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A Case Report of Oblique Vaginal Septum Syndrome with Difficulty in Defecation as the Initial Symptom

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Abstract

Oblique vaginal septum syndrome (OVSS) is a rare congenital disorder characterized by uterus didelphys, hemivaginal septum, and unilateral renal agenesis. Its clinical characteristics vary among different patients. Herein we report a case of OVSS with difficulty in defecation as the initial symptom and diagnosed with ultrasound and laparoscopic exploration. This 13-year-old girl was admitted to our hospital with the complaint of difficulty in defecation for more than 1 month and difficulty in urination for 6 days. Her ultrasound test showed that the left kidney was not displayed, and the right kidney was compensatory enlargement. A giant cystic-solid mass (110 mm × 96 mm × 87 mm) was seen in the pelvic cavity. Under laparoscopy, a huge mass was seen in the abdominal cavity. The surface resembled the seromuscular layer of the uterus. A uterine organ was seen on both sides of the tumor, and both fallopian tubes and ovaries were connected. The size of the right uterus was about 3 cm × 2 cm. The right fallopian tube and ovary were normal in appearance. The left uterus was enlarged (about 6 cm × 5 cm). The left fallopian tube was enlarged and contained a large amount of brown blood stains, and blood stains were seen flowing out from the umbrella end. The hysteroscopic incision and repair surgery were then performed. Most patients with OVSS presented with dysmenorrhea, abdominal pain, pelvic mass, and foul mucopurulent discharge, while in this case, difficulty in defecation was the initial symptom. Thus, this case report provided evidence supporting the heterogeneity of this disorder.

Keywords: Oblique vaginal septum syndrome; Laparoscopy; Hysteroscopy; Infertility; Ultrasonography

1. INTRODUCTION

Oblique vaginal septum syndrome (OVSS), also referred to as Herlyn-Werner-Wunderlich syndrome (HWWS) or obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome, is a rare congenital disorder characterized by uterus didelphys, hemivaginal septum, and unilateral renal agenesis [1]. It usually occurs in post-menarche female adolescents

with dysmenorrhea, irregular menses, abdominal pain, and pelvic mass [2]. The incidence of OVSS is very low, estimated to be between 0.4% to 6.7% among the general population according to previous reports [3, 4]. Due to the low prevalence, the diagnosis of OVSS is usually difficult. However, early diagnosis and early surgical resection of obstructing vaginal septum are critical to relieve the pain of patients and prevent other complications, such as endometriosis and infertility [5]. Thus far, our knowledge of this disorder is mainly based on case reports from different medical centers. Its clinical characteristics including the initial symptoms, diagnostic approaches, as well as surgical treatments vary among different individuals.

In this case report, we shared our experience with an OVSS patient who had difficulty in defecation as the initial symptom and was diagnosed by ultrasound and laparoscopic exploration as well as hysteroscopic incision.

2. CASE PRESENTATION

A 13-year-old girl was admitted to our hospital with the complaint of difficulty in defecation for more than 1 month and difficulty in urination for 6 days. More than 1 month ago, she had unresolved stools without obvious incentives, with a small amount, accompanied by abdominal pain; her urination was normal at that time. Her symptoms improved after oral administration of Berberine and compound *Lactobacillus acidophilus* tablets. However, she had difficulty in defecation again, accompanied by difficulty in urinating and abdominal pain, and was admitted to the outpatient department of our hospital for related examinations with urinary catheter intubation. She started menstruating when she was 12. Her physical examination revealed that the hymen was bulging downward and there was no vaginal opening. During anal diagnosis, a mass about 16cm×9cm in size could be palpated with mild tenderness. Her two-dimensional color Doppler ultrasound showed that the left kidney was not displayed (the congenital absence was not excluded), and the right kidney was compensatory enlargement (120 mm × 47 mm). A giant cystic-solid mass (110 mm × 96 mm × 87 mm) was seen in the pelvic cavity, with a clear boundary, liquid inside with echoes of small light spots, cloudy, and fluid level. A polycystic mass was seen above the mass, and there were many uneven separations within it, which may have originated from the left appendix. The size of the uterus was 38 mm × 28 mm × 19 mm, and the thickness of the endometrium was 4 mm. The right ovary was visible, and part of the left ovary was visible. We then suspected the patient had OVSS and surgical exploration was performed.

