



PEDIATRIC UROLOGY CASE REPORTS

ISSN:2148–2969

journal homepage: <http://www.pediatricurologycasereports.com>

Role of OHVIRA syndrome in renal agenesis: a case report

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Abstract A 13-year-old female was admitted with abdominal pain and dysmenorrhea. Ultrasonography and magnetic resonance imaging revealed OHVIRA syndrome (uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis). A left hysterotomy was performed. The patient recovered fully following treatment with vaginal septum resection and drainage of a hematometrocolpos.

Key Words Renal agenesis, Herlyn–Werner–Wunderlich syndrome; Müllerian anomaly; obstructed vagina; OHVIRA syndrome.

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Introduction

The triad of uterine didelphys, obstructed hemivagina, and ipsilateral renal agenesis, known as OHVIRA syndrome or Herlyn-Werner-Wunderlich syndrome, is a rare congenital anomaly represented by a Müllerian duct associated with mesonephric duct anomalies [1,2].

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Accepted for publication

28 February 2014

The incidence of this anomaly has been reported to be between 0.1% and 3.8% [3]. Usually, it is diagnosed at puberty with nonspecific symptoms, such as pelvic pain, recurrent severe dysmenorrhea, and a palpable mass associated with hematocolpos or hematometra [4]. Hematocolpos or hematometra occurs with occlusion of the contralateral vaginal canal due to the fusion of the septum with the vaginal wall. Here, we describe the case of a 13-year-old girl who was diagnosed with OHVIRA syndrome after presenting to our department with recurrent

abdominal symptoms during menstruation and abdominal pain, as well as a review of the literature.

Case report

A 13-year-old female was referred to our department with complaints of acute abdominal pain and a mass on the left side of the abdomen. Her menstrual cycles had been regular, lasting for 3 to 4 days and occurring at 30-day intervals. Laboratory test results, including a complete blood cell count and urinalysis, were normal. External genital inspection showed an intact hymen and menstrual discharge. At the abdominal examination, there was lower abdominal tenderness and a palpable pelvic mass. Trans-abdominal ultrasound (US) of the pelvis revealed a 60×55 mm heterogeneous hyperechoic cystic lesion containing internal echoes in the pelvis. In addition, US revealed the absence of a left kidney and a normal right kidney. Both ovaries were normal. The renal function tests and systemic clinical expected findings were found normal. Magnetic resonance imaging (MRI) revealed uterus didelphys and a single vagina. The right uterus, cervix, and vagina were normal. There was a 6 cm mass filled with fluid suggestive of hematometrocolpos (Fig. 1a–e). Under general anesthesia, an examination with an appropriate-

size speculum revealed a single normal vagina and cervix. Transvaginally, a hemivaginal septal resection did not reveal hematometrocolpos. In surgical exploration, hematometrocolpos and hematosalpinx were observed on the left side, and the right uterus and the salpinx were normal (Fig.2 a, b). Both ovaries were normal. A unilateral hysterotomy was performed, and the obstructed hemivaginal septum was resected. Approximately 1000 ml of bloody fluid were drained from the hematocolpos and hematometra. No complications developed during the procedure. The patient was discharged on the postoperative 5th day. She continued to have regular menstrual periods, with no complications. She was asymptomatic. Two months later, the patient was admitted with complaints of dysmenorrhea. A pelvic ultrasound examination was performed for evaluation of the vagina and uterus. The septoplasty was found to have closed, with recurrence of the hematocolpos. Therefore, septoplasty was performed again, and a vaginal stent was placed. The vaginal stent was kept in place for 3 weeks. Since the removal of the vaginal stent, the patient has remained well.

