



Case report

Obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) syndrome: A case report

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ABSTRACT

Introduction: Obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) is a rare urogenital malformation. The clinical symptoms of OHVIRA include uterine morphology abnormality, persistent vaginal discharge, and renal anomaly or agenesis. Delayed diagnosis can lead to complications, such as pelvic inflammatory disease, adhesion to the oviduct, and endometriosis.

Presentation of case: We report the case of a 12-year-old girl who presented with severe dysmenorrhea and abnormal vaginal discharge. The patient was diagnosed with OHVIRA based on magnetic resonance imaging findings. The patient underwent combined transvaginal and laparoscopic surgery for drainage of hematocolpos and adhesiolysis of the pelvic cavity. The patient had an uncomplicated recovery and normal menstrual cycle after surgery.

Discussion: OHVIRA is a rare syndrome, of which delayed diagnosis may result in the development of endometriosis.

Conclusion: We report that a combined laparoscopic and transvaginal approach was useful for treating OHVIRA with oviductal hematoma.

1. Introduction

The Müllerian ducts develop to form the fallopian tubes, uterus, cervix, and upper one-third of the vagina at 6 weeks' gestation. In addition, The Wolffian ducts have crucial roles in developing the Müllerian ducts and kidneys. Thus, abnormal development of the Wolffian ducts leads to uterine and vaginal malformations complicated by urologic abnormalities. Obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) syndrome, Herlyn-Werner syndrome, and Wunderlich syndrome are similar diseases: their embryopathogenesis is thought to be caused by Wolffian duct dysplasia and Müllerian ducts fusion failure [1]. They are rare diseases: the incidence of OHVIRA is reported to be 1/20,000 [2]. There are two types of OHVIRA syndrome depending on whether it involves complete vaginal obstruction or contralateral vaginal traffic [3]. Since menstrual blood accumulates in the uterine lumen, OHVIRA with complete vaginal obstruction is often diagnosed after menarche due to recurrent severe dysmenorrhea.

Treatment of OHVIRA usually involves blood drainage or removal of the affected uterus. However, if diagnosis is difficult, a laparoscopic examination may be useful for confirming uterine morphology. In cases of delayed diagnosis, patients may develop endometriosis owing to retrograde menstruation, which may require surgical treatment. Herein, we report a case of OHVIRA syndrome complicated by complete vaginal obstruction and tubal hematoma. This case was reported based on the latest Consensus Surgical Case Report (SCARE) Guidelines [4], and emphasizes the usefulness of laparoscopic examination in the treatment and diagnosis of OHVIRA syndrome.

2. Presentation of case

The patient, a 12-year-old girl, had no family history of note, without remarkable medical or surgical history. The patient's first menstruation occurred at 11 years of age. Although the patient had regular menstrual cycles, dysmenorrhea occurred, for which medication for pain relief was

Abbreviations: OHVIRA, obstructed hemivagina and ipsilateral renal agenesis.

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administered. Thereafter, because the symptoms of dysmenorrhea became more severe, the patient visited a nearby obstetrics and gynecology department.

The patient experienced lower abdominal pain only during menstruation. Moreover, there was no tenderness in the Douglas fossa on examination. Transabdominal ultrasonography revealed large blood retention in the uterus (Fig. 1). Magnetic resonance imaging revealed an enlarged left fallopian tube, two cervixes, and a duplicated uterus with blood retention in the left uterus (Fig. 2). The uterus, cervix, and vagina on the right side were normal, whereas the left side vagina was quite short and obstructed. Both ovaries appeared normal. They were not examined with regard to the kidneys. The patient was referred to our hospital, wherein transabdominal ultrasonography could not identify the left kidney. We suspected a rare disease of Müllerian origin and performed a computed tomography, which confirmed the kidney defect (Fig. 3). Taken together, OHVIRA syndrome was considered as the patient had left renal agenesis, two cervixes, a duplicate uterus, and obstructed hemivagina. Drainage of blood retention by the vaginal approach combined with laparoscopic examination was planned for confirming uterine morphology and treat abdominal adhesions.

