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Research Letter

Obstructed hemivagina and ipsilateral renal anomaly with uterus didelphys and vaginal discharge

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Vaginal discharge is one of the most common vaginal symptoms whose causative conditions include vaginal candidiasis, chlamydial cervicitis, bacterial vaginosis, and cervical carcinoma [1].

The syndrome of obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) was first reported in 1922, and it is also known as Herlyn–Werner–Wunderlich syndrome. The concomitant uterine anomaly has been described as didelphys or duplicated uterus [2,3]. The typical patient is an adolescent girl with nonspecific symptoms such as pelvic pain or dysmenorrhea.

This is a case report of a 19-year-old female patient who presented to our obstetrics and gynecology department with frequent, copious amounts of blood-tinged, foul-swelling vaginal discharge. Since the age of 14 years, the patient has had frequent consultations with physicians at various clinics regarding this vaginal discharge. Symptoms would resolve after every antibiotic treatment.

Menarche occurred at the age of 12 years, and thereafter the patient claimed to have a regular monthly menstrual interval every 28–30 days lasting for 5–7 days. The patient stated that her father and aunt have polycystic kidney disease. The gross appearance of her external genitalia was normal. A speculum examination was not performed because of the absence of a history of sexual intercourse. Other physical and systemic examinations were unremarkable.

Abdominal ultrasound revealed a large, well-defined cystic lesion with a hypoechogenic elliptical cystic mass (Fig. 1A). A detailed sonogram revealed a double uterine fundus with its corresponding endometrial canals (Fig. 1). Both ovaries appeared normal. In addition, absence of the right kidney and a left cystic kidney were noted.

Magnetic resonance imaging (MRI) revealed two uterine horns, two cervices, and two vaginal cavities. The collapsed left hemivagina was seen to communicate with the left cervix and corresponding uterine horn. The right hemivagina was markedly distended, whereas its right cervix and corresponding uterine horn were not distended. The right cervix was seen to communicate with the upper end of a half-moon-shaped, distended right hemivagina on a sagittal MRI scan (Figs. 2 and 3).

The patient was scheduled for surgery under intravenous anesthesia. As the speculum was inserted, a bulging right vaginal wall was noted with a puncta on the posterior apex of the vaginal bulge (Fig. 4), through which pus was escaping continuously. A needle was introduced through the puncta and a 3-cm incision was made anteriorly from the puncta. Pus squirted from the incision site (Fig. 5). An additional circular incision was made around the area. Pus was drained completely, and continuous suture was used to close the incision (Fig. 6). Pus culture revealed the presence of *Escherichia coli*. Histopathology of the excised tissue revealed a vaginal septum. Follow-up MRI 1 year after surgery showed double uterine horns and a collapsed cervix without a distended mass (Fig. 7).

The incidence of Müllerian defects is 1.1–3.5% [4]. OHVIRA is a rare congenital anomaly constituting 0.16–10% of all Müllerian duct abnormalities [5]. In OHVIRA, a didelphic uterus is suggestive of embryologic arrest during Week 8 of gestation that ultimately affects the Müllerian and metanephric ducts [6]. Generally, the paramesonephric ducts appear at 44–48 days of gestation as longitudinal invaginations of the surface epithelium along the mesonephric ridge lateral to mesonephric ducts [7]. An early failure of the metanephric diverticulum to develop (at ~5 weeks) from the mesonephric duct results in agenesis of the ureteric bud, which leads to agenesis of the ipsilateral ureter and kidney [8].

