with mucopurulent vaginal discharge common in patients with incomplete obstruction. Pyocolpos can occur in patients with HWWS due to

secondary infection of retained menstrual blood in the obstructed hemivagina. The two vaginal cavities can communicate through partially fenestrated septum, or the two cervicies can communicate through fistula.[18] Additionaly, women with complete obstruction are more susceptible to complications such as hematometra, hematosalpinx, hemoperitoneum and even pelveoperitonitis, as obstruction causes retention of the menstrual flow in the internal female genitalia.[19] Another uncommon presentation is urinary retention which occurs because the mass of hematocolpos is quite large. At that size, it causes urethra angulation which leads to an obstruction.[20] In the absence of treatment, the natural course of the disease also includes complications such as endometriosis and pelvic adhesions.[21] The fertility of patients with HWWS with early diagnosis and treatment is usually not impaired.[22] Some authors mentioned that in 87% of HWWS cases pregnancy could be achieved and 62% of them had delivery at term.[23] However, premature labour in patients with HWWS occurs more commonly compared than in the general population, although the prevalence of misscarriages does not differ.[24] Approximately 84% of pregnancies require caesarean section. The most common indication is breech presentation, which is reported in 51% of cases.[22] Fetal growth restriction is a possible complication that can increase the rate of caesarean section.[22] Clinical suspicion and awareness of the syndrome are imperative to make a timely diagnosis and prevent complications. Ultrasound and MRI are the most widely used diagnostic tools.[24-26] It is reported to note that CT imaging is not recommended for syndrome diagnosis as it less accurate and subjects the patient to ionizing radiation.[26] Ultrasound examination is an inexpensive and convenient method of screening for HWWS. Sonographic features include uterine anomalies (didelphis/bicornuate uterus), hematometra, hematocolpos, pelvic fluid collection (often hemoperitoneum), and ipsilateral renal agenesis with compensatory hypertrophy of the contralateral kidney.[27] MRI is considered to be more sensitive for imaging soft-tissue anatomy and delineating subtle findings seen in congenital anomalies. MRI is the imaging modality of choice for the diagnosis and classification of HWWS as it provides details about uterine morphology, including contour and intrauterine cavity shape and continuity with each vaginal lumen, and nature of the fluids in these cavities. Evaluation of the genital tract using MRI scanning is recommended in all girls with known renal abnormalities detected antenatally or after that, before the onset of menstruation. This enables us to diagnose some patients before menarche and carry out a surgical correction of the obstruction before any damage has occured because of hematocolpos, hematometra and retrograde menstruation.[27-29] It can also identify associated pathologies such as endometriosis, pelvic inflammation and adhesions, as well as renal abnormalities. Laparoscopy is not mandatory but could help confirm the diagnosis when radiologic imaging is inconclusive.[1] Resection of the vaginal septum is the treatment of choice for obstructed hemivagina. Candiani and coauthors have suggested marsupializing the vaginal margins after excision of the septum to allow ample drainage of the purulent material and better expose the cervix.[30] Smith and Laufer reported of 27 cases, there were six cases underwent two-stages vaginoplasty due to anatomical distortion, infection and stenosis.[1] Rarely, unilateral hysterectomy may be considered in case of reccurent stenosis.[31] Surgery is important to relieve the obstruction, alleviate symptoms and prevent complications of retrograde flow. Some authors prefer a hysteroscopic incision. The advantages of the last method include, among others, avoiding the risk of the use of general anesthesia in an operating room setting and the possibility of performing in the office. Boyraz and coauthors performed laparoscopic resection of the vaginal septum to preserve virginity.[32] Regular gynecological control is important in postoperative management, aimed at assessing the patency of the residual vagina. In some cases, it is necessary to expand the formed passage to avoid its secondary closure.

Prompt and accurate diagnosis of female reproductive tract disorders, including HWWS, is necessary to prevent complications and preserve future fertility. Early recognition of this relatively rare syndrome would lead to the immediate, proper surgical intervention. A multidisciplinary approach guided by a gynecologist, radiologist, pediatric specialist and pediatric surgeon is fundamental to avoid complications and achieve a better outcome.

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