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REVIEW PAPER

Vomiting, constipation, and abdominal pain during menstruation as first symptoms of Herlyn-Werner-Wunderlich syndrome: case report and literature review

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ABSTRACT

Herlyn-Werner-Wunderlich syndrome, also known as OHVIRA (Obstructed Hemivagina and Ipsilateral Renal Anomaly), is an extremely rare abnormality of the Müllerian ducts, characterized by the presence of double uterus, unilateral cervico-vaginal obstruction and renal agenesis and/or other urinary tract anomalies. Most cases are diagnosed after menarche through an MRI, CT or ultrasound. There are no specific symptoms of this syndrome. Patients presented fever and vomiting during menstruation, intermenstrual bleeding and palpable abdominal, pelvic or vaginal mass. Thorough history taking is extremely important as common symptoms in pediatrics (vomiting, constipation, abdominal pain) may be a part of a more complex condition, not only within the GI tract but also including other systems.

KEY WORDS:

constipation, children, didelphys uterus, hematometrocolpos, renal agenesis.

INTRODUCTION

Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome (also known as Herlyn-Werner-Wunderlich syndrome) is a rare condition with very few cases reported. The abbreviation OHVIRA stands for obstructed hemivagina with ipsilateral renal agenesis. The third component of this disorder is didelphys uterus. The syndrome is the result of an impaired fusion of the Müllerian ducts. This fusion takes place during embryogenesis, between the 6th and 9th week after conception. The malformation is usually diagnosed after menarche [1].

Herlyn-Werner-Wunderlich syndrome is extremely rare. However, vomiting and constipation are relatively common in the pediatric population and usually indicate gastrointestinal (GI) tract disease. The case presented here was reported to emphasize that such widespread symptoms can be a part of a more complex condition that may occur within the GI tract, but also may include other systems. A short literature review was prepared to complement our report.

CASE REPORT

A 12-year-old girl was admitted to the Department of Pediatric Gastroenterology and Nutrition because of constipation (her last defecation was observed 2 days before admission with a very small amount of stool), dehydration, and abdominal pain. The patient reported pain in the left lower quadrant. It was still, non-radiating, and with no exacerbating factors. The pain began 2 days before admission.

On her first day in the hospital, the patient vomited (6 times) and refused to eat and drink because of the pain.

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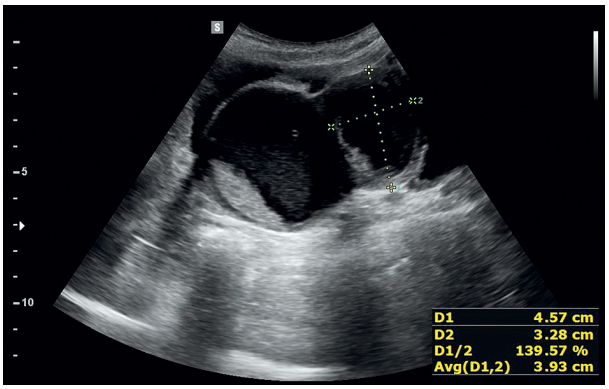

FIGURE 1. Ultrasonography of cystic lesions localized in the lesser pelvis

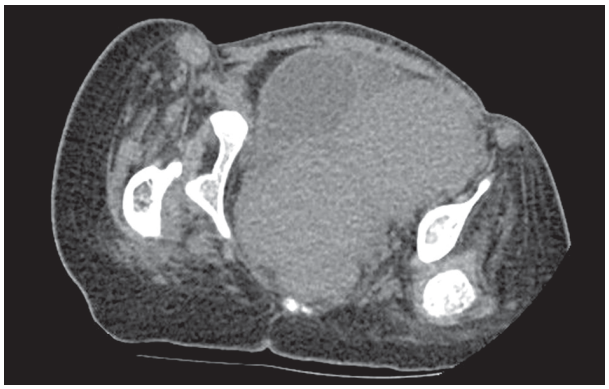

FIGURE 2. Contrast CT presenting a double-bodied uterus with hematometrocolpos on the left side

The patient was experiencing menstruation (which started 2 days before admission and was the third menstruation cycle in the patient's life). A gradual reduction in menstrual bleeding was observed. Physical examination revealed a palpable mass in the left iliac fossa. This was suspected to be a mass of stool. The patient was previously hospitalized because of myelomeningocele and hydrocephalus. She has a ventriculoperitoneal shunt, scoliosis, kyphosis, left kidney agenesis, and neurogenic bladder dysfunction. She is also underweight (below 1 percentile for her age group based on the growth chart).

The ultrasonography performed at the Emergency Department revealed numerous cystic lesions localized in the lesser pelvis, some of which were filled with hyperechogenic content (Figure 1). It also showed the presence of dense content in the uterine cavity and some fluid in the peritoneal cavity. Evaluation of the abdomen was difficult because of the peculiar anatomical conditions, thus CT scan was scheduled. Contrast CT was performed and showed a double-bodied uterus. The right side of the uterus had no pathologies, while hematometrocolpos was present in the left side of the uterus (Figure 2). CT scan also revealed fluid in the peritoneal cavity, ventriculoperitoneal drainage, ovarian cysts in the left ovary (the biggest measuring 4 cm), agenesis of the left kidney, and multiple skeletal deformations.

