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Case Report

Herlyn-Werner-Wunderlich syndrome with double hematocolpos in one side hemivagina

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Abstract

Background: Herlyn-Werner-Wunderlich syndrome (HWWS) is an extremely rare Mullerian anomaly consisting of uterus didelphys with obstructed hemivagina and ipsilateral renal agenesis. Although few HWWS cases had been reported in the gynecological literature, most of the reported cases were combined with single hematocolpos. **Case**: We report here on a case of HWWS with double hematocolpos on one side of a hemivagina combined with ipsilateral renal agenesis. This is the first report of HWWS with double hematocolpos on one side of a hemivagina that was successfully treated with the resection of the vaginal septum. **Conclusion**: It is important to understand the pathogenesis and clinical features of these anomalies for diagnosis.

Keywords: Hematocolpos; Hereditary renal agenesis; Urogenital abnormalities; Herlyn-Werner-Wunderlich syndrome; Mullerian anomaly

1. Introduction

Herlyn-Werner-Wunderlich syndrome (HWWS) has been associated with the combination of uterine didelphys, blind hemivagina, and ipsilateral renal agenesis [1]. Providing appropriate treatment through precise identification is important. We report here on a first case of HWWS with double hematocolpos on one side of a hemivagina divided by the transverse septum.

2. Case report

An 11-year-old nulligravid girl who presented with the complaint of lower abdominal pain for a week was referred to our hospital for a suspicious uterovaginal anomaly. She had attained menarche six-month before, after which she had had regular menstrual cycles every 25 days with a normal menstrual flow and without significant dysmenorrhea. Medical, surgical, and social histories were unremarkable. On pelvic exam, the external genitalia appeared grossly normal. The hymen was patent, and pubic hair was tanner scale IV. On rectal exam, a tense fluid collection suggestive of hematocolpos was palpable within the presumed vaginal canal, extending inferiorly to 3 cm above the introitus.

An abdominal ultrasonography revealed that she had a double uterus, double cervix, and a longitudinal vaginal septum; an elongated lobulated lesion with thickened walls extended from the perineal surface into the pelvic cavity in close proximity to the uterus (Fig. 1A). Her right kidney was normal, however the left kidney was absent. Magnetic resonance imaging of the pelvis and kidney, done to confirm the ultrasound findings, revealed that there were two vaginal septums (Fig. 1B). A 2.3-mm thick transverse vaginal septum confining a 4.9 × 3.3 cm hematocolpos and a 4.4-mm thick oblique vaginal septum confining a 9.0 × 3.2 cm hematocolpos were seen in the left hemivagina (Fig. 1C). HWWS with a double hematocolpos on one side of the hemivagina was diagnosed (Fig. 2B). On the laboratory test, electrolytes including renal function test were within the normal range.

Resection of the vaginal septum was chosen for the evaluation and correction of the uterovaginal anomaly. The adhesiolysis of the transverse septum and resection of both vaginal septums were done, and the edges of the septums were marsupialized. The oblique vaginal septum was thick and completely obstructed, whereas the transverse vaginal septum was thin and had a pinpoint hole on the medial side. Blood flowed through the tiny hole in the transverse septum to the lower hematocolpos. After surgery, HWWS with a double hematocolpos on one side of the hemivagina was confirmed. The pathological results of the oblique vaginal septum were squamous epithelial hyperplasia with focal erosion and granulation tissue, and the transverse vaginal septum was squamous metaplasia of the endocervix.

The patient was comfortable in the postoperative period and was discharged two days after the operation. She then had regular menstruation, and no dysmenorrhea was combined. She became absolutely asymptomatic one month later.

