Herlyn-Werner-Wunderlich Syndrome with Hematopyocolpos and Cystocele: A Case Report

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Abstract: Introduction: Herlyn-Werner-Wunderlich Syndrome (HWWS) is a rare congenital anomaly of female urogenital tract caused by abnormalities in the development of the Mullerian ducts resulting in uterus didelphys, obstructed hemivagina, and associated ipsilateral renal anomaly. HWWS patients are usually diagnosed around or after puberty. We present the case investigation of HWWS patient with hematopyocolpos and cystocele. Case Report: A 14-year-old girl came with abdominal enlargement and lower abdominal pain. A cystic mass with tenderness on the right iliac fossa was found on abdominal examination. Per-rectal examination revealed a cystic mass with a smooth surface in the right lateral region of the midline. The abdominal ultrasound revealed a single left kidney, uterus didelphys, and hematocolpos in right hemivagina. Bulging cystic mass with smooth surface on right lateral vaginal wall was found from vaginal examination under general anesthesia. The patient underwent right vaginal wall excision, neovaginal reconstruction, and anterior colporrhaphy. One month post-operation follow-up, there’s improvement condition and menstruation without lower abdominal pain. Conclusion: Lower abdominal pain with a cystic mass finding on abdominal examination and accompanying hematocolpos after menarche suggests HWWS. Early detection and management can prevent the later complications of these syndrome.

Keywords: Herlyn-Werner-Wunderlich Syndrome, Uterus Didelphys, Hematopyocolpos, Cystocele

1. Introduction

The paramesonephric or Mullerian ducts are an essential aspect of the development of the urogenital system. The Mullerian ducts develop into the uterus, fallopian tubes, cervix, and the upper third of the vagina [1]. Mullerian malformations are abnormalities that are originated during the development of the paramesonephric ducts and are characterized by failures in the fusion of these structures in the middle line when they connect to the urogenital sinus. They occur due to alterations in the formation of the upper vaginal lumen and the uterine lumen, and also because of the non-absorption of the septum in the fusion of the ducts. The forms vary from the very light ones, generally asymptomatic but that can cause serious obstetrical disorders, to the most serious cases such as vaginal and uterine agenesis [2].

2. Literature Survey

Herlyn-Werner-Wunderlich Syndrome or also referred to obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) is a rare congenital anomaly of the female urogenital tract caused by abnormalities in the development of the Mullerian ducts resulting in uterus didelphys, obstructed hemivagina, and associated ipsilateral renal anomaly. The exact cause, pathogenesis and embryologic origin of HWWS are unclear and remain a subject of discussion [3]. This syndrome is regarded as Class III Mullerian dysgenesis and constitutes around 5%-10% of the reported Mullerian duct dysgenesis cases [4]. Renal anomalies range from ectopic ureteric insertion and renal dysplasia to complete ipsilateral renal agenesis [5], [6]. The prevalence of HWWS is unknown. Literature data are ambiguous [7]. Tuna et al. state 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 those with obstructed hemivagina [8]. According to Nishu et al., the frequency is 0.1–3% while Fachin et al. state that it is 5% [9], [10].

3. Problem Definition

Patients with HWWS are usually diagnosed around or after puberty [11]. Clinical manifestations are unspecific and most often include abdominal pain, painful menstruation and a palpable mass in the abdomen, secondary to hematometocolpos from retained menstrual blood [7], [8], [12]. 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 [13]. Ultrasound, magnetic resonance imaging (MRI) and laparoscopy are used to establish the diagnosis of HWWS [7]. The management of HWWS requires thorough and careful anatomic considerations. Surgical reconstruction of the internal genitalia with restoration of normal menstruation and maintenance of a patent genital tract is challenging [12].

4. Approach

A fourteen year old girl came to gynecologic department with her parents complaining abdominal enlargement and lower abdominal pain since 1 month ago. The pain does not radiate and is not associated with fever, nausea, vomiting, diarrhea or micturition problem. The patient also complained of foul smelling brownish vaginal discharge since 2 weeks ago. The patient menarche in the middle of 12 years but the cycles was irregular about every 2 months but without dysmenorrhea. The patient denies any past medical or surgical history. The patient was born atterm pregnancy and without complications. The patient does not have a family history of congenital diseases. The patient is not sexually
active and is not taking contraceptive pills or hormone therapy. The patient belongs to the middle class family.

