Case Report

Herlyn-Werner-Wunderlich syndrome: A rare cause of pain in the left iliac fossa

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Abstract

Alterations of the Müllerian ducts are rare but often treatable causes of infertility. Uterus didelphys is caused by a complete or almost complete lack of fusion of the Müllerian ducts during embryological development. As a result, two separate symmetrical uterine cavities develop with two cervices and no communication between these cavities; this is often associated with a vaginal septum, which can have a transverse wall that blocks one of the hemivaginas. Symptoms begin to develop in menarche, and complications related to a retrograde menstrual flow arise, along with pelvic adhesions and endometriosis. Some kidney abnormalities may occur. Magnetic resonance imaging can be used to diagnose and distinguish surgically correctable forms of Müllerian duct alterations. The surgical approach depends on the imaging findings. The objective of this article is to describe the radiological characteristics of the Herlyn-Werner-Wunderlich syndrome.

Keywords: Herlyn-Werner-Wunderlich syndrome, Müllerian duct anomaly, obstructed hemivagina and ipsilateral Renal Anomaly syndrome, uterus didelphys

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INTRODUCTION

Uterus didelphys accounts for about 5% of Müllerian anomalies[1] and is due to total failure of the paramesonephric ducts to fuse at about 9 weeks of gestation. This results in the development of two hemiuteri with separate endometrial cavities, with their own cervices, and no communication with the contralateral uterus. Up to 75% of patients also have a longitudinal septum dividing the vagina into two hemivaginas,[1] one of which not only has a bind end but is also associated with ipsilateral renal agenesis [Figure 1]. This association is known as Herlyn-Werner-Wunderlich syndrome, in honor of the first authors who described this syndrome; it is also called obstructed hemivagina and ipsilateral renal anomaly.[2,3] However, the latter term is broader because apart from renal agenesis, it includes other urinary tract anomalies such as double collecting system, renal duplication, and horseshoe kidney.

Herlyn-Werner-Wunderlich syndrome clinically manifests with progressive pelvic pain, dysmenorrhea, and a pelvic palpable mass due to hemihematocolpos,[4] and diagnosis is usually made after menarche. Ultrasonography has an advantage of low cost with no radiation exposure and is a useful to establish a suspected diagnosis although the diagnosis can be confirmed with magnetic resonance imaging (MRI) which is also crucial for proper surgical planning.[5]

In this article, we present a case of 12-year-old girl whose clinical and imaging features confirmed Herlyn-Werner-Wunderlich syndrome. The aim of this

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article is to describe the radiological findings that lead to the suspicion of this diagnosis.

CASE REPORT

A 12-year-old girl presented at the emergency service because of 4-day pain located in the left iliac fossa. Her menstrual cycle was regular, and her last menstruation was 3 days earlier. Her menarche occurred 8 months earlier. She had right inguinal herniorrhaphy many years earlier.

On physical examination, the abdomen was distended, globular, and tender in the hypogastrium and left iliac fossa.

Neither cervical palpation nor a speculum examination was performed because of an intact hymen.

An urgent transabdominal ultrasonography showed a left renal agenesis, a large retrovesical pelvic cystic mass with interior echoes, producing a significant mass effect on adjacent structures [Figure 1], and a small amount of free fluid in the pouch of Douglas. A provisional diagnosis of salpingitis associated with a pelvic inflammatory disease was made.

MRI examination showed left renal agenesis [Figure 2], uterus didelphys with two hemiuteri and two hemivaginas (the left hemivagina was blind), left hematometrocolpos, and hematosalpinx [Figure 3a and b]. A moderate amount of free fluid that was hyperintense on T1-weighted images was observed between the bowel loops and the pouch of Douglas.

A diagnosis of Herlyn-Werner-Wunderlich syndrome was, therefore, made. At hysteroscopy, normal right hemivagina, right hemiuterus, and right ovary and a small amount of hemoperitoneum were observed; the left hemivagina was swollen. The hematic content was released after performing a longitudinal incision on the left hemivagina. Subsequently, a marsupialization of the incision was performed, and drainage was maintained.

The clinical outcome was favorable and without complications. The patient was discharged 4 days after the operation.

