Case Report

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A Rare Cause of Abdominal Pain: Herlyn-Werner-Wunderlich Syndrome

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Abstract

Herlyn-Werner-Wunderlich (HWW) syndrome is a congenital urogenital syndrome. Typical features are unilateral blind-ending hemivagina with uterine didelphy and ipsilateral renal agenesis. Patients usually progress asymptomatically until menarche, and they are diagnosed with radiological imaging by applying to the hospital with these conditions such as abdominal pain, abdominal bloating, primary amenorrhea along with menarche. In this case report, it is aimed to present a patient who applied to our emergency department with abdominal pain and is diagnosed with HWW syndrome. In this case, it is emphasized that emergency physicians should regard to HWW syndrome in the differential diagnosis of abdominal pain.

Keywords: Herlyn-werner-wunderlich, müllerian anomaly, abdominal pain

Introduction

Abdominal pain constitutes an important part of emergency admissions. It has been observed that 40% of these not complying with the known symptoms and concluded any clear and understandable outcome and come up with a spontaneous sight (1). It is very difficult to determine the prevalence and incidence of uterine anomalies due to the use of different classifications, diagnosis with different methods and the absence of symptoms of many anomalies. On the other hand, in all studies conducted between 1950 and 2007, it was determined as 6.7% in the general population and 7.3% in the infertile population and women with recurrent pregnancy loss, this rate varies between 3% and 25% (2). No matter how the exact factors that caused these anomalies are not clear, it has been determined in studies that ionizing radiation, infective processes and some medicine (Diethylstilbestrol (DES), Thalidomide, etc.) exposed during the genital development process causing Müllerian anomalies (3).

Herlyn-Werner-Wunderlich (HWW) syndrome is among the uterine anomalies and important for early diagnosis of complications and permanency of fertility. Most of the patients consult with these complaints such as abdominal pain, abdominal mass, bloating, inability to menstruate and acute abdominal pain(4). Dysmenorrhea is the main symptom of HWW syndrome and it is usually established after puberty. The increase in pain is related to a rise in the volume of hematocolpus caused by an obstructed hemivagina. Endometriosis, inflammation, twisted cysts, and appendicitis are other differential causes of pelvic pain and these must be excluded. Although it is possible to interfere diagnoses with similar symptoms and there is not a typical examination finding, differential diagnosis can be easily made with scanning methods. Observing unilateral blind-ending hemivagina and uterus didelphy and ipsilateral renal agenesis on computed tomography (CT) or ultrasonography (USG) imaging performed on the patient is important for diagnosis(5). Making diagnosis in time makes it possible to prevent possible complications with further examination, treatment and surgical procedures depending on the condition of the anomaly (6).

In this case, it is aimed to present the HWW Syndrome that emergency physicians detect in a 19-year-old female patient who applied to the emergency department with the abdominal pain.

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A 19-year-old female patient has consulted to the emergency department with the abdominal pain. On admission to the

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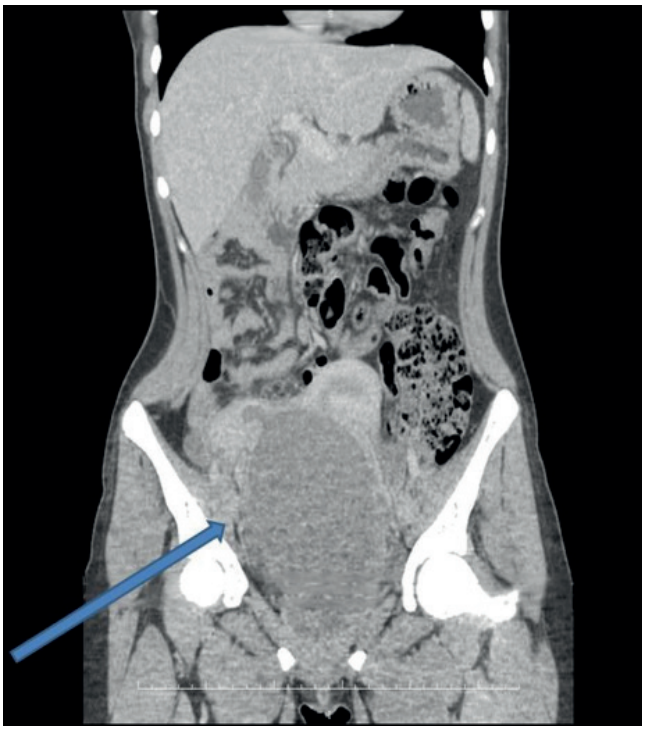

Figure 1.

emergency service, it has been observed that the general health of the patient is good and conscious. First vital signs; blood pressure: 115/75 mmHg, saturation: 96%, heart rate: 88 beats/min, fever: 36.6oC.In physical examination; there is tenderness and defense in the right inguinal and hypogastric area of the abdomen. Advanced vaginal examination could not made to the patient in the emergency room, but it is learned that she has the virginity in this statement. No significant finding is detected in the patient's other systemic examination. In her background, it is learned that the patient has occasional abdominal pain, but not having consistent dysuria or dysmenorrhea. The patient has not applied to any health institution with this complaint beforehand.