First, the hemivaginal septum was incised with an electrocautery scalpel under transabdominal ultrasonography guidance, and the left-side cervix was identified. The left cervix was dilated, after which blood retention was drained using forceps. Next, a duplicated uterus and left tubal hematoma were confirmed via laparoscopic examination (Fig. 4). The omentum was adherent around the left fallopian tube. These adhesions were peeled with electrocautery. There were no findings of adhesions or suspected endometriotic lesions in the abdominal cavity other than those around the left fallopian tube. Pathological examination was not performed. These procedures were performed without obvious complications. The patient was discharged on post-operative day four. Nine months after surgery, monthly menstruation occurred without complications or medication. Furthermore, the left oviductal hematoma improved.

3. Discussion

Female genitourinary tract malformations are common: they are found in approximately 3–4 % of the general population and in 15 % of patients experiencing recurrent miscarriage [1]. In contrast, OHVIRA syndrome, which is characterized by a duplicated uterus, unilateral vaginal closure, and ipsilateral renal aplasia, is considered a rare disorder. Similar to OHVIRA syndrome, Wunderlich syndrome and Herlyn-Werner syndrome are diseases caused by Wolffian duct dysplasia. The



Fig. 1. Transabdominal ultrasonography (Ta-USG): Ta-USG shows blood retention in the uterus (indicated with arrows).

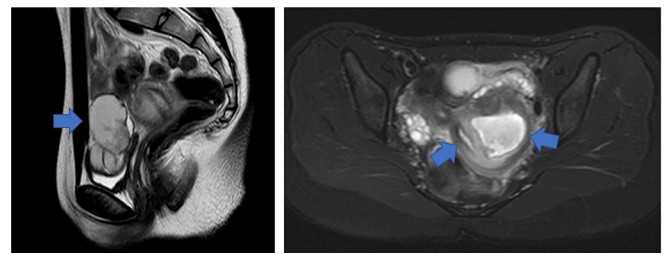


Fig. 2. A) T2 MRI sagittal view: Oviductal hematoma is found on the ventral side of the uterus shows the left oviductal hematoma (indicated with arrows). B) T2 MRI in axial view: The two-uterine cavities and mass filled with fluid suggestive of blood retention on the left side resulting in the formation of the left oviductal hematoma.

MRI, Magnetic resonance imaging.

Wolffian duct is involved in the development of the kidney and induction of the Müllerian duct: their defect would manifest with a duplicated uterus and kidney defect. The difference between OHVIRA and Wunderlich, which manifests a duplicated uterus and renal aplasia, is the presence or absence of a duplicated vagina. The present OHVIRA case could be categorized as classU3 b (bicorporeal uterus) according to the ESHRE/ESGE classification, with a blind end at the vaginal septum [5]. The risk of vaginal wall perforation during surgery is increased in patients with Wunderlich syndrome, which has a single cervix, if misidentified as OHVIRA syndrome. Therefore, laparoscopic examination could be of great help to confirm uterine morphology for precise diagnosis and reduction in the risk of complications.

In patients with OHVIRA syndrome, diagnosis is usually made based on abdominal pain after menarche due to increased blood retention due to obstructed hemivagina. However, the boundary between the dilated cervix and vagina is often indistinct, making diagnosis of OHVIRA syndrome difficult. This syndrome may remain unrecognized if it is not suspected because of normal menstruation through the unobstructed hemivagina. These cases can possibly be treated as simple dysmenorrhea and overlooked in routine practice. If this syndrome left untreated, retrograde menstruation results in tubal hematoma and endometriosis in the abdomen, consequently leading to infertility [6]. Therefore, aggressive surgical excision of the vaginal septum is preferable to symptomatic treatment for diseases manifesting with dysmenorrhea and blood retention.

On the contrary, a good prognosis of pregnancy outcome was shown in patients with OHVIRA syndrome treated with appropriate therapies [7,8]. Laparoscopic examination should be combined for fertility preservation in cases where presence of adhesions and endometriosis are suspected. In addition, in this case, laparoscopic observation of the abdominal cavity ruled out endometriosis and indicated that there was no need for drug therapy (progesterone or oral contraceptives).