An enema was used to relieve constipation (for 3 days), laxatives were changed from lactulose to macrogols, and the patient was discharged home. A visit to the Gynecological Outpatient facility was scheduled. Gynecological consultation resulted in the initiation of continuous oral contraceptives (microgynon). Surgical treatment was also scheduled. Surgical treatment of the syndrome usually consists of opening the blind hemivagina and performing fluid drainage. Two weeks after the gynecological consultation (but still prior to surgery), the patient returned to the Department of Pediatric Gastroenterology and Nutrition with acute abdominal pain in the same area as previously observed. She was transported to the Department of Gynecology for urgent surgery.

DISCUSSION

The first case of a patient with OHVIRA syndrome was described in 1950 [3]. A more in-depth description of this disease was conceived more than 20 years later by Herlyn and Werner, and subsequently expanded upon by Wunderlich in 1976 [4, 5]. On this basis, the syndrome was given its second name, Herlyn-Werner-Wunderlich syndrome.

The etiology of OHVIRA syndrome is not clear. However, it is hypothesized to be the result of the failed fusion of the Müllerian ducts during embryogenesis (8th week). These ducts develop to form female reproductive organs. The newest theory claims that the maldevelopment of the Müllerian ducts can also result in the absence of the kidney and the ureter [1, 2]. There hasn't been found any genetic mutation or karyotype abnormality predisposing to the development of the syndrome.

The incidence of congenital uterine anomalies is estimated to be 4.3-6.7%. OHVIRA accounts for 0.1-3.8% of congenital uterine anomalies and is the most rare type [2, 6]. In the vast majority of cases, OHVIRA syndrome is diagnosed in young, adolescent girls after the first menstrual bleeding [7, 8]. An analysis of 27 cases revealed that the median age at diagnosis was 14 years old [9]. It has been previously reported that this syndrome can also be diagnosed during pregnancy, birth process [2, 10, 11, 12], or incidentally during diagnostic imaging for other reasons [13, 14].

Obstructed hemivagina and ipsilateral renal anomaly syndrome, beyond the typical presentation as intermenstrual bleeding, abdominal pain, pelvic or abdominal mass, and renal agenesis, can include symptoms from multiple organs [1]. Additional symptoms described in the literature include acute abdomen symptoms [15], ectopic ureter [8, 16, 17], pyocolpos [2, 18], skeletal anomaly (ectrodactyly [19], kyphoscoliosis [20]) and wandering spleen [21]. In our case, constipation and vomiting were an unusual first presentation.

Ultrasonography is the predominant first-choice imaging method. It is very helpful in the detection of hematocolpos, but this method cannot identify the type of Müllerian duct anomaly [22]. The advantages of ultrasonography include availability, cost, and the absence of radiation. However, ultrasonography has a high operator dependency and such rare malformations as OHVIRA syndrome may go undiagnosed. The best diagnostic choice for ultrasound access is transvaginal. Given the fact that in most cases the symptoms occur in childhood and early puberty, transvaginal ultrasonography cannot be performed before the beginning of sexual activity.

Magnetic resonance imaging (MRI) is the gold standard for diagnosing Müllerian duct anomaly (MDA) and allows for the identification of the specific type of MDA [22]. MRI is most useful in cases where differentiation of the uterine anomaly is not easily achievable by ultrasonography or physical exam. This method is characterized by higher sensitivity in the detection of the uterine morphology and the continuity of the vagina when compared to ultrasonography [15]. Moreover, MRI is the best method to identify associated pathologies (endometriosis, pelvic adhesions, other viscera anomalies) [22].

Laparoscopy should be considered when MRI imaging is not available or ultrasonography and MRI diagnosis is not clear. Indications for laparoscopy include: interval between menarche and diagnosis, severity of the symptoms, presence of hematometra or pyometra [22].

An additional advantage of laparoscopy is that it can be used for therapeutic purposes, such as drainage of hematocolpos/hematometrocolpos, septectomy, or marsupialization of the blind hemivagina [22].

The treatment of choice for OHVIRA syndrome is single-stage vaginoplasty. In rare cases, such as extreme proximal vaginal septum or infectious complications, hemihysterectomy may be an alternative [23]. In recent years, there has been a trend towards publications on hymen-sparing surgeries. Sleiman describes safe and successful laparoscopy-assisted resection for the management of high vaginal septum while maintaining an intact hymen [24]. Another hymen-sparing management was demonstrated recently, namely the transrectal ultrasound (TRUS)-guided vaginoscopic septoplasty supported by pre- and postoperative diagnostics with the use of a novel ultrasound technique: three-dimensional saline-solution infusion contrast sonovaginocervicography (3D-SVC) with virtual speculoscopy [25].

Due to the development of diagnostics and treatment methods, patients can manage to avoid complications such as infertility. Ugurlucan et al. reported the follow-up after surgical treatments in which 7 cases out of 32 (21.9%) had married and 5 of these (71.4%) were pregnant or had delivered [23]. After hysteroscopy, patients achieve a close to normal prognosis for the outcome of their pregnancies (75% term delivery rates and 85% live birth rates) [26]. Successful pregnancy outcomes in OHVIRA syndrome are estimated at 57%–68% [2].

In our case in differential diagnosis must be consider infectious diseases of the digestive tract, functional constipation, irritable bowel syndrome, gynecological tumors, Mayer-Rokitansky-Kuster-Hauser syndrome (MRKH) or another MDA.

The present case shows that differential diagnosis is not always obvious and that a common list of manifestations can also indicate a rare disease. Thanks to solid and accurate history taking, unique disorders may be overlooked less frequently.

DISCLOSURE

The authors declare no conflict of interest.

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