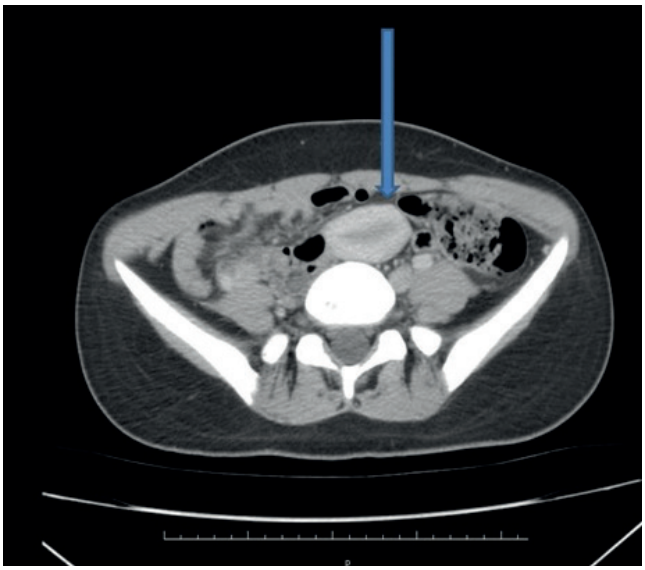

Figure 2.

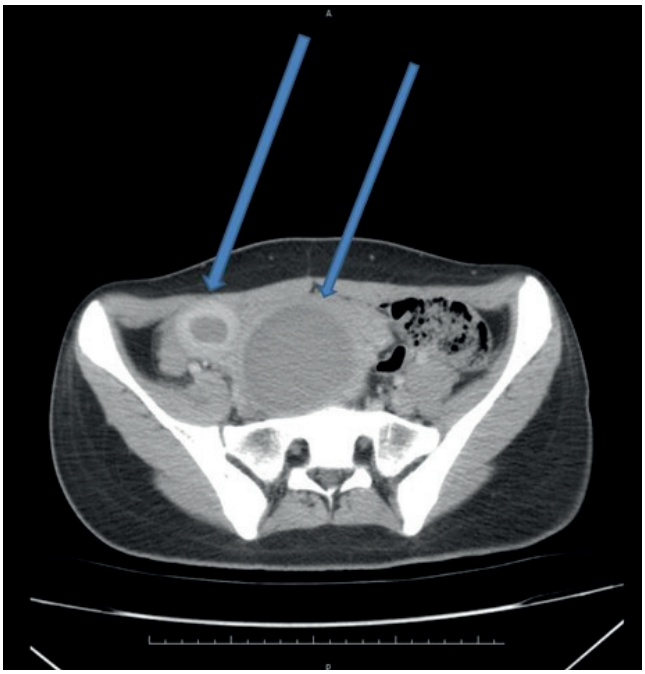

Figure 3.

By closing the oral intake of the patient in the emergency room, IV hydration is started, and necessary laboratory tests are requested. In the test results; b-Hcg<2 mIU/mL. White blood cell (WBC): 11360/mL C-reactive protein (CRP): <0.5 mg/dL and urinalysis: No leukocytes are observed. Little bit of erythrocyte is observed. The patient who has constant complaints, abdominal USG is taken for advanced diagnostic purposes. On USG, edematous didelphis uterus and concominant hematocolpos are observed. As the level of the cecum and ileocecal valve pushed to the left lower quadrant of the abdomen and renal pathologies could not be evaluated clearly, abdominal CT is performed for definitive diagnosis. On abdominal CT, a compatible sight is observed with hematocolpos, which fills the pelvis associated with right renal agenesis, uterine didelpys and right uterine cavity.

The patient who consulted with abdominal pain is diagnosed with HWW syndrome in the emergency room. Afterwards, she is discharged from the service with recommendation of outpatient control. The patient is transferred to obstetrics and gynecology service for further examination-treatment and follow- up process.

Horizontal and vertical one-centimeter incision is made in the hymen of the patient, who evaluated by the department of obstetrics and gynecology. Approximately 200 cc of old bleeding material is aspirated. Afterwards, the patient who followed up for the service is discharged next day and the outpatient clinic follow-up has started.

