Herlyn-Werner-Wunderlich syndrome - a case report

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Summary

This is a case report of Herlyn-Werner-Wunderlich syndrome in a 28-year-old patient. She was admitted to hospital for surgical treatment of the pelvic mass accompanied by painful menstruation periods. This syndrome was diagnosed by US and MRI and it was treated by hemi-hysterectomy with vaginectomy. After the surgery, the patient has had regular and painless menstruation.

Key words: Müllerian duct anomalies; Hemivagina; Hematocoplos; Ipsilateral renal agenesis.

Introduction

Herlyn-Werner-Wunderlich (HWW) syndrome is a rare congenital anomaly of the female genital tract characterized by obstructed hemivagina, hematocolpos, ipsilateral renal anomaly, and uterine anomaly, more commonly uterus didelphys than uterus septus. Recently, this syndrome has been referred to as OHVIRA syndrome [1-3]. We still cannot accurately define the frequency of HWW syndrome. The incidence of obstructive Müllerian duct anomalies is 0.1-3.8% [4], 2-3% [5]. Isolated anomaly, like uterus didelphys is present in 1/2,000 to 1/28,000 cases, and ipsilateral renal agenesis is present in approximately 43% cases [6]. Presence of a pelvic mass in association with pelvic pain after menarche points to this syndrome even in women with regular menstrual periods [7]. Ipsilateral renal agenesis with pelvic mass should imply that this syndrome is to be seriously considered. The age of women with this syndrome ranges from 10 to 29 years (mean age: 14) [2]. Magnetic resonance imaging (MRI) will help render an accurate diagnosis, whereby the early endoscopic resection of the septum can be the final treatment followed by good and long-term outcome and preserved fertility [8].

Case Report

We report the care of a 28-year-old patient with HWW syndrome who was admitted to hospital for surgery due to a pelvic mass and severely painful menstrual periods. Six months prior to hospitalization, the patient had undergone laparoscopic hysterectomy because of an endometriotic cyst on the left ovary. During the laparoscopic treatment, the uterus duplex and pelvic mass were visualized below the right uterus. The patient had menarche at the age of 14. Menstrual cycles were regular, every 30 ± 4 days and severely painful. Upon being admitted to hospital, she was examined rectally as she was virgo intacta. Soft tumefaction was palpable below the uterus on the right side, parallel to the vaginal wall, and ellipsoid in shape. The shape of the uterus was irregular, slightly enlarged. The adnexal findings aroused no suspicion. Blood and urine laboratory analyses were within the reference ranges. Transabdominal ultrasound revealed absence of the left kidney, the existence of two normal size uteri, with blood retention in the right one and with pelvic cyst 60 x80 mm filled with hyperechogen content. Each uterus had one adequately developed ovary of normal size. In order to confirm the ultrasound diagnosis, i.e., the existence of a congenital urinary tract anomaly, intravenous pyelography was performed. It revealed that the left kidney filtered and concentrated the contrast; the collecting system functioned properly, its morphology being normal; a proximal ureter of normal diameter; bladder contours not well demarked from the uterus. The right kidney did not filter the contrast. MRI was done and it showed that there were two uteri of normal size with blood retention in the right one. Each uterus had communication with the related hemivagina. There was a cyst formation (60 x 80 mm) between the bladder and rectum filled with fluid, while the signal intensity implied dilated blood-filled right hemivagina obstructed by the transverse septum. Each uterus had well-developed adnexa. Given the imaging findings and clinical history, the diagnosis was Herlyn-Werner-Wunderlich syndrome. Preoperative preparations being completed, the patient underwent surgery. Hemihysterectomy with vaginectomy was performed because of the position of two hemivaginas, since drainage of a blind vagina and ipsolateral uterus would not be possible by simple septum resection. The operative field was prepared and abdomen was opened by a low transverse Pfannenstiel incision. The genital organs appeared, two uteri and adnexa in each uterus (Figure 1). Once the Lig ovarii proprium and Lig rotundum were clamped, they were ligated and cut while the right adnexa was conserved. Peritoneum and plica vesicovaginalis were dissected first sharply, then bluntly and the bladder was retracted towards the vagina. The hemiuterus was cut by thermocautery after ligation of uterine arteries and the uterus had been clamped, ligated, and cut from the right side. The right hemivagina was opened and chocolate-colored fluid was drained from a 4 cm long blind vagina (Figure 2). The right hemivagina was sharply divided from the paracolpium and removed together with the hemiuterus (Figure 3). Hemostasis was checked and the right adnexa was fixed to the left hemiuterus (Figure 4). Postoperative treatment was uneventful. The patient was discharged from hospital five days after surgery in good condition. Her menstrual periods have been regular and painless.