4. Conclusion

OHVIRA is a rare syndrome, its delayed diagnosis may result in the development of endometriosis. The combined use of laparoscopy enables precise diagnosis and safe surgery, as well as treatment of intra-abdominal lesions.

Patient perspective

The patient reports to be satisfied by the procedure. She was suffering before the surgery from dysmenorrhea and feels no menstrual pain now.

Ethical approval

Ethical approval by the Tokushima University Hospital was not



Fig. 3. Abdominal CT (coronal section): CT shows a defect in the left kidney, and the right kidney is mildly enlarged. CT, computed tomography.

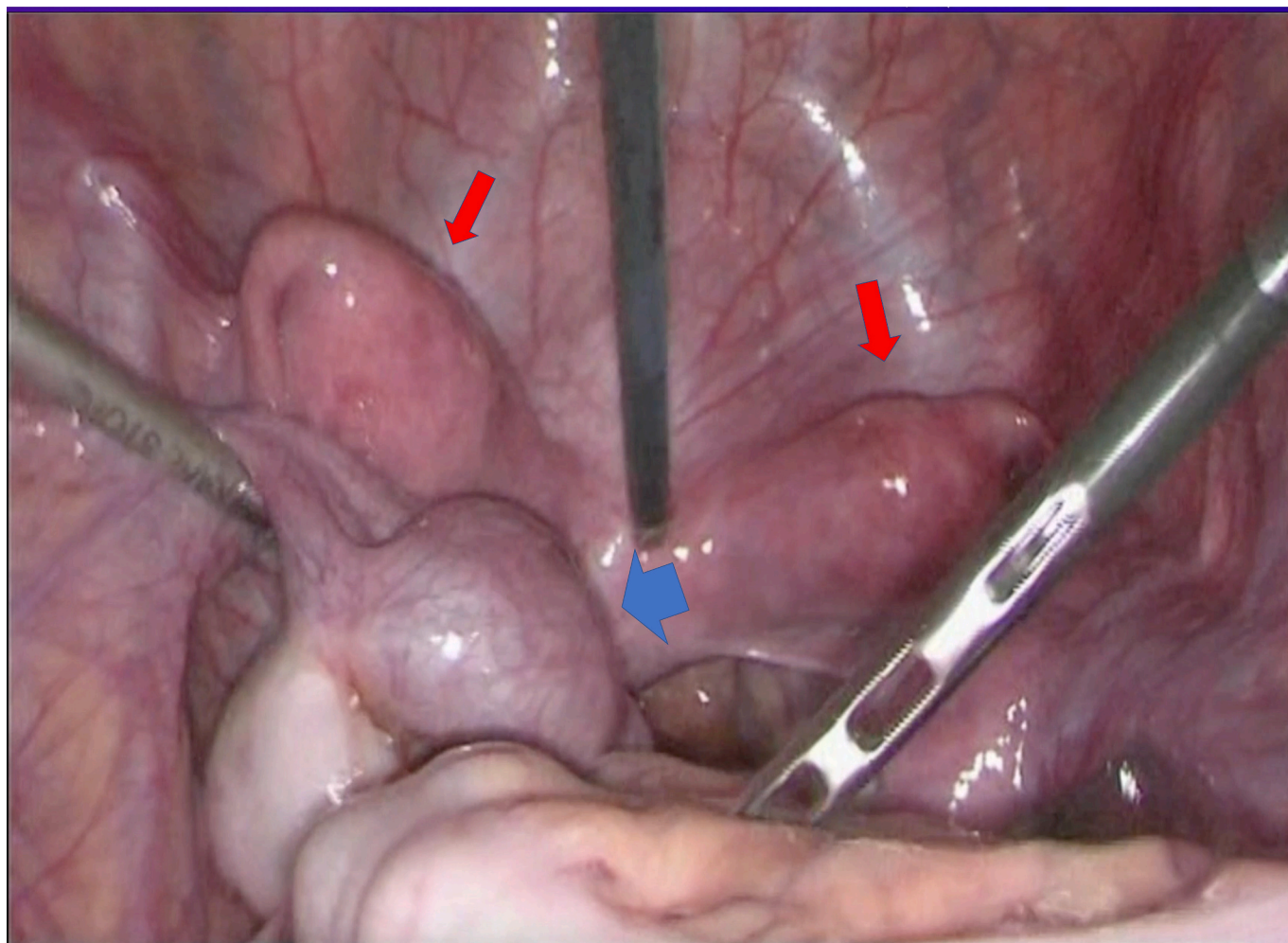


Fig. 4. Laparoscopic image: Two hemiuteruses (indicated by red arrows) and the left oviductal hematoma (indicated by blue arrows) is observed. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

required because written informed consent was obtained from the patient and patient's parents/legal guardian for publication of this case report and accompanying images.

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CRedit authorship contribution statement

Ryosuke Arakaki and Kanako Yoshida designed and executed the research and wrote the manuscript.

Takeshi Kato contributed to the concept and helped to write the manuscript.

Takahi Kaji is co-supervisor and edited the manuscript.

Junki Imaizumi made substantial contributions in the management of the patient.

Takeshi Iwasa is a supervisor and edited the manuscript.

Guarantor

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Parental consent for minors

Written informed consent was obtained from the patient and patient's parents/legal guardian for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Declaration of competing interest

None declared.

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References