Under laparoscopy, a huge mass (about 12 cm×11 cm) was seen in the abdominal cavity (Figure 1A). The surface resembled the seromuscular layer of the uterus. A uterine organ was seen on both sides of the tumor, and both fallopian tubes and ovaries were connected. The size of the right uterus was about 3 cm × 2 cm. The right fallopian tube and ovary were normal in appearance. The left uterus was enlarged (about 6 cm × 5 cm). The left fallopian tube was enlarged and contained a large amount of brown blood stains, and blood stains were seen flowing out from the umbrella end. Adhesion of the left fallopian tube and the surrounding bowel could be seen. The left ovary appeared to be normal.

Under hysteroscopy, a cyst was seen in the vagina, but the cervix was not seen. Deep into the right side, the structure of the cervical canal was seen alternately with ravines. Deep

A Abdominal mass (under laparoscopy)**B** Puncture aspiration**C** Abdominal mass after aspiration (under laparoscopy)

Figure 1. Laparoscopic exploration and puncture aspiration of the patient. (A) Abdominal mass seen under laparoscopy. (B) Puncture aspiration. (C) After aspiration, the abdominal mass reduced seen under laparoscopy

into the right uterine cavity, a right fallopian tube opening could be seen in the right uterine cavity. The vaginal cyst was punctured, the non-coagulated fluid was drawn out (Figure 1B), and the pelvic mass was gradually reduced observed under laparoscopy (Figure 1C), which was considered to be the oblique vaginal septum. The oblique vaginal septum was incised and about 600 ml of fluid was completely released. The hysteroscope was penetrated into the oblique septum on the left side of the vagina and the cervical canal-like organ could be seen. The opening of the left fallopian tube could be seen when entering the left uterine cavity. The oblique vaginal septum was excised, and the incisional margin was sutured with a 2-0 tendon line.

Her postoperative diagnosis was vaginal oblique septum syndrome type 1 with the left kidney agenesis.

3. DISCUSSION

In this case with OVSS, the patient's initial symptom was difficulty in defecation, followed by difficulty urinating. Although routine laparoscopy is not essential to management of OVSS (PMID: 17320871), laparoscopic exploration was performed in this case for diagnosis clarification. Combined with ultrasound and hysteroscopic incision, the diagnosis of this patient was confirmed, and she was promptly treated.

OVSS syndrome is a rare malformation of the female genital tract in the clinic, manifesting as double uterus, double cervix, double vagina, complete or incomplete atresia of one vagina, and often accompanied by atresia lateral urinary tract malformation (unilateral

renal agenesis). Because during embryonic development, both the mesonephric duct and the paramesonephric duct originate from the urogenital ridge. Paramesonephric duct development is dependent on mesonephric duct development. Agenesis of one mesonephric duct affects the development of the ipsilateral paramesonephric duct, resulting in a series of malformations of the kidneys, ureters, uterus, and vagina.

OVSS syndrome can be divided into three types. Type 1 is a non-porous oblique partition. It is manifested as complete obstruction of the unilateral vagina, and the uterus behind the septum is completely isolated from the outside world and the contralateral uterus. There is no passage between the two uterus and between the two vaginas. The blood in the uterine cavity accumulates in the vaginal cavity after the septum. Type 2 is a perforated oblique septum. It manifests as incomplete atresia of one side of the vagina. There is a small hole in the oblique septum through which menstrual blood can drip, but drainage is partially blocked. Type 3 is a non-porous oblique septum combined with a cervical fistula. It is characterized by complete atresia of one side of the vagina. There is a small fistula between the two sides of the cervix or between the retroseptal vaginal canal and the contralateral cervix. The blood in the retroseptal cavity can be drained through the other side of the cervix, but it is partially blocked as well.

OVSS syndrome is easily misdiagnosed in the clinic. With a normal uterus and vagina on one side, patients often have normal menstrual cycles, masking the presence of obstruction. Early diagnosis can remove the obstructed oblique vaginal septum timely, thereby relieving symptoms and preventing complications, and preserving fertility. Without timely treatment, patients may develop pelvic endometriosis, pelvic adhesions, and pelvic infection. In severe cases, empyema and vaginal empyema may occur.

The clinical symptoms of oblique vaginal septum syndrome depend on its type. Type 1 patients mostly complain of dysmenorrhea, younger age of onset, and shorter time from menarche to onset. Types 2 and 3 mainly complain of vaginal purulent or bloody discharge. Most of the patients are virgins, and vaginal examination is rare. Such as vaginal examination can be seen on one side of the vagina has a small hole. There may be pus out. Vaginal wall masses can be palpated, and these masses are generally low-lying, unlike common pelvic masses, which are fixed on one side of the vaginal wall and fornix.