The patient was afebrile and her vital signs were stable on the day of admission. Signs of secondary sexual growth were well developed. On abdominal examination found no distension was noted, bowel sounds were normal and palpable a cystic mass with tenderness on the right iliac fossa with size 10 cm x 10 cm and smooth surface. From external gynaecology examination was within normal limits. Per rectal examination revealed a cystic mass with a smooth surface in the right lateral region of the midline with proximal border and its origin was difficult to be determined. The results of her other general physical examinations were within normal limit.

The patient’s complete blood count revealed mild anemia with haemoglobin 9.7 g/dL, normal leukocyte count (7.41 x 10^9/µL) and platelet count 448 x 10^9/µL. The patient has been done evaluated by upper and lower abdominal ultrasound. The ultrasound result were found that: the right kidney wasn’t visualized (Figure 1), the left kidney was visualized with size larger than normal with echo parenchyma and cortex thickness configuration were normal but there was dilatation of pelviccalyceal system grade 2 (Figure 2), the double uterus were visualized with two cervical canal which attached together (Figure 3), there weren’t any abnormality visualized in right and left adnexe and there was accumulation of fluid with internal echo was visualized intravaginal with volume about 530.77 ml corresponding to hematocolpos (Figure 4 and 5).

A provisional diagnosis of uterus didelphys, hematocolpos, and agenesis of the right kidney was made. We planned Magnetic Resonance Imaging (MRI) examination for the abdominal and pelvic areas. However, due to MRI limitation in our hospital and insurance issues, we did not do it. We prepared the patient for vaginal excision to evacuation of the accumulation of fluid which causing the lower abdominal pain with informed consent to the patient and her parents about hymenadamage during the procedure. The periooperative examination wasunremarkable and generalanaesthesia was used during the surgery and gave prophylaxis antibiotic with 2 gramecefotaxime intravenous.

5. Results

We performed gynaecology examination under general anesthesia. We found vaginal introitus with an intact hymen and bulging cystic mass with smooth surface on right lateral vaginal wall from vaginal examination (Figure 6). We performed excisionon right lateral vaginal wall and evacuated about 800 ml foul smelling blood containing pus corresponding to hematopyocolpos (Figure 7). Evaluation after excision of the right lateral vaginal wall we found right and left vaginal introitus with their respective cervix and cystocele (Figure 8). We decided performed a right neovaginal reconstruction to maintenance drainage of menstrual blood from right functional uterus. Anterior colporrhaphy is performed to repair anterior vaginal wall to relieve the cystocele (Figure 9).
There were no complications during surgery. The patient’s recovery after operation was very well and after 3 days had been discharged from hospital. One week after operation, patient came for follow-up and reported there was no complained of lower abdominal pain again and did repeated upper and lower abdominal ultrasound. The ultrasound results showed that there was no accumulation fluid visualized intravaginally and the dilatation of the pelvicocalyceal system of the left kidney was reduced. In one month after operation patient reported got menstruation without lower abdominal pain but increasing amount of menstrual blood compare before operation. The amount of menstrual blood were much more than before operation as consequences of there were two functional uterus.

6. Discussion

Female genital malformations arise from any abnormalities at different stages of development of the paramesonephric ducts or the Mullerian ducts. The Mullerian ducts migrate to the midline then fuse to form the uterus, cervix and upper vagina at 8 weeks of pregnancy [14], [15]. Although the Mullerian ducts develop into the uterus, the fallopian tubes, and the upper third of the vagina, anomalies of the fallopian tubes are rarely observed among Müllerian malformations [7].

Herlyn-Werner-Wunderlich Syndrome is a rare form of Mullerian malformations. This syndrome is regarded as Class III Mullerian dysgenesis and constitutes around 5%-10% of the reported Mullerian duct dysgenesis cases [4]. The characteristic triad of this syndrome includes didelphys uterus, obstructed hemivagina, and ipsilateral renal agenesis hence, also known as OHVIRA [16]. Incomplete or non-existent merging will result in two uterus. Renal agenesis usually follows the Mullerian ducts dysgenesis because kidneys, fallopian tubes, cervix and proximal vagina are derived from the same ureteric buds [15]. The reported variations of this syndrome are 72.4% with classical variants of uterus didelphys and 27.6% rare variants with uterus septum or cervical agenesis [14], [17]. The abnormalities of this syndrome is 60% of occurred on the right side [18].