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Fig. 1

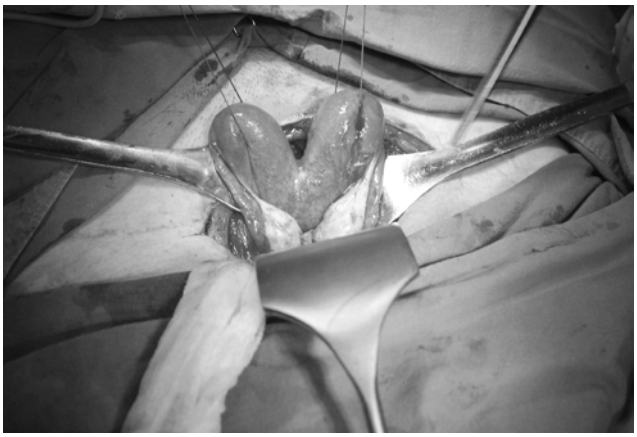

Fig. 2

Fig. 3

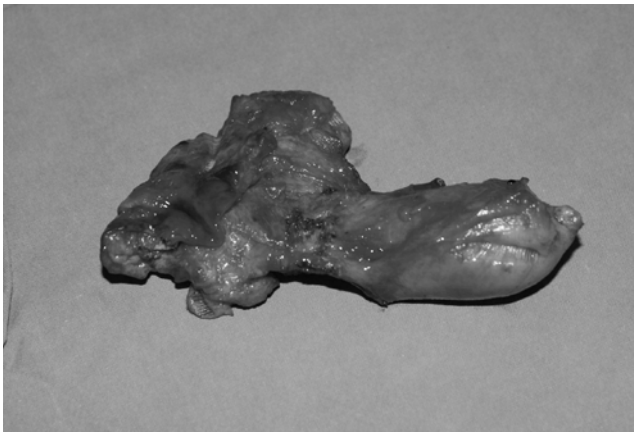

Fig. 4

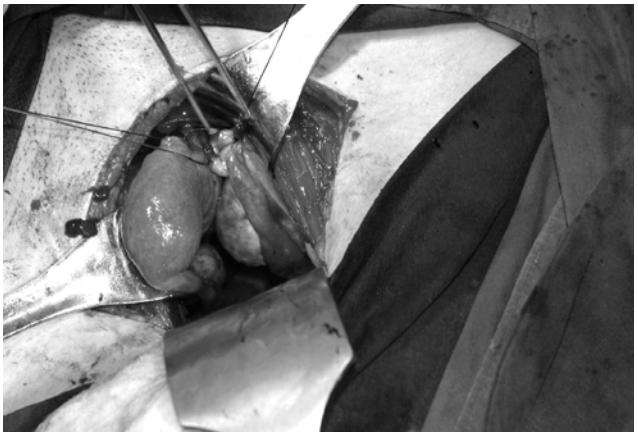

Figure 1. — Uterus didelphys.
Figure 2. — Blind hemivagina opened and haematocolpos.
Figure 3. — Hemiuterus and blind vagina are removed.
Figure 4. — Hemiuterus and both adnexa.

Discussion

HWW syndrome is a rare anomaly manifested immediately after menarche. This syndrome is a result of developmental disorders of genital organs in the period from the 6th to 17th gestational week. The cause is unknown. It is difficult to precisely determine the incidence of the syndrome. The incidence of congenital anomalies of the Müllerian duct is about 2-3% [5]. Congenital Müllerian duct anomalies result from nondevelopment or nonfusion of Müllerian ducts. These abnormalities include double uterus, uterus didelphys, uterus bicornuate, and uterus septus. Uterus didelphys represents a complete duplication of the uterus in two separate horns, two cervices and two vaginas. Uterus didelphys with obstructed hemivagina is one of the rarest congenital anomalies of the Müllerian ducts, occurring between the 12th and 16th gestational week. It is the result of lateral fusion defects of caudal Müllerian ducts, and includes abnormalities caused by failed septum resorption after fusion of the ducts. This anomaly is accompanied by renal agenesis on the side of the obstructed hemivagina. Renal agenesis occurs due to the failure of the uteric bud to differentiate from the mesonephric duct. As two-thirds of the upper vagina develop from Müllerian ducts, defects in fusion of the ducts can lead to vagina duplex, and failed wall resorption between the ducts causes vagina septum. This condition is often associated with uterus didelphys or uterus septus [9]. Uterus didelphys is most frequently accompanied by transverse vaginal septum [10]. Transverse vaginal septum can be complete or incomplete and is not usually associated with other urologic and Müllerian anomalies [4]. We have reported a 28-year old patient with HWW syndrome admitted to hospital for surgery because of a pelvic mass and severely painful menstrual periods. Genital tract anomalies often occur in association with urinary tract anomalies. Abdominal ultrasound evaluation can easily reveal the absence of a kidney, accompanied by functional hypertrophy of the other kidney due to which laboratory analyses of renal functions remain within the reference ranges. It is important to make a timely diagnosis of the syndrome. This diagnosis must be suspected in girls complaining of dysmenorrhea and recurrent pains in the lower abdomen between the periods and subsequent pelvic mass. Accurate and final diagnosis can be rendered not only by vaginal examination but must be accompanied by ultrasound and MRI, or by laparoscopy and hysteroscopy.

