Pelvic inflammatory disease due to Herlyn-Werner-Wunderlich syndrome

ABSTRACT

Background: Herlyn-Werner-Wunderlich syndrome is a congenital urogenital malformation associated with a uterus didelphys and a longitudinal vaginal septum, resulting in a blind hemivagina and ipsilateral renal agenesis. Clinical presentation is highly variable, delaying diagnosis and leading to important complications.

Clinical case: We present the case of a 13-year-old female who was diagnosed with Herlyn-Werner-Wunderlich syndrome following acute abdomen due to a right tubo-ovarian abscess. She had a vaginal septum giving rise to a right blind hemivagina. It was microperforated, causing intermittent genital bleeding. This hematocolpos was colonized by microorganisms that ascended into the pelvic cavity, causing a right tubo-ovarian abscess. Nuclear magnetic resonance imaging provided the most diagnostic information. We performed a vaginal septum resection and both hemiuterus communicated with a single vagina, resulting in an asymptomatic patient.

Conclusion: Herlyn-Werner-Wunderlich syndrome is a little known entity and can be presented atypically, resulting in diagnostic difficulty and treatment delay. It is important to be aware of this syndrome in order to avoid irreversible complications.

Key words: Herlyn-Wener-Wunderlich, OHVIRA, hematocolpos.
BACKGROUND

Herlyn-Werner-Wunderlich syndrome (HWWS) is a rare urogenital malformation caused by alterations both in lateral and vertical fusion of the Müllerian ducts. It consists of a combination of didelphys uterus with a longitudinal vaginal septum that forms a blind hemivagina and ipsilateral renal agenesis to the vaginal obstruction. Because of this it is also called OH-VIRA syndrome (Obstructed HemiVagina and Ipsilateral Renal Anomaly). There are different forms of clinical presentation described, although the most common presentation includes nonspecific symptoms such as dysmenorrhea or abnormal genital bleeding in adolescent females, which occurs soon after menarche. We report a case with atypical presentation in a 13-year-old female.

CLINICAL CASE

We report the case of a 13-year-old female who presented to the Emergency Department at our hospital due to abdominal pain and fever. There was no clinically significant past history, only occasional intermenstrual “spotting” shortly after menarche. During physical examination a pelvic tumor in the right iliac fossa was noted, painful to palpation and with bulging of the right lateral face of the vagina. Abdominal and transrectal ultrasound demonstrated right renal agenesis with suspected didelphys uterus and a 54-mm heterogeneous cystic pelvic tumor without vascularization that appeared to arise from the right adnexa, suggestive of an endometrioma. Blood count reported leukocytosis and increased C-reactive protein. With the possibility of a diagnosis of acute abdomen, a diagnostic/therapeutic laparoscopy was performed, demonstrating an abscess adjacent to the right horn along with signs of pelvic inflammatory disease. The adhesions were freed and abscess of the drainage was carried out with clear symptomatic improvement in the immediate postoperative period.

DISCUSSION

According to Smith and Laufer, HWWS was described for the first time in 1922 and in the decade of the 1960 the first cases began to be reported; however, in the last 20 years the figure has increased considerably due to the diagnosis being supported with imaging tests.

During outpatient follow-up, the patient did not report abdominal pain but did state she continued to have “intermenstrual spotting.” On physical examination and transrectal ultrasound, bulging on the right vaginal face and image of a pelvic cystic mass on the right iliac fossa continued to be visualized. However, both ovaries had normal morphology. Nuclear magnetic resonance (NMR) reported right renal agenesis with didelphys uterus and a fluid collection suggestive of hematocolpos in the right hemivagina of 50 mm (Figure 1). The patient was operated on after the diagnosis of HWWS. An exhaustive vaginal examination was carried out under anesthesia and an opening on the right lateral face was seen where prior bloody material was draining. The complete vaginal septum was resected with a bipolar resectoscope and both cervices were connected so as to leave only one vagina (Figure 2). At present, the patient remains asymptomatic.