Discussion

Herlyn-Werner-Wunderlich (HWW) syndrome is a congenital malformation among Müllerian anomalies. In this

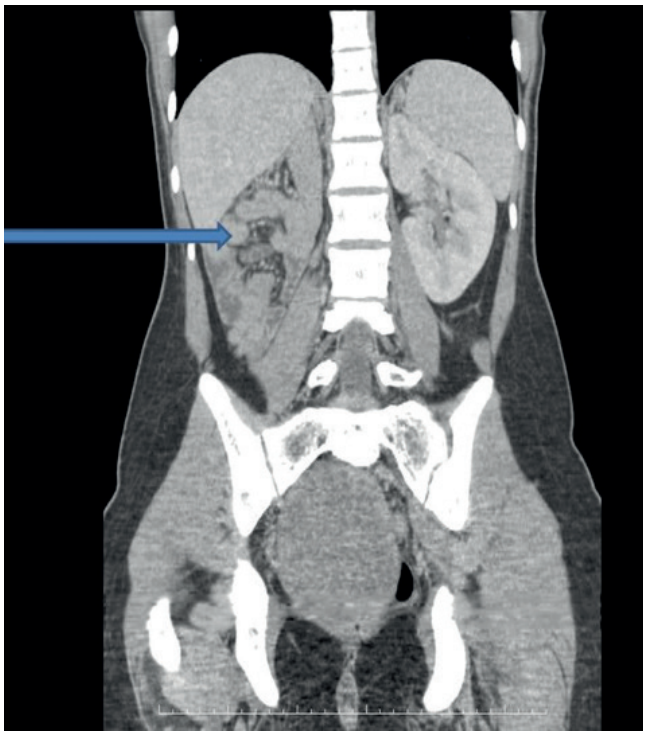

Figure 4.

syndrome, which generally progresses asymptomatically until puberty, it can be diagnosed rarely before puberty, and in some patients in the neonatal period with the appearance of a distended and protruding hemivaginal mass due to the effect of maternal estrogen (7). Symptoms of hematocolpos distension starting with puberty and menarche, bloating and abdominal pain varying from patient to patient depending on the degree of distension can be seen. There are many clinical manifestations that may cause infertility, such as fever, pyohematocolpos, peritonitis, which may occur with accompanying infective processes, and, albeit rare, urinary system stenosis and endometriosis (8-9).

In the developmental pathophysiology of the anomaly, the Müllerian canal shows migration to the midline at the 8th week of pregnancy and then there is a defect in its union with the uterus, cervix and superior part of the vagina (10). The incompleteness of this union causes the formation of two uteruses. The kidneys, fallopian tubes, cervix, and proximal vagina all develop from the same ureteral bud. Therefore, renal agenesis occurs following Müllerian duct dysgenesis (10).

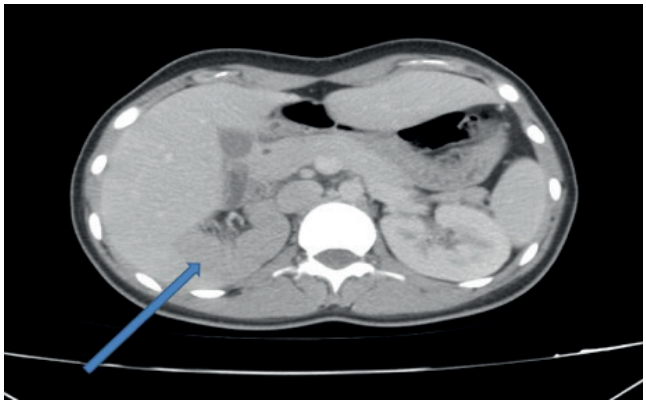

Figure 5.

Scanning methods play an important role during the diagnosis process. It can be diagnosed by visualizing genitourinary anomalies, uterine didelphis, bicornuate uterus, vaginal septum, renal agenesis, hematocolpos, free fluid in the pelvis in USG, CT, hysterosalphingography methods. However, magnetic resonance imaging (MRI) is accepted as the high standard in the diagnosis of Müllerian anomalies (6). Laparoscopy can be performed as an auxiliary intervention in the diagnosis and treatment when other imaging methods are insufficient (11). In treatment, vaginal septum resection can be applied to prevent obstruction, and unilateral hysterectomy can be applied in recurrent stenosis (12-13).

Conclusion

The patient, who presented to the emergency department with abdominal pain, is diagnosed with HWW syndrome as a result of the examinations. HWW is a rare disease caused by Mullerian canal anomaly. Emergency physicians should also keep in mind the causes of vaginal stenosis and HWW syndrome in female patients who apply with abdominal pain apart from the menstruation time.

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