In order to understand the complexities of the HWWS, it is important to review the currently postulated theory of its embryogenesis. The mesonephric ducts on each side pouch out to form the mesonephric diverticulum by the fifth week of pregnancy [19]. Immediately after that, the diverticulum will bud up the ureter which triggers the formation of the kidney by the mesonephric blastema. The paramesonephric
duct begins to develop in the seventh week of pregnancy. The lateral mesonephric duct then migrates medially to it. It has been suggested that the development of the paramesonephric ducts and their position and fusion are highly dependent on the factors released by the mesonephric duct [4]. Subsequent fusion of the mesonephric duct occurs and resorption of the midline septum results in normal uterine formation. Therefore, any developmental abnormality of the Mullerian duct will affect the ipsilateral hemivagina, ureteral budding, and uterine fusion [4].

Figure 9: Post anterior colporraphy

Clinical manifestations of patient with HWWS usually include acute or chronic lower abdominal pain that occurs immediately after menarche due to hematocolpos with the mean age is about 15 years [7], [9]. However, the course of the HWWS can be asymptomatic, which is associated with the normal flow of menstrual blood through patent collateral hemivagina [7]. The other manifestations can present with a pelvic or vaginal mass, abnormal vaginal discharge, acute retention of urine, fever, vomiting, infertility, complicated pregnancy and labor, or endometriosis [9]. Our patient is a 14 years old girl which is quite similar with the mean age of all patient with HWWS. Accumulation a lot of hematocolpos in obstructed right hemivagina we finding like a cystic mass on abdominal examination in our patient. It should be noted that the presence of the obstructed right hemivagina in our patient and the slight outflow of menstrual blood from the right uterine cavity to obstructed right hemivagina, were probably the cause of a asymptomatic period. A long time of blood retention in the blocked vagina could contribute to the development of bacterial infection and hematopyocolpos.

Due to the normal appearance of the external genitalia, this syndrome often remains undiagnosed and asymptomatic in childhood [20]. The diagnosis is determined based on radiological examination. Although MRI usually allows a confirm diagnosis of anomalies but MRI is not as accessible as ultrasound. Therefore, ultrasound usually helps in making the diagnosis, allows detection of blood retention in the hemivagina and/or uterine cavity and determination of the presence of two uterine bodies [7]. So far, this has helped in determining the initial diagnosis in our patient. Based on the morphology of the vagina, HWWS has been classified as class 1 (completely obstructed hemivagina) and class 2 (incompletely obstructed hemivagina). Both classes have two subclasses [21]. Our patient’s case represents subclass 1.1 with uterus didelphys and blind hemivagina (Figure 10). Patient with complete obstruction are more susceptible to complications such as hematometra, hematosalpinx, hemoperitoneum, and even pelviperitonitis as obstruction cause retention of menstrual fluid in the internal female genitalia [10]. In our patient, we only found hematocolpos during ultrasound examination without hematometra and the others complications.

Figure 10: HWWS ’s subclass classification 1.1 with blind hemivagina

The management of HWWS requires careful anatomical considerations. Surgical reconstruction of internal genitalia with restoration of normal menstruation and maintenance of patent genital tract is a challenge. Full excision and marsupialization of the vaginal septum or obstructed vagina are the surgical treatments of choice for uterus didelphys with unilateral vaginal obstruction [12]. In our patient, we performed right lateral vaginal wall excision to evacuated accumulation of hematopyocolpos fluid and continued with right neovagina reconstruction to maintenance patent genital tract for right and left functional uterus and then continued with anterior colporrhaphy because there was cystocele. Those procedure relief of the symptoms in our patient and get menstruation without lower abdominal pain.

7. Conclusion

Lower abdominal pain with a cystic mass finding on abdominal examination and accompanying hematocolpos from ultrasound examination in patient after menarche suggests HWWS as the cause of these symptoms. This finding must be confirmed by MRI to confirm the diagnosis of HWWS (uterus didelphys, hematocolpos in hemivaginaandipsilateral renal agenesis) if possible. Early detection and management can prevent the later complications of these syndrome. The goal of surgery management for HWWS is to relieve the obstruction of the internal genital tract.

References