Figure 1. Pelvic nuclear magnetic resonance where the following are visualized. A) Double uterus and the communication of each hemiuterus with each cervix (arrows) and in B) the hyperintense blood collection in the right hemivagina (arrow), causing bulging in the free vagina.
The etiology and pathogenesis of this syndrome are unknown although it is believed that the embryological disorder is produced during the eighth week of gestation and that its origin is multifactorial. Lack of development of one of the mesonephric or Wolffian ducts causes a renal agenesis on that side and, in turn, induces lateral displacement of the ipsilateral Müllerian duct, preventing its contact and fusion with the contralateral Müller duct, thereby developing a double uterus. This hemiuterus will not be in contact adequately with the proximal portion of the urogenital sinus and will form a blind hemivagina, with alteration of the lateral and vertical fusion of the Müllerian ducts that predominate on the right side (Figure 3).

The actual incidence of this disease is unknown and perhaps is underdiagnosed because some cases are asymptomatic. Some series report figures of between 0.1 and 3.8% of the general population, being slightly greater in Finland for still unknown reasons.

The majority of cases are described in young women at the beginning of the menstrual cycle; the median age at diagnosis is 14 years. The most common clinical manifestation is chronic pelvic pain with regular menstrual cycles due to

Figure 2. A) Vaginoscopy and localization of the longitudinal vaginal septum (black arrow). B) Excision of the septum (white arrow) using a resectoscope with bipolar current. C) Surgical specimen for histological study.

Figure 3. Herlyn-Werner-Wunderlich syndrome. Double uterus, longitudinal vaginal septum that form a blind hemivagina and ipsilateral renal agenesis.
the chronic accumulation of blood in the blind hemivagina and the ipsilateral hemiuterus. If the diagnosis is delayed, hematocolpos-hematometra may cause compressive symptoms in the neighboring organs such as urine retention and pyelocalyceal dilatation.\(^5\)

The obstructed vaginal wall may be perforated and cause intermittent abnormal vaginal bleeding and could lead to colonization and the rise of microorganisms ascending to the internal genital apparatus, which give rise to pyocolpos with pelvic inflammatory disease in women who do not engage in sexual relationships, such as in the case of our patient.\(^6,7\) It is important to carefully examine the vagina to observe the opening at the wall level where the hematocolpos bulges and be able to partially drain it.

Due to the wide variety of clinical manifestations with which this syndrome may occur and because it is a rare condition and correct diagnosis in the majority of cases is delayed, the following complications may arise: compressive symptoms, altered renal function, acute abdomen due to tubo-ovarian abscesses, etc. Also, the most common symptoms are very nonspecific such as chronic abdominal pain or dysmenorrhea that they are treated empirically in most cases before other imaging tests are performed.

The continuous retrograde reflux of endometrial material towards the internal genital apparatus causes endometriosis, causing chronic abdominal pain, adhesions and sterility.\(^3,8\) For this reason it is necessary that this syndrome be recognized. When there is diagnostic suspicion, physical examination should be performed using procedures such as NMR so as to carry out an early diagnosis and thereby avoid complications.

Complementary tests that are most cost effective for the diagnosis of this syndrome are ultrasound and NMR. The first is an accessible, safe technique that easily evaluates the renal system. Transvaginal study evaluates the uterine and adnexal morphology with high precision. Nevertheless, it is not as precise in the evaluation of the soft tissues as NMR, which is considered to be the most sensitive test for this syndrome.\(^8\)

Laparoscopic inspection is the only technique capable of assessing possible pelvic complications arising from this disorder (endometriosis, adhesions) and, for some authors, it is considered to be the most cost-effective test for the diagnosis of the HWWS.\(^3\) However, it should be reserved for cases where the diagnosis is unclear with the rest of the imaging tests.\(^1\)

Treatment should be carried out as early as possible to avoid complications and consists of resection of the vaginal septum and drainage of hematocolpos and hematometra, communicating both uterine necks with one vagina, which can be performed with a vaginal approach or with vaginoscopy using a resectoscope.\(^3\)

Hemi-hysterectomy with or without salpingectomy should be avoided, with the intent of improving the patient’s fertility. Pregnancy rates are similar to that of patients without this disorder. However, there is a greater rate of obstetrical complications because of the double uterus, such as preterm labor or intrauterine growth delay.\(^4\)

Cases of adenosis in the histological study of removal of the vaginal wall have been described and the relation with cervical cancer is not yet clear. However, some authors recommend annual Pap smears and serial colposcopic studies.\(^4,9\)

In conclusion, HWWS is a little known disorder that can cause severe sequelae in patients if diagnosis is delayed. Therefore, it is fundamental to maintain a high diagnostic suspicion and to carry out an exhaustive physical examination to be able to request the necessary imaging tests and thereby perform timely surgical treatment.
REFERENCES


