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Case Report

Significance of expeditious diagnosis and treatment following menarche in obstructed hemivagina and ipsilateral renal agenesis syndrome: A case report☆,☆☆

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ABSTRACT

Obstructed hemivagina and ipsilateral renal agenesis syndrome is a rare complex of structural abnormalities of female urogenital tract. Moreover, delayed treatment of this condition can reportedly lead to pelvic infection, endometriosis, and infertility. We present a rare case of obstructed hemivagina and ipsilateral renal agenesis syndrome wherein appropriate treatment was initiated early due to the identification of left renal agenesis. A 14-year-old girl visited the gynecology department due to worsening left lower abdominal pain after menarche. She was diagnosed with renal dysplasia in the pediatric department at the ages of 3 and 9 years, and it was recommended that she visits the gynecology department right after menarche. She experienced her first menarche at the age of 14 years. Imaging studies revealed uterine didelphys with occlusion of the left vagina and formation of hematocolpos. Additionally, hematomas were observed in the left uterine cavity and the left fallopian tube; left renal agenesis was confirmed. Based on these findings, we diagnosed this patient with obstructed hemivagina and ipsilateral renal agenesis syndrome and performed a surgical incision and excision of the closed left vaginal wall, resulting in the disappearance of the hematocolpos. Approximately 2 years postoperatively, the patient has remained recurrence-free. Early diagnosis and treatment are crucial for preserving fertility in patients with obstructed hemivagina and ipsilateral renal agenesis syndrome, and early referral to the obstetrics and gynecology department is recommended if renal dysplasia is detected during childhood.

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Introduction

Obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) syndrome is a congenital abnormality of the urinary and reproductive systems. In 1971, Herlyn and Werner [1] reported a case of a double uterus, ipsilateral blind vagina, and renal agenesis. Similarly, Wunderlich [2] reported a case of a bicornuate uterus, ipsilateral blind hemivagina, and kidney aplasia in 1976. In these aforementioned reports, the same condition was referred to as Herlyn-Werner or Wunderlich syndrome. Consequently, to avoid confusion often encountered with the name of the condition, the term "Obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) syndrome" was proposed by Smith and Laufer [3]. The exact incidence of this condition is unknown, but reports suggest a prevalence of approximately 0.1%-3.8% [4]. Owing to its rarity, unnecessary diagnostic laparoscopy and open surgery may be performed, and a delayed diagnosis can lead to complications, such as pelvic infections and endometriosis, which can cause infertility. In this study, we present a case in which OHVIRA syndrome was diagnosed immediately after the onset of menarche, influenced by the identification of left renal agenesis during a pediatric examination at ages of 3 and 9 years. We observed that expeditious diagnosis allows for appropriate treatment at an early stage.

Case report

A 14-year-old girl visited the gynecology department due to worsening left lower abdominal pain after menarche. She had no related medical family history. At 3 years old, she underwent an abdominal ultrasound examination owing to fever during a visit to the pediatric clinic that revealed renal agenesis. At 9 years old, during another visit to the pediatric clinic, the possibility of renal agenesis and abnormalities in the internal genitalia was mentioned. After her first menstrual period, she began experiencing worsening left lower abdominal pain and sought a gynecological consultation. Although no internal examination was performed, abdominal palpation revealed a soft and tender mass in the left lower abdomen.

On magnetic resonance imaging (MRI), the uterus showed a bicornuate appearance with a double cervix. A cystic tumor measuring approximately 8.5 cm in diameter was observed in the left lower abdomen, containing fluid with a high T1 signal intensity (Fig. 1). The left uterine cavity and fallopian tube were also enlarged and filled with fluid with a high T2 signal intensity (Figs. 2 and 3). Left renal agenesis was also observed on MRI (Fig. 4). Based on the clinical course and imaging findings, the patient was diagnosed with OHVIRA syndrome. Upon examination under general anesthesia, the right vaginal canal and external uterine opening of the right uterus were observed, with the left vaginal wall bulging into the right vaginal canal. When incised, gelatinous blood flowed out. To prevent adhesion of the incision to the vagina, a portion of the vaginal wall was excised, and the surrounding area of the incision was sutured with absorbable sutures (Fig. 5). Postoperatively, the abdominal pain resolved. Follow-up MRI performed 6 months postoperatively showed the disappearance of the hematoma in the left vagina. There has been no recurrence of the hematoma formation in the left vagina or uterus 2 years postoperatively.