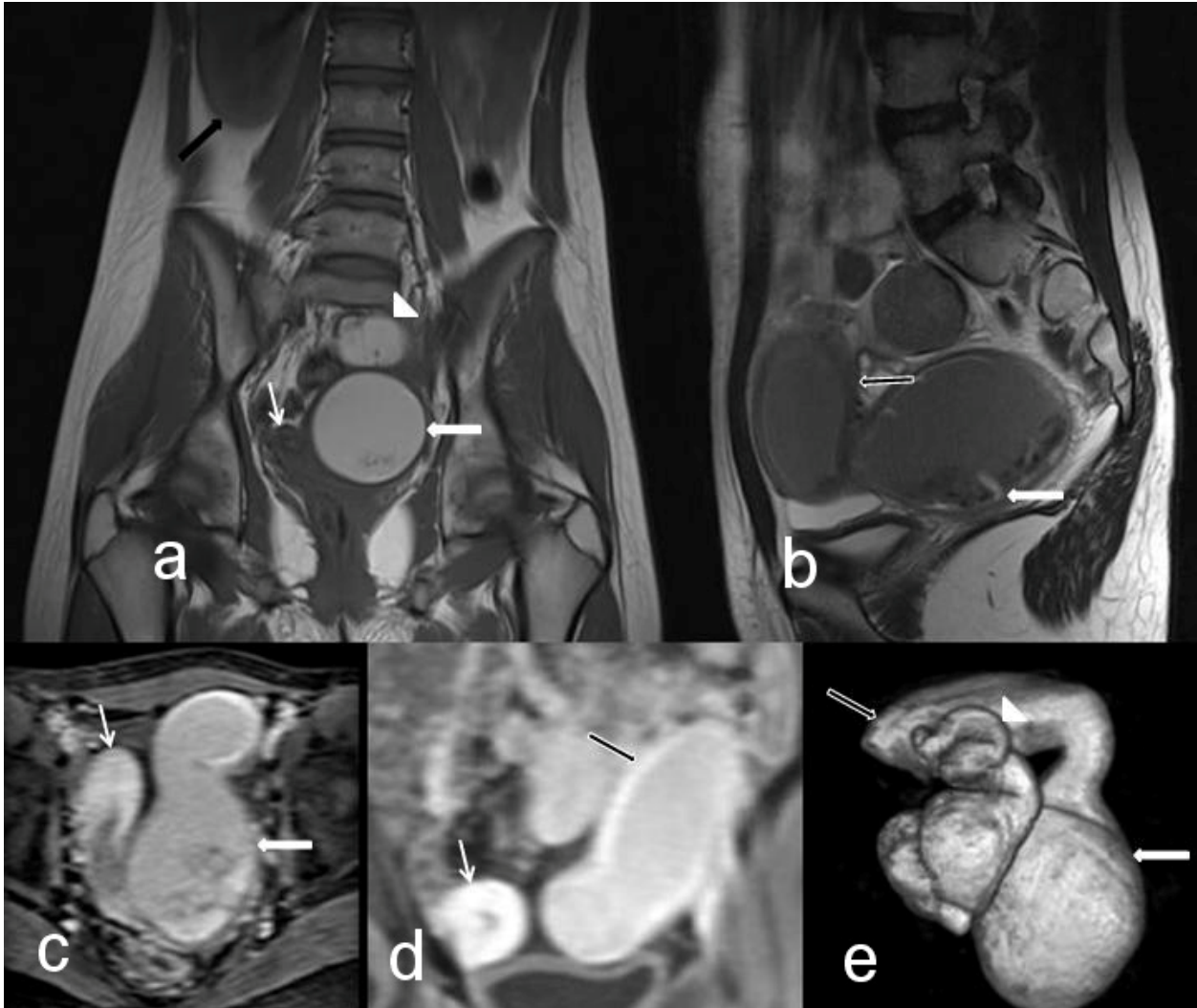


Fig. 1. (a) Coronal T1A image shows absence of the left kidney. The distended hemivagina is seen on the left side (thick white arrows) and the normal collapsed right hemivagina (thin white arrow). (b) Sagittal view of magnetic resonance imaging (T2A) showing distended left hemivagina (thick white arrows). (c,d) Axial T1A image of the pelvis shows bright signal intensity of the left hemivaginal collection (thick white arrows). The collapsed left hemivagina is seen adjacent to it (thin white arrow). (e) Volume rendering image shows the distended hemivagina on the left side (thick white arrows) and the normal collapsed right hemivagina (thin white arrow).