DISCUSSION

Paramesonephric or Müllerian ducts are the embryological structures from which the uterus, fallopian tubes, and posterior two-thirds of the vaginas (the anterior third is formed from the urogenital sinus) are formed. The development of these ducts occurs during various embryological stages of organogenesis:[6]

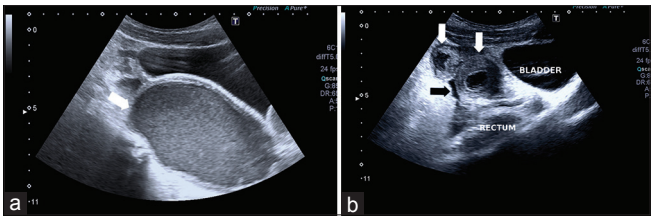

Figure 1: (a and b) Abdominal and pelvic ultrasound. A large cystic mass (white arrows) with echogenic content and thickening of the wall is shown. A small amount of anechoic free fluid is present in the pelvis (black arrow)

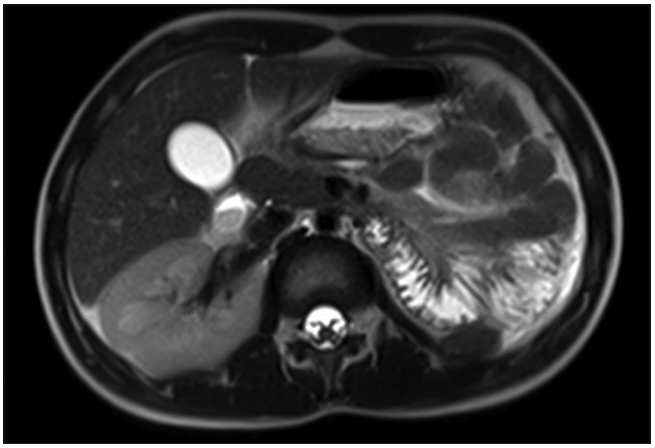

Figure 2: Abdominal magnetic resonance imaging. A HASTE sequence axial section shows the left renal agenesis

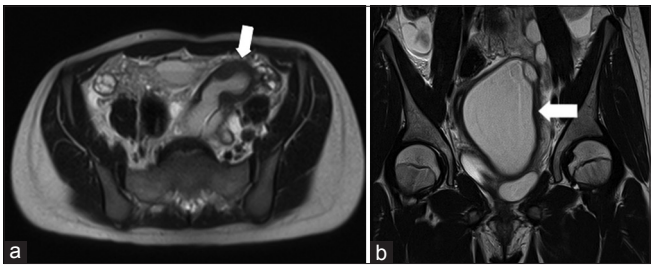

Figure 3: Pelvic magnetic resonance imaging. (a) A high-resolution, T2-weighted sequence in the axial plane shows hematometrocolpos in the left hemiuterus (white arrow). (b) Coronal plane shows a large cystic mass (white arrow)

The first stage is the invagination and elongation of the ducts at about 6 weeks of gestation; the second stage (at 9 weeks' gestation) is the caudal migration of the Müllerian duct toward the urogenital sinus and distal fusion of the ducts, forming a fibrous wall between the two fused ducts; the third stage (at 10–11 weeks' gestation) is the reabsorption of the wall to form a single endometrial cavity; and the fourth stage (at 12–22 weeks' gestation) is the formation of the vagina, with epithelialization of the vagina by cells migrating from the urogenital sinus.

Wolffian ducts, in addition to forming the kidneys, are inductors of the fusion of Müllerian ducts. Therefore,

a defect in the caudal formation of the left Wolffian duct is likely responsible for the renal agenesis in our case. In addition, this absence of the Wolffian duct is likely responsible for the lateral displacement of the left Müllerian duct, which could not be fused with the contralateral side, resulting in uterus didelphys. Furthermore, if the Müllerian duct cannot make caudal contact with the urogenital sinus, which is responsible for the formation of the anterior portion of the vagina, a blind hemivagina is formed.[7]

This impairment mainly affects the right side, but no explanation for this phenomenon is known.[8,9] However, in our case, both renal agenesis and the blind hemivagina occurred on the left side. Herlyn-Werner-Wunderlich syndrome is commonly associated with early pelvic endometriosis and can be complicated by infections (pyocolpos, pyometra, or pyosalpinx).[7]

MRI is the most reliable diagnostic imaging method available for the diagnosis of this pathology. It provides information on uterine and endometrial cavity morphology while generating a precise anatomical map for subsequent reconstructive surgeries.[10]

After resection of the vaginal wall by hysteroscopy, it is important to conduct follow-up of the patient to ensure that no decrease in vaginal caliber occurs over time.[11]

CONCLUSIONS

Sonographic demonstration of renal agenesis coexisting with a cystic mass in the pelvis strongly suggests Herlyn-Werner-Wunderlich syndrome, and this can be confirmed with MRI.

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Conflicts of interest
There are no conflicts of interest.

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