Ultrasonography can reveal urogenital malformations and corresponding obstruction images due to oblique vaginal septum. It is non-invasive and inexpensive, making it the preferred method of secondary examination. MRI examination can accurately display the anatomical structure of the genitourinary system at all levels, accurately distinguish uterine malformations, distinguish between the myometrium that is difficult to distinguish by ultrasound and the adnexal mass with hemorrhage, and directly show the oblique vaginal septum. However, it is more expensive and therefore less used as a diagnostic tool.

Surgery is the only effective treatment for OVSS. The purpose of surgery is to relieve symptoms and preserve fertility. Transvaginal oblique septum resection is the most ideal surgical method, and it is also the most effective and simple method to relieve genital tract obstruction. The vast majority of patients can be cured with oblique vaginal septumectomy alone. The key to the operation is to reduce the adhesion and atresia of the oblique septum incision.

The timing of surgery is better during the menstrual period, when the vaginal wall mass

is tense and easy to locate [6]. During the operation, the puncture is performed through the small hole in the capsule wall or the most prominent part of the vaginal mass. After the blood or pus was drawn, the vaginal septum was incised longitudinally along the needle until it was long enough, up to the fornix and down to the lowest point of the cyst, so that the drainage could be smooth. Zhu Renlie et al. [7] proposed that the side length of the excess diaphragm is 1.5–2.0 cm after diamond-shaped excision, and electrocoagulation to stop bleeding after excision. After fully exposing the retroseptal cervix, the rough surface of the incision edge was sutured intermittently with a catgut, and then the cyst cavity and the incision were filled with iodoform gauze, which could not only compress the hemostasis, but also prevent the incision retraction and adhesion.

Once the deformity is corrected in OVSS patients, their fertility is the same as that of normal women. Both sides of the uterus can have normal pregnancy and childbirth. However, there are also a small number of patients who experience miscarriage, embryo arrest, and ectopic pregnancy [8].

In summary, we present an OVSS case with difficulty in defecation as the initial symptom. Laparoscopic exploration and hysteroscopic incision were performed for further diagnosis clarification and treatments. Future studies focusing on the heterogeneity of this disorder are warranted.

References

- [1] Angotti R, Molinaro F, Bulotta AL, Bindi E, Cerchia E, Sica M, Messina M. Herlyn-Werner-Wunderlich syndrome: An "early" onset case report and review of Literature. *Int J Surg Case Rep.* 2015;11:59–63. doi: 10.1016/j.ijscr.2015.04.027. Epub 2015 Apr 24. PMID: 25932973; PMCID: PMC4446687.
- [2] Kabiri D, Arzy Y, Hants Y. Herlyn-Werner-Wunderlich syndrome: uterus didelphys and obstructed hemivagina with unilateral renal agenesis. *Isr Med Assoc J.* 2013 Jan;15(1):66. PMID: 23484247.
- [3] Saravelos SH, Cocksedge KA, Li TC. Prevalence and diagnosis of congenital uterine anomalies in women with reproductive failure: a critical appraisal. *Hum Reprod Update.* 2008 Sep-Oct;14(5):415–29. doi: 10.1093/humupd/dmn018. Epub 2008 Jun 6. PMID: 18539641.
- [4] Stampe Sørensen S. Estimated prevalence of müllerian anomalies. *Acta Obstet Gynecol Scand.* 1988;67(5):441–5. doi: 10.3109/00016348809004257. PMID: 3265253.
- [5] Piccinini PS, Doski J. Herlyn-Werner-Wunderlich syndrome: a case report. *Rev Bras Ginecol Obstet.* 2015 Apr;37(4):192–6. doi: 10.1590/SO100-720320150005077. PMID: 25992503.
- [6] Zhu Lan, Wang Shu, Lang Jinghe. Chinese expert consensus on the diagnosis and treatment of female genital malformations [J]. *Chinese Journal of Obstetrics and Gynecology*, 2015, 50(10).

- [7] Zhu Renlie. Diagnosis and management of oblique vaginal septum syndrome [J]. Chinese Journal of Practical Gynecology and Obstetrics, 1994, 10:4.
- [8] Ke Jianchu, Huang Wei. Clinical analysis of 7 cases of congenital oblique vaginal septum[J]. Journal of Practical Obstetrics and Gynecology, 2004, 20(6):371-372.