Discussion

OHVIRA syndrome is a genitourinary malformation characterized by an obstructed hemivagina and ipsilateral renal agenesis. Symptoms include abdominal and menstrual pain after the onset of menstruation and, in some cases, a palpable abdominal mass. These symptoms arise because of the accumulation of blood in the closed vagina and uterus on

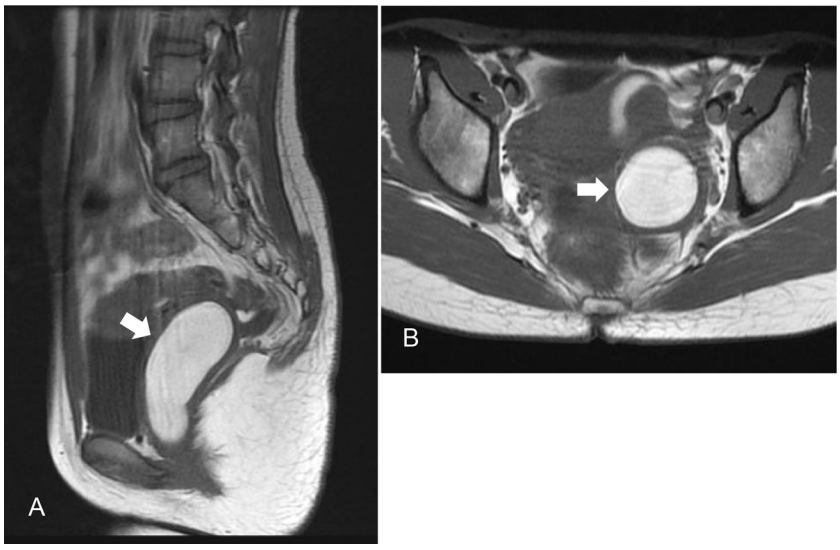

Fig. 1 – T1-weighted magnetic resonance images, (A) sagittal section and (B) transverse section. A high T1 signal indicating hematoma in obstructed hemivagina (white arrow).

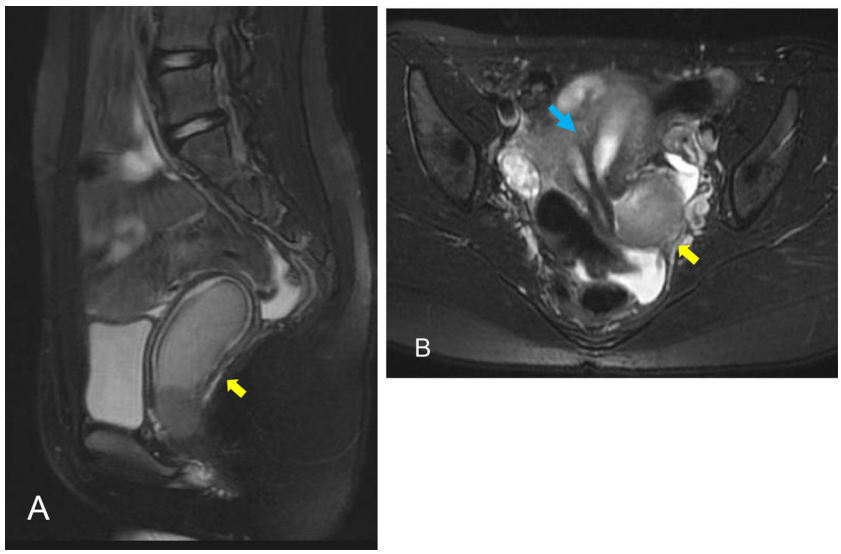

Fig. 2 – T2-weighted magnetic resonance images, (A) sagittal section and (B) transverse section. The image shows uterus didelphys (blue arrow) and blood signals within the closed left vaginal cavity. Yellow arrow indicates obstructed hemivagina filled with hematoma.