Patients with a double uterus with OHVIRA can generally be categorized into three diagnostic groups: Group 1 patients

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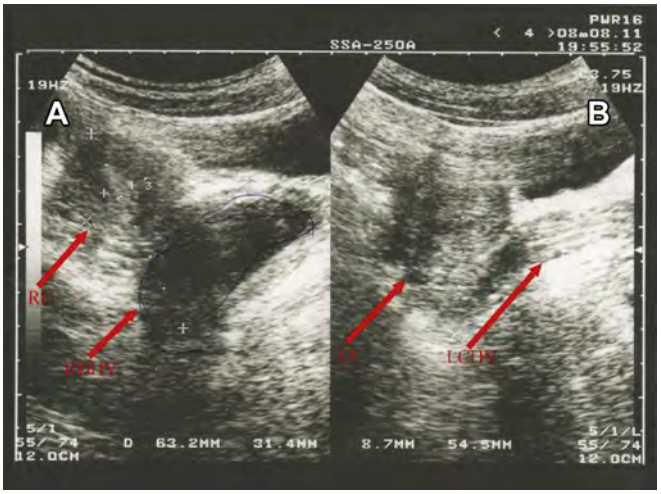

Fig. 1. Sonographic image, transverse view of (A) right uterine cavity with endometrium appearing normal and a cystic hypoechoic lesion, located anterior to the cervix, and (B) normal left uterine cavity and endometrium. LCHV = left collapsed hemivagina; LU = left uterus; RDHV = right distended hemivagina;. RU = right uterus.

have complete unilateral vaginal obstruction without uterine communication; Group 2 patients have an incomplete unilateral vaginal obstruction without uterine communication; and Group 3 patients have complete vaginal obstruction with a laterally communicating double uterus. Its accompanying symptoms include lower abdominal pain, severe dysmenorrhea, excessive foul and mucopurulent discharge, intermenstrual bleeding, and a paravaginal mass [9].

The patient in this case report probably belonged to Group 2. There was a continuous foul-smelling discharge oozing from the puncta, which resulted from an ascending infection. The puncta arose from incomplete obstruction of the right

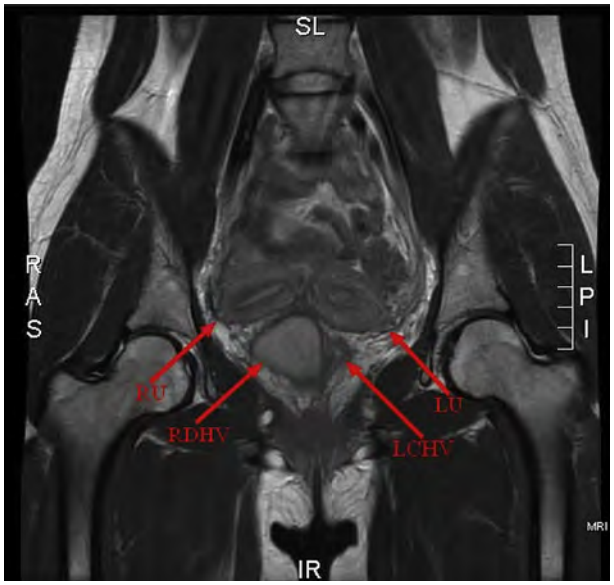

Fig. 2. Magnetic resonance imaging of the pelvis. The distended hemivagina is seen on the right side and an adjacent left collapsed hemivagina. Image also shows right and left uterine horns. LCHV = left collapsed hemivagina; LU = left uterus; RDHV = right distended hemivagina; RU = right uterus.

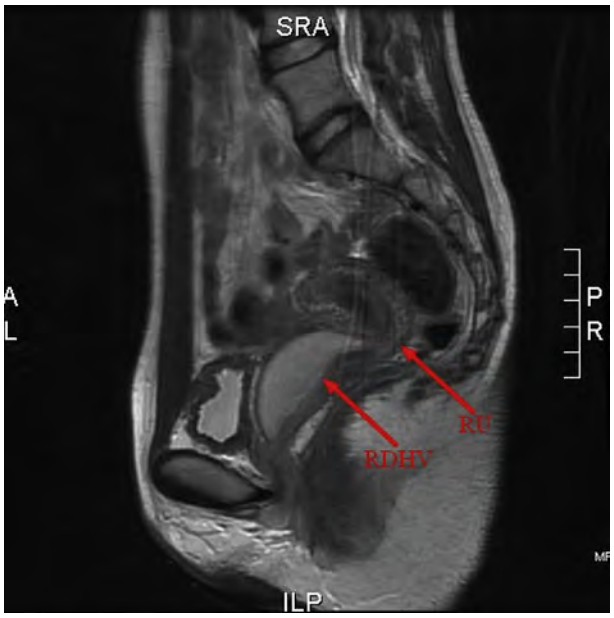

Fig. 3. Sagittal view of magnetic resonance imaging showing a half-moon-shaped, distended right hemivagina. RDHV = right distended hemivagina; RU = right uterus.

