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CLINICAL VINGNETTE

Dysmenorrhea in OHVIRA syndrome

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INTRODUCTION

In literature, the OHVIRA syndrome (Obstructed HemiVagina and Ipsilateral Renal Anomaly) is also referred to as Herlyn–Werner–Wunderlich (HWW) syndrome or the “nameless” syndrome. It is a rare congenital malformation which affects less than 1 in 1,000,000 girls. It is characterized by uterine didelphys, obstructed hemivagina, unilateral renal agenesis (Fig. 1) [1, 2].

The most common symptoms of OHVIRA include pain which increases during menstruation despite the visible flow of menstrual blood and later becomes constant, predominantly unilateral. The pain does not respond to oral contraceptives or analgesics [2].

CASE REPORT

A 12-year-old female accompanied by her mother presented during her period with severe lower abdominal pain. The pain had been present for 2– 3 days without any additional complaints. The girl was undergoing antibiotic therapy for a suspected urinary tract infection

(UTI). The patient had been menstruating regularly for 2 years, with normal bleeding; dysmenorrhea had been present for 3 months.

An ultrasound examination revealed: two corpus uteri — the left one was 26 x 47 mm, with endometrium 8 mm, and the right one 40 x 24 mm with an enlarged (blood-filled) uterine cavity (hematometra); each uterine corpus had an anatomically correct ovary and a uterine cervix. A well-defined, smooth isoechoic lesion of 86 x 75 mm was found adjacent to the right uterine corpus, consistent with hematocolpos. The right kidney was absent (Fig. 2–4).

The patient was referred to the Gynecological Ward at the Holy Trinity Hospital in Plock to undergo surgery. The procedure involved an incision of the occluded vagina to release the accumulated menstrual blood. The incision was marsupialized with single sutures and a foley catheter was placed into the right vagina. The drain was removed after 4 days. The patient was discharged home without any complaints. Two years after the procedure, the patient remains in good condition, she menstruates regularly and painlessly.

CONCLUSION

The OHVIRA syndrome results from abnormal formation of the Müllerian and Wolffian ducts during the fetal stages of development [3]. First-line treatment consists of a vaginal access procedure involving the incision and marsupialization of the occluded vagina. Incorrect treatment may lead to retrograde blood flow through the fallopian tubes into the abdomen (retrograde menstruation) resulting in endometriosis and infertility [4, 5]. There is insufficient data regarding the fertility in women with OHVIRA. In literature, there is one case report discussing a pregnant patient who earlier underwent the marsupialization of the occluded right vagina. In this case, the pregnancy located in the right uterus progressed normally and concluded in a vaginal delivery in the 36th week of gestation. The premature labor was probably due to the small size of the uterus [2, 6].

Article information and declarations

Ethics statement

Ultrasoud examination and surgery procedure were performed as integral parts of routine clinical care for which informed consent had been previously given by mather' s patient. Data were anonymized.

Author contributions

Kinga Ksiezakowska-Lakoma — concept, assumptions, corresponding author, analysis and interpretation of data, article draft; Malgorzata Soroka — acquisition of data, revised article critically; Marek Brzezinski — analysis and interpretation of data, revised article critically.

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None.

Conflict of interest

None.

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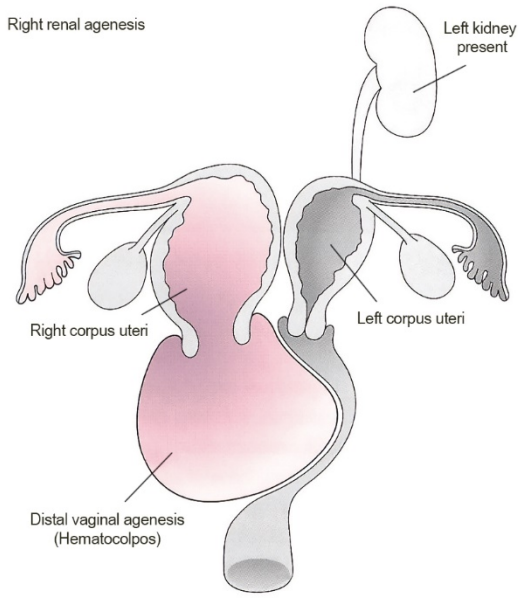