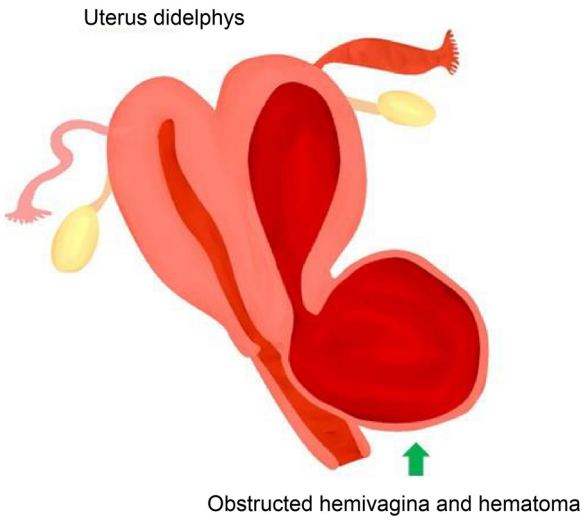

Fig. 3 – An image showing uterus didelphys and obstructed hemivagina filled with hematoma (green arrow).

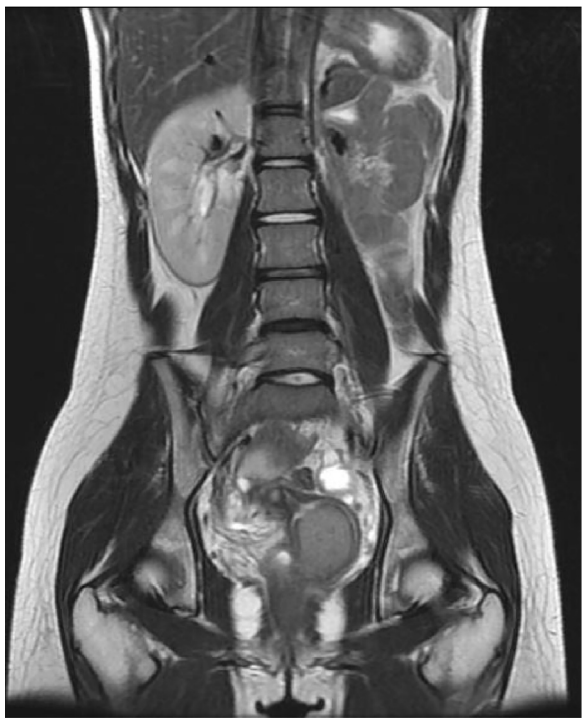

Fig. 4 – T2-weighted magnetic resonance images showing agenesis of the left kidney.

the affected side. Notably, in cases of complete obstruction on the affected side of the vagina, these symptoms occur early after menstruation and can be severe. Conversely, in incomplete obstruction cases, symptoms are slow, and the condition may be diagnosed in late adolescence or in the 20s [5]. MRI is highly effective in its diagnosis, as it allows for the observation of uterine anomalies and vaginal and tubal hematomas [6]. Transabdominal ultrasonography and CT are useful for detecting abnormalities in the urinary system, particularly renal agenesis, and a laparoscopic examination may be necessary for diagnosis in cases with pelvic infection or endometriosis. The principal challenge with this condition is that unnecessary laparoscopic or open abdominal surgeries are often performed for its diagnosis due to its rarity. Smith and Laufer [3] reviewed 27 cases referred to 2 large referral centers and found that laparoscopy was performed in 4 cases and open abdominal surgery in one case before referral, but none led to a correct diagnosis. Another issue is the potential development of tubal hematoma, pelvic infections, pelvic