MRI is a very efficient, non-invasive diagnostic method that can show anatomic visualization of the organs at all levels and help differentiate myometrium from fibrous septum. MRI helps render an accurate diagnosis and determination of the follow-up treatment [11, 12]. HWW syndrome is to be suspected in young girls complaining of painful menstrual periods and having pelvic and paravaginal masses. Diagnosis is usually rendered after menarche. All the symptoms for this syndrome, like dysmenorrhea, pelvic mass, and hematocolpos are even more obvious after menarche [13]. This syndrome is diagnosed in patients at the age of 20 to 30. HWW syndrome has been found in 12 patients (mean age: 13 years). Pelvic mass and abdominal pain were present in 11 of the girls out of 12, whereas four of them had dysmenorrhea. Pain lasted for 0.5-12 months [1]. We have presented a case of uterus didelphys with obstructed hemivagina and pyocolpos and ipsilateral renal agenesis where the final diagnosis was postponed till pregnancy [12]. Symptoms of this syndrome have already been described in girls even prior to menarche. One patient with renal agenesis and microscopic hemathuria had these symptoms before menarche [14]. All the characteristics of this syndrome were described in a four-year old girl [8]. Ipsilateral renal genesis accompanied by a pelvic mass is always a step further in diagnosing this syndrome. Transvaginal ultrasound (TVUS), especially 3D TVUS is important in diagnosing this syndrome. There was a case of incomplete syndrome with asymmetric obstructed uterus didelphys diagnosed by 3D ultrasound [11]. HWW syndrome is not an anomaly resulting in female sterility. It is a syndrome found in women with primary infertility, although rarely [15]. Uterus didelphys, obstructed right hemivagina, pyocolpos and ipsilateral renal agenesis were found in a 25-year old patient suffering from infertility with dysmenorrhea when examined by ultrasound and MRI. Vaginal septum excision and drainage were performed. The patient got pregnant spontaneously three months after surgery [3]. According to the literature, 23% of all the patients suffer spontaneous miscarriages, 15% of them undergo pre-term delivery, and 62% of them undergo full-term deliveries successfully [16]. If septum of the obstructed vagina is timely recognized and surgically removed and drained, it can contribute to the disappearance of symptoms and prevention of complications relating to chronic cryptomenorrhea, like endometriosis, pelvic adhesion, and infectious pyocolpos [1, 17]. The aim of the surgery is to preserve normal fertility. Treatment of these anomalies includes hematocolpos drainage and vagina septum excision which is traditionally done by scalpel or scissors. A less invasive procedure is hysteroscopic resection of the vagina septum which improves visualization [18]. Surgical removal of the septum of the obstructed vagina with drainage is the adequate treatment of the syndrome [1, 17] with good long-term results and preserved fertility [8]. Tracheobronchial stent is inserted in patients with bilateral hemivagina and uterus didelphys, hematometra, and hematocolpos after vaginal septum excision to preserve communication. Six months later the stent was removed. Twelve months later the patient had regular menstrual periods which were painless [19]. Out of 12 patients with HWW syndrome undergoing vaginal septectomy, 11 had no more symptoms, and one patient had irregular menstrual periods [1]. There was one case of a minimally invasive combination of laparoscopy and vaginoscopy in a ten-year old girl. After laparoscopic incision of the horn of the uterus through its cavity and cervical canal, a Maryland dissector was introduced – a hole was made on the septum of the obstructed hemivagina and the dissector placed in the vaginal canal; a Penrose drain was inserted from the vagina, through the cervical canal into the uterine cavity up to the horn. Incision was made on the ipsilateral Fallopian tube and the drain was inserted [20].

Conclusion

Combined laparoscopy and hysteroscopy represent the best approach leading to accurate diagnosis and adequate treatment of the syndrome.

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