- [1] P. Acien, M. Acien, The presentation and management of complex female genital malformations, *Hum. Reprod. Update* 22 (2016) 48–69, <https://doi.org/10.1093/humupd/dmv048>.
- [2] P. Acien, M.I. Acien, The history of female genital tract malformation classifications and proposal of an updated system, *Hum. Reprod. Update* 17 (2011) 693–705, <https://doi.org/10.1093/humupd/dmr021>.
- [3] J.A. Rock, H.W. Jones Jr., The double uterus associated with an obstructed hemivagina and ipsilateral renal agenesis, *Am. J. Obstet. Gynecol.* 138 (1980) 339–342, [https://doi.org/10.1016/0002-9378\(80\)90260-4](https://doi.org/10.1016/0002-9378(80)90260-4).

- [4] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, SCARE Group, The SCARE 2020 guideline: updating consensus surgical CAse REport (SCARE) guidelines, *Int. J. Surg.* 84 (2020) 226–230, <https://doi.org/10.1016/j.ijssu.2020.10.034>.
- [5] G.F. Grimbizis, S. Gordts, Sardo A. Di Spiezio, S. Brucker, C. De Angelis, M. Gergolet, T.C. Li, V. Tanos, H. Brölmann, L. Gianaroli, R. Campo, The ESHRE-ESGE consensus on the classification of female genital tract congenital anomalies, *Hum. Reprod.* 28 (2013) 2032–2044, <https://doi.org/10.1093/humrep/det098>.
- [6] N. Güdücü, G. Gönenç, H. İşçi, A.B. Yiğiter, İ. Dündar, Herlyn-Werner-Wunderlich syndrome—timely diagnosis is important to preserve fertility, *J. Pediatr. Adolesc. Gynecol.* 25 (2012) e111–e112, <https://doi.org/10.1016/j.jpap.2012.05.013>.
- [7] G.B. Candiani, L. Fedele, M. Candiani, Double uterus, blind Hemivagina, and ipsilateral renal agenesis: 36 cases and long-term follow-up, *Obstet. Gynecol.* 90 (1997) 26–32, [https://doi.org/10.1016/S0029-7844\(97\)83836-7](https://doi.org/10.1016/S0029-7844(97)83836-7).
- [8] D.M. Albulescu, A.E. Ceașescu, L.M. Sas, M.C. Comănescu, C. Constantin, Ș. Tudorache, The Herlyn–Werner–Wunderlich triad (OHVIRA syndrome) with good pregnancy outcome – two cases and literature review, *Romanian J. Morphol. Embryol.* 59 (2018) 1253–1262.