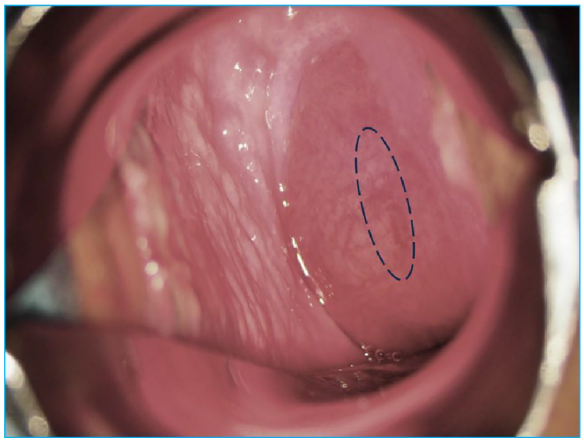

Fig. 5 – Vaginal examination under general anesthesia. The closed vagina on the left is bulging. The vaginal wall was excised along the dotted line.

adhesions, and endometriosis due to the accumulation and retrograde flow of menstrual blood into the uterus and fallopian tubes, which can lead to infertility. Tong et al. [7] reported the presence of endometriosis in 17% of OHVIRA syndrome cases and a history of pelvic inflammatory disease in 20% of their cases. Kudela et al. [8] conducted a systematic review of 734 OHVIRA syndrome cases and reported the presence of endometriosis in 14% of the patients who underwent laparoscopic examination. They mentioned that laparoscopic examination was only performed in a small number of patients and that there might be a higher incidence of endometriosis in more cases [8], suggesting that early diagnosis and treatment are important to preserve fertility. In the present case, renal agenesis was detected on an abdominal ultrasound that was performed when the patient was 3 years old. When she was 9 years old, the possibility of renal agenesis and internal genital anomalies was mentioned in the pediatric department, and the patient was advised to visit the obstetrics and gynecology department after menstruation onset, leading to expeditious diagnosis and treatment immediately after menarche. Recently, abdominal ultrasonography has been performed more frequently in pediatric departments, and renal agenesis can now be detected during childhood, and it has been reported that more than 30% of these cases have accompanying Mullerian anomalies [9,10]. Thus, if a pediatrician identifies unilateral renal agenesis in a female child, it is advisable to inform the child's parents to visit the obstetrics and gynecology department as soon as menstruation begins [11]. Generally, the most commonly performed treatment is single-stage vaginoplasty, which involves the incision and excision of the closed vagina and marsupialization, and re-excision is recommended in cases of restenosis at the excision site. Regarding fertility, Kudela et al. [8] analyzed multiple case series and stated that pregnancy was achieved in 62%-95% of patients who desired to conceive. In another report, of the 33 patients who desired to conceive among 79 patients with OHVIRA syndrome, 52 pregnancies occurred, with 19 (37%) pregnancies on the side with vaginal closure and 33 (64%) on the opposite side [5]. The pregnancy outcomes included 23% miscarriages, 15% preterm births, and 62% full-term births [6]. Another report mentioned that out of 15 patients who desired to conceive in cases of duplicated uterus, duplicated vagina, and ipsilateral renal agenesis, 87% achieved pregnancy and 77% had normal deliveries [12]. Early diagnosis and treatment can potentially help preserve the fertility of the patient. However, due to its rarity, pediatricians are not yet familiar with OHVIRA syndrome.

Conclusion

Delayed treatment of OHVIRA syndrome can reportedly lead to pelvic infection, endometriosis, and infertility. If renal agenesis is detected during the fetal or pediatric period, early visits to the obstetrics and gynecology department promptly after menarche are recommended in order to preserve fertility in patients with OHVIRA syndrome.

Ethical approval

This study was approved by the Institutional Review Board of Nagano Municipal Hospital (Nagano, Japan).

Patient consent

Informed consent was obtained from the patient and guardian.

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