Journal of Bursa Faculty of Medicine
e-ISSN:

Case Report

Obstetrics and Gynecology

Received
December 18, 2022
Accepted
December 18, 2022
Published online
May 29, 2023

J Bursa Med 2023;1(1) 65-68

Ovarian hyperstimulation syndrome developed in a patient who underwent neovagen with the diagnosis of Müllerian agenesis and was a candidate for uterine transplantation: Case Report

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ABSTRACT

We aimed to present OHSS that developed after OPU in a patient who underwent neovagen 10 years ago with the diagnosis of Müllerian agenesis and was awaiting uterine transplant. We wanted to draw attention to successful approaches in such groups of patients with Müllerian agenesis.

A 35-year-old sexually active patient who applied to the Gynecology and Obstetrics Emergency Service of our institution with the complaint of abdominal pain had zero gravida. In her anamnesis, a 15-year-old patient who applied to the obstetrics clinic with the complaint of primary amenorrhea was diagnosed with MRKH syndrome in an external center. Neovagen was performed by laparoscopic method due to vaginal agenesis of the patient in an external center in 2012. The patient was informed and admitted to the gynecology service for follow-up with the diagnosis of mild OHSS.

The patient was informed and admitted to the gynecology service for follow-up with the diagnosis of mild OHSS. During the follow-up period, waist circumference and weight did not increase, the follow-up was balanced, no free fluid was observed in the abdomen in the ultrasonography, there was no electrolyte imbalance, there was no deterioration in liver-kidney functions and hemoconcentration did not develop. The patient was discharged on the 3rd day of hospitalization due to the regression of abdominal pain.

OHSS may be one of the difficulties experienced by all Müllerian agenesis patients who decide to become a real family. These patients should be given up-to-date information about neovagen production, IVF processes and uterine transplantation by all obstetricians. Considering all of these approaches will raise awareness in this patient population, which faces a variety of lifelong challenges in starting a family.

Keywords: ohss, neovagen, müllerian agenesis, uterine transplatation

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is a congenital disorder characterized by agenesis of the müllerian ducts (uterus, cervix, and upper two-thirds of the vagina).

The external genitalia and secondary sex

How to cite this article

Bilgin Yüce E, Gumus Satici Y, Koca E, Yenigul NN. Ovarian hyperstimulation syndrome developed in a patient who underwent neovagen with the diagnosis of Müllerian agenesis and was a candidate for uterine transplantation: Case Report J Bursa Med 2023;1(2):65-68

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characteristics of these 46, XX women are normal [1]. MRKH syndrome occurs in approximately one in 4500 live births and is the second most common cause of primary amenorrhea after gonadal dysgenesis [2]. It has been reported to affect approximately 10% of women with primary amenorrhea. [2-3]. It may be associated with renal, cardiac, skeletal and other anomalies. These patients have been able to become sexually active for many years thanks to neovagen operations. Especially with the developments in endoscopic surgery, neovagen techniques have been advanced and this has given hope to women with this diagnosis.

Until recently, legal adoption was the only option for women with MRKH syndrome. Uterine transplantation gave hope to these women. Uterine transplantation performed on a patient with mullerian agenesis performed by Ozkan and friends in 2013 changed a lot [4]. Now these women have the chance to have their own children with embryo transfer after uterine transplant.

Pharmacological ovarian stimulation has been used as the gold standard since the beginning of in vitro fertilization (IVF). Ovarian Hyperstimulation Syndrome (OHSS), which was defined in the 1930s; It is a syndrome that can develop during ovulation induction or rarely in the natural cycle, is characterized by massive ovarian growth and multiple ovarian cysts, and progresses with electrolyte disturbance and protein loss as a result of excessive steroid hormone and capillary permeability [5]. Of those who develop OHSS, the incidence of which has increased in recent years, clinically significant ones are 2-3%, but milder forms of OHSS develop in 20-30% of all IVF patients [6]. Agents such as clomiphene citrate (CC), human menopausal gonadotropin (hMG), follicle stimulating hormone (FSH), gonadotropin-releasing hormone (GnRH), GnRH analogs, human chorionic gonadotropin (hCG), which are used for ovulation induction, are held responsible for the development of OHSS.

In this article, we aimed to present OHSS that developed after oocyte pick up (OPU) in a patient who underwent neovagen 10 years ago with the diagnosis of Müllerian agenesis and was awaiting uterine transplant. We wanted to draw attention to successful approaches in such groups of patients with Müllerian agenesis.

CASE

A 35-year-old sexually active patient who applied to the Gynecology and Obstetrics Emergency Service of our institution with the complaint of abdominal pain had zero gravida. In her anamnesis, a 15-year-old patient who applied to the obstetrics clinic with the complaint of primary amenorrhea was diagnosed with MRKH syndrome in an external center. Neovagen was performed by laparoscopic method due to vaginal agenesis of the patient in an external center in 2012. In her anamnesis, it was learned that oocyte pick-up was performed in an external center 4 days ago, 20 oocytes were collected and the patient was during uterine transplantation in another center.

The vulva and vagina examination performed by us was normal. Vaginal length was 3 centimeter (cm), cervix was not observed. Bilateral ovarian dimensions were slightly increased in transvaginal ultrasonography (right ovary 82 millimeter (mm) (Figure 1), left ovary 72 mm (Figure 2)), and no uterus was observed. There was no free fluid in the abdomen. There was no sign of acute abdomen in the physical examination. The patient's weight was 67 weight (kg) and waist

Figure 1. Right ovary

Figure 2. Left ovary

circumference was 71 cm. In the pelvic MRI of the patient, a suspicious appearance of uterine tissue (rudimentary uterus?) measuring 15x8 mm was observed at the level of the uterine lodge. In addition, bilateral kidneys were normally located and no accompanying renal anomaly was observed. In the blood tests studied from the patient, hemogram 13.1 g/dl, leukocyte 14.680/mcl, hematocrit 38.6%, beta hcg 10.9 mIU/l, creatinine 0.6 mg/dl, AST 13 U/l, ALT 9 U/ l and it was observed that there was no electrolyte imbalance.

The patient was informed and admitted to the gynecology service for follow-up with the diagnosis of mild OHSS. Daily waist circumference, weight, ultrasonography, complete blood count, liver-kidney function test, electrolytes and intake and output were followed up. Thromboprophylaxis was administered during hospitalization. During the follow-up period, waist circumference and weight did not increase, the follow-up was balanced, no free fluid was observed in the abdomen in the ultrasonography, there was no electrolyte imbalance, there was no deterioration in liver-kidney functions and hemoconcentration did not develop. The patient was discharged on the 3rd day of hospitalization due to the regression of abdominal pain. The examinations and tests performed on the 3rd, 10th and 15th days after discharge were found to be normal.

DISCUSSION

OHSS