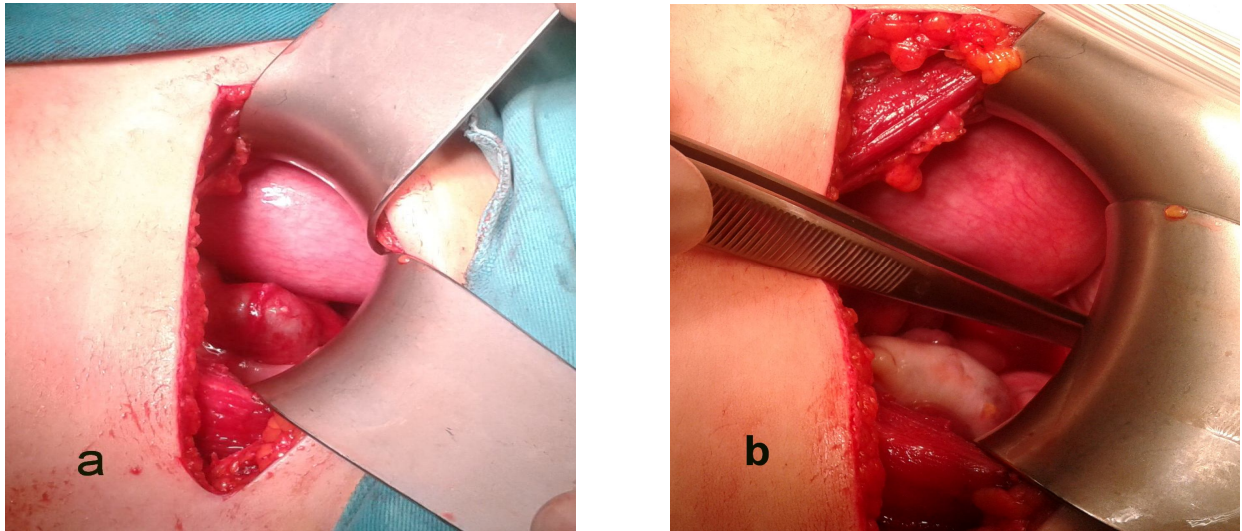


Fig. 2. (a,b) Intraoperative photograph showing uterine didelphys.

Discussion

OHVIRA syndrome is the result of the abnormal development of paramesonephric (Müllerian) and mesonephric (Wolffian) ducts. In the first 6 weeks of gestational life, the Müllerian ducts develop into the female genital tract [5,6]. Nondevelopment or nonfusion of the Müllerian ducts or failed resorption of the uterine septum causes a wide-ranging series of reproductive ducts anomalies [7,8]. The urinary and genital systems arise from a common ridge of mesoderm occurring along the dorsal body wall. Therefore, abnormal differentiation of the mesonephric and paramesonephric ducts may also be associated with anomalies of the kidneys

[9]. Renal agenesis is the most common associated nongenital anomaly, and it is always seen on the same side as the obstructed hemivagina [10].

Typically, the most common finding of a patient with OHVIRA syndrome is pelvic pain shortly following menarche in association with vaginal or pelvic mass and normal menstrual periods. A large hematometra, a dilated cervix, and hematocolpos may also be present. Pelvic examination may show a bulging vaginal mass. However, the vaginal mass may be small and difficult to determine [3,11]. Patients with OHVIRA syndrome rarely present with

infertility, vomiting, fever, and acute retention of urine [5]. The case described in this study was admitted with abdominal pain and an abdominal mass. On physical examination, a mass in the lower abdomen was evident. However, a vaginal mass could not be determined.

Although US, computerized tomography scans, and hysterosalpingography are commonly for the diagnosis of OHVIRA syndrome, MRI is the most accurate diagnostic method [12]. Laparoscopy is not needed for the diagnosis of most of the cases [13]. Renal agenesis including the more common right side and Müllerian ducts anomaly often coexists [2]. So, in all patients with Müllerian ducts anomaly, it is important to investigate the urinary tract anomaly. Obstructive genital lesions may be associated with other anomalies, such as coarctation of the aorta, atrial septal defects, and abnormalities of the lumbar spine. Hence, a complete physical examination and abdominal tests are required in all cases [14]. The incidence of unilateral renal agenesis in OHVIRA syndrome is 81%, and 25–50% of affected women exhibit associated

genital abnormalities [15]. As in other studies, the diagnosis of the patient in the current case was made with US and MRI. The patient was diagnosed with OHVIRA syndrome based on the presence of left renal agenesis, uterus didelphys, and a unilateral obstructed hemivagina, with resultant hematometrocolpos.

Resection of the vaginal septum is the treatment of choice for an obstructed hemivagina. The treatment invariably requires surgical intervention in the form of excision of the vaginal septum to relieve the obstruction. Hemihysterectomy is no longer preferred, as the reported incidence of pregnancy is almost the same with hemihysterectomy and transvaginally repair. Reconstruction of the vagina can be accomplished by careful excision of the vaginal septum, with precautions taken to avoid injury to the urethra, bladder, and rectum. An abdominal approach may be necessary in cases of failed septum resection or when the anatomy is difficult to determine [2,3,10,14]. In our patient, transvaginal septum resection was insufficient. Hence, a unilateral hysterotomy was performed, and the obstructed hemivaginal septum was resected. A successful pregnancy occurs in 87% of the patients with OHVIRA syndrome.

In young females with renal anomalies presenting with a pelvic mass, abdominal pain, and acute urinary retention, OHVIRA syndrome should be considered in the differential diagnoses. In addition, we recommend evaluation of the genital tract in all girls with renal agenesis.

CONFLICT OF INTEREST

None declared.

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