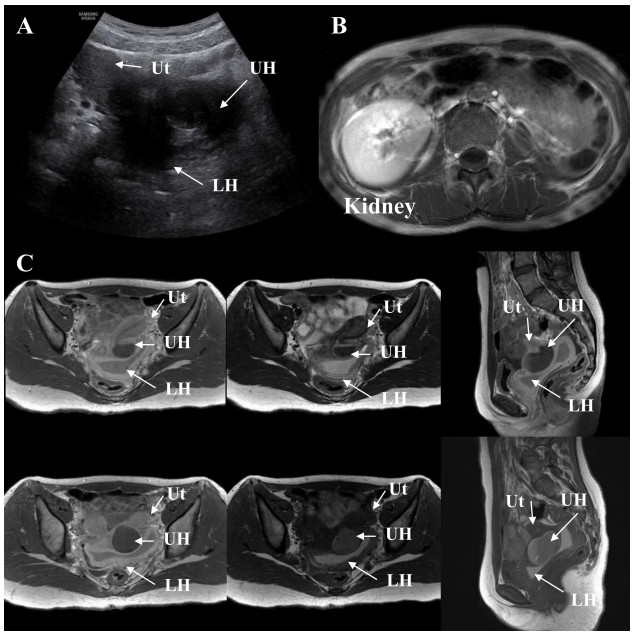

Fig. 1. Image findings. (A) Sonographic finding. Left-side uterus and double hematocolpos are shown. (B) MRI finding. The single right kidney is shown. No other urinary tract abnormalities except the ipsilateral renal agenesis were noted. (C) MRI finding. A double uterus, double cervix and two vaginal pouches are shown. The upper vaginal pouch containing an old hematoma connected with the left-side uterus about 4.9 × 3.3 cm in size. The lower vaginal pouch containing a subacute stage hematoma is wrapping the upper vaginal pouch. Ut, uterus; UH, upper hematocolpos; LH, lower hematocolpos; HWW, Herlyn-Werner-Wunderlich.

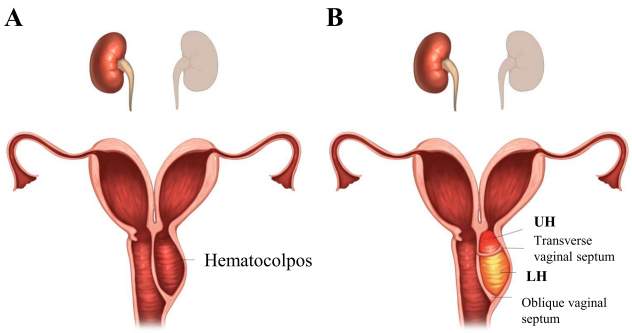

Fig. 2. Schematic representation. (A) Schematic representation of usual HWW syndrome. Single hematocolpos is presented. (B) Schematic representation of this case. Double hematocolpos are shown in one side hemivagina. Ut, uterus; UH, upper hematocolpos; LH, lower hematocolpos; HWW, Herlyn-Werner-Wunderlich.

3. Discussion

HWWS is an extremely rare congenital female reproductive malformation syndrome that is characterized by a triad of uterine didelphys, blind hemivagina, and ipsilateral renal agenesis also known as obstructed hemivagina and ipsilateral renal anomaly; OHVIRA (Fig. 2A) [1,2]. Although previous HWWS case series with vertical fusion or transverse canalization defects have reported, there have been no cases with this double hematocolpos combined HWWS have been reported. In this study, the extremely rare case of HWWS with double hematocolpos on one side of a hemivagina was successfully treated by resection of the vaginal septum.

Several case reports of HWWS [3,4] have reported a hematocolpos in both vaginas; however, no HWWS with two hematocolpos in a hemivagina had been observed. Himadri *et al.* [3] reported that a case of HWWS was composed of a hematotrachelos and hematometra in the left side of the uterus. Deligeoroglou *et al.* [4] reported a case of combined unicornuate uterus, imperforate hymen, and transverse vaginal septum which suggested agenesis of one half of the Mullerian ducts, resulting in a unicornuate uterus and not a vertical fusion defect. However, our case arose from failures of vertical fusion and canalization at the level of the hemivagina, unlike previous studies [3,4]. There is a rare congenital anomalies of female reproductive organs; hematocolpos caused by transverse septum and hymen [4]. Thus, this study is the first report on double hematocolpos caused by transverse septum and oblique septum in one side hemivagina that has never been reported so far.

The age at diagnosis of HWWS has decreased as neonatal screening tests have increased, however, many patients were still diagnosed with symptoms of hematocolpos after menarche began. Because menstruation is mostly regular in cases of HWWS, early diagnosis is difficult. Delayed diagnosis may induce progression of the disease to pyohematocolpos or pyosalpinx, pelvic peritonitis in severe cases [5]. Furthermore, in pediatric patients with HWWS, diagnosis is especially important because a protective approach to a single kidney is required. Therefore, early recognition and diagnosis are important. When a single kidney is first discovered during newborn screening with ultrasound, HWWS should also be considered [6–8].

This extremely rare case, HWWS with double hematocolpos on one side of the hemivagina, was successfully treated with surgery. It is important to understand the pathogenesis and clinical features of these anomalies for diagnosis.

Author contributions

YJK, JBY, YWJ and HJY designed this clinical study. YJK and SYS performed the case review. GWL provided help and advice on the clinical report. YJK analyzed the data. All authors contributed to editorial changes in the manuscript. All authors read and approved the final manuscript.

Ethics approval and consent to participate

The patient gave her informed consent for this case report before her medical chart was reviewed. This case report was conducted in accordance with the Declaration of Helsinki. Study's registration and ethics committee approval were unnecessary due to the nature of the study.

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Conflict of interest

The authors declare no conflict of interest. HJY is the Editorial board member of this journal, given his role as Editorial board member, HJY had no involvement in the peer-review of this article and has no access to information regarding its peer-review. Full responsibility for the editorial process for this article was delegated to Michael H. Dahan.

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