Figure 1. Schematic representation of OHVIRA syndrome

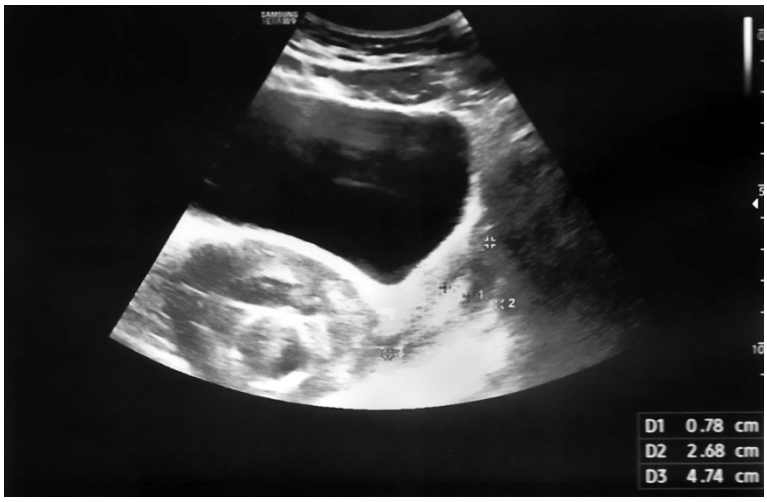

Figure 2. The Left corpus uteri