hemivagina, which explained the presence of continuous vaginal discharge without severe dysmenorrhea. Whether the formation of the puncta was congenital or acquired was unclear.

In imaging studies, pelvic sonography would often show an elliptical cystic mass with or without a distended uterine horn

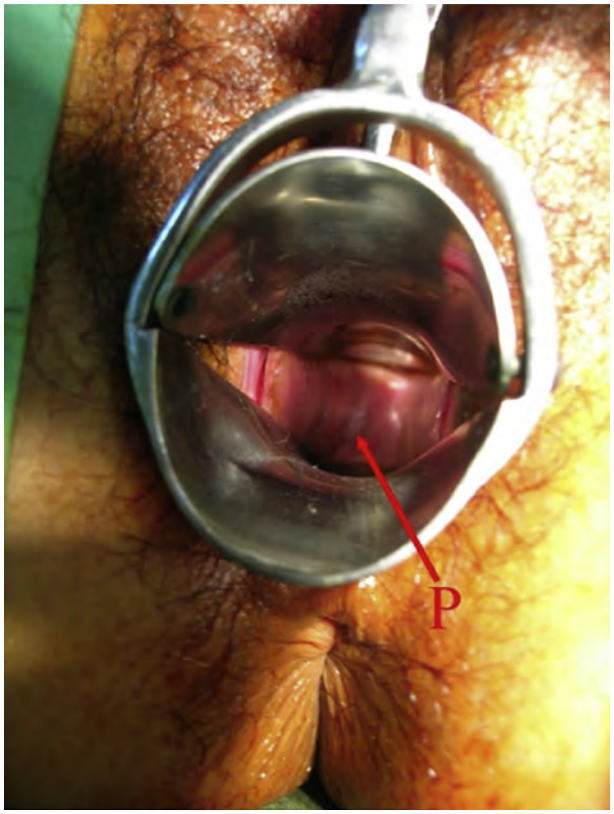

Fig. 4. A bulging right vaginal sidewall with a puncta. P = puncta.

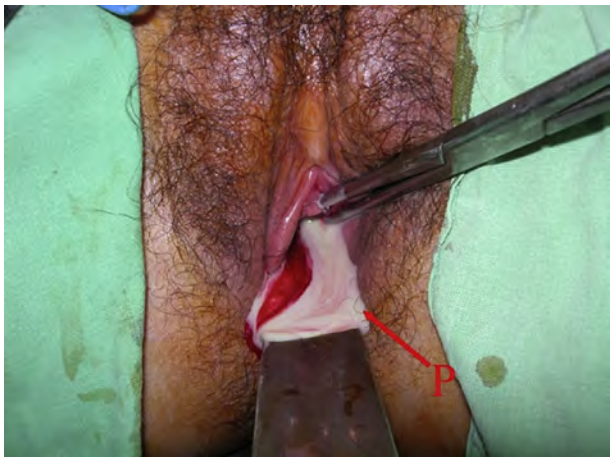

Fig. 5. Pus escaping from the incision site. P = pus.

and fallopian tube (hematometrocolpos with hematosalpinx) and another nondistended uterine horn. An elliptical or half-moon-shaped, distended cystic mass in sagittal view that connects with the cervix is suggestive of a completely or partially obstructed vagina. In addition, renal agenesis or absence is always seen on the side with the obstructed hemivagina [3].

MRI is superior to ultrasound because it allows better characterization of anatomic relations, given its multiplanar capabilities and wider field of view that are important for surgical planning [10]. Many authors have reported a high accuracy of MRI in these cases [2]. Smith and Laufer [2] reported correct MRI diagnoses in 15 of 16 cases from referrals, with one incorrect diagnosis and one case of ovarian cyst combined with OHVIRA.

A correct diagnosis could be made with history taking and physical examination in combination with appropriate imaging. Smith and Laufer [2] have suggested that laparoscopy should be reserved for cases in which the diagnosis is not clear after imaging or when MRI is not available, rather than being performed as a routine procedure.

Treatment of this syndrome is with vaginoplasty to relieve obstruction, prevent further complications, and decrease severity of dysmenorrhea. Traditionally, two methods are

Fig. 6. Continuous suture to close the incision site. S = septum.

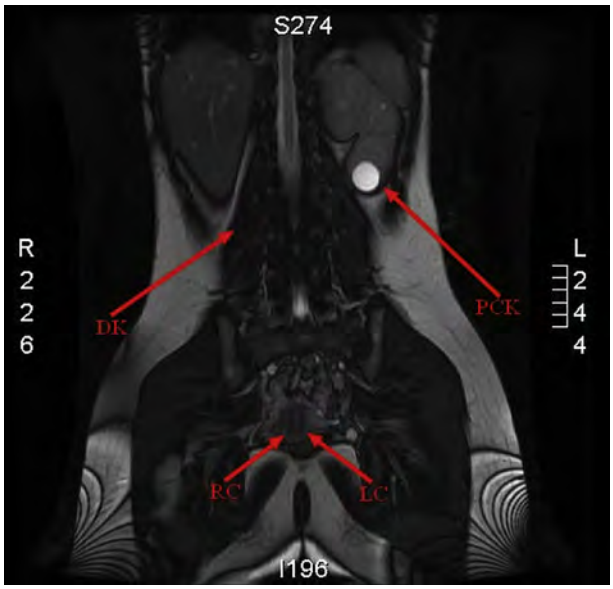

Fig. 7. Follow-up of magnetic resonance imaging 1 year later showing double cervices with absence of a previous right distended mass, right renal dysplasia, and left polycystic kidney. LC = left cervix; RC = right cervix; RDK = right dysplastic kidney; RPCK = right polycystic kidney.

suggested. A single-stage vaginoplasty includes marsupialization and complete resection of the septum, if there is no infection or other complication [2]. A two-stage procedure includes limited resection (3 cm) sufficient for adequate drainage, followed by removal of any remaining vaginal septum 1 month later, if inadequate resection or possible postoperative stenosis occurs [9]. Chao-Lan et al [11] reported resectoscopic excision of the vaginal septum as an alternative procedure for virgin females.

In this case, we decided to perform a two-stage procedure initially. However, after primary resection of the vaginal septum, the patient felt better and no stenosis was found. In addition, the patient did not wish to undergo another surgical procedure. One year later, a repeat MRI showed no further complications.

A vaginal septum and its complications were hypothesized to be associated with an obstructive lesion. Smith and Laufer [2] found that only 23% of patients with OHVIRA syndrome had endometriotic implants. Routine laparoscopy is not essential for management except when complications such as pyocolpos, pelvic inflammatory disease, or tubo-ovarian abscess are present. Antibiotic treatment and surgery may be necessary, and postoperative complications are uncommon. Occasional vaginal stenosis is noted and may be associated with adenosis [2]. In the presence of pyocolpos, hysteroscopy could result in increased infection risk due to regurgitation of pus into the pelvis or bloodstream. Therefore, a hysteroscopic procedure is not suitable in this situation.

OHVIRA is a rare congenital anomaly. Its diagnosis requires careful history taking, physical examination, and appropriate imaging studies. Vaginal discharge is a common symptom in most gynecological conditions. However, the

presence of recurrent vaginal discharge mandates further investigation for an accurate diagnosis [12,13].

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