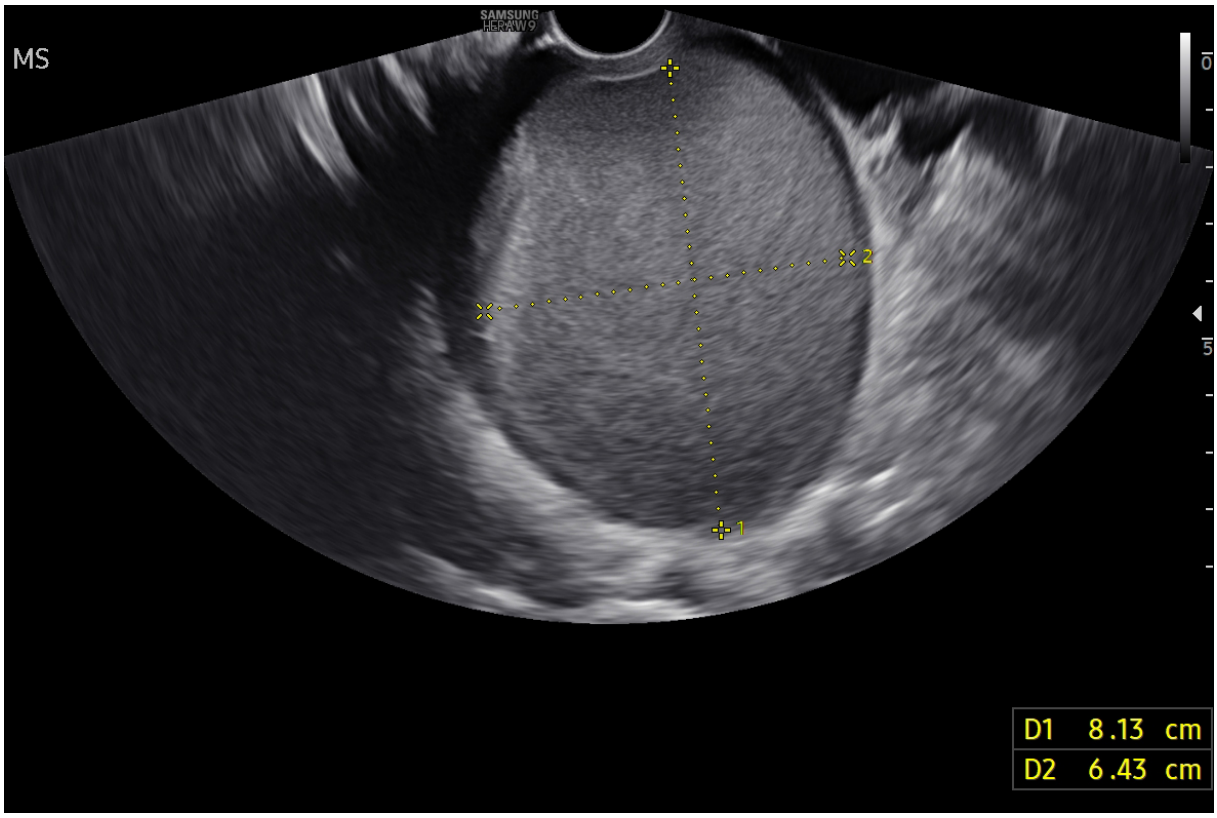

Figure 3. Hematocolpos

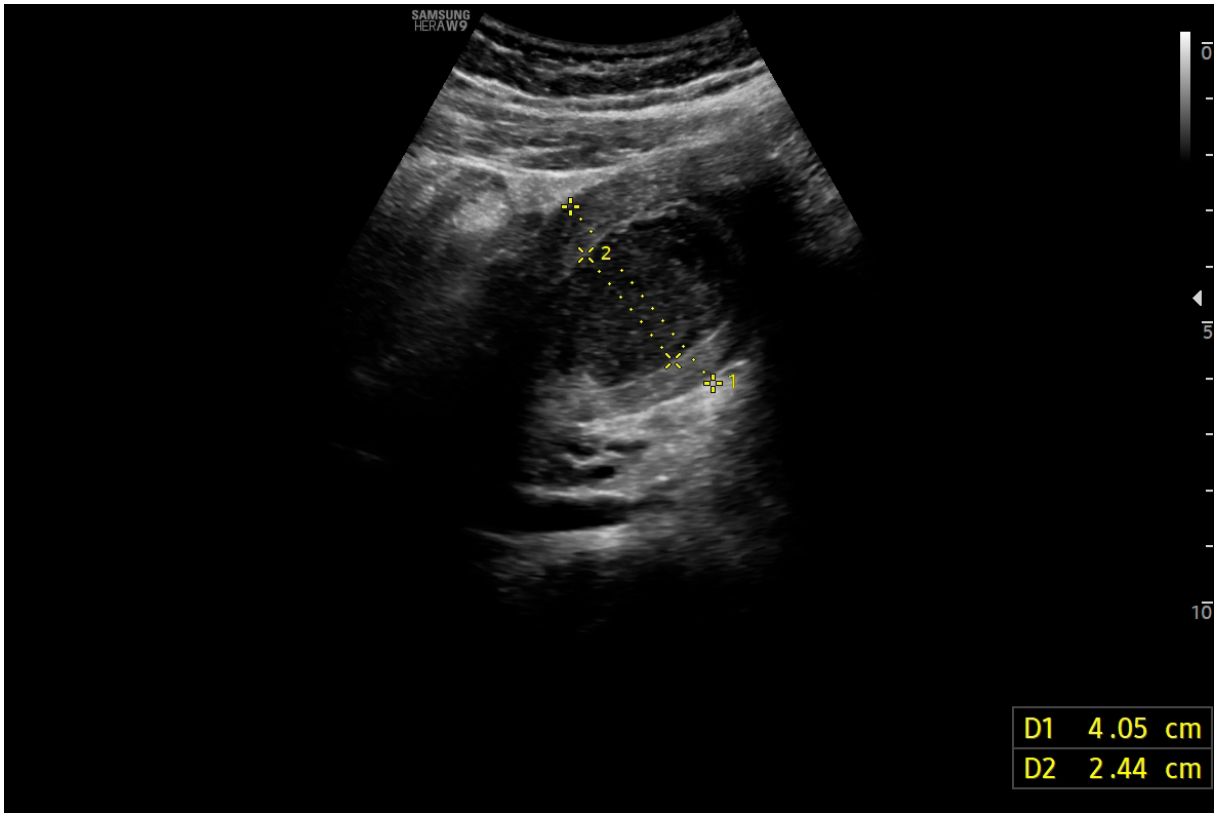

Figure 4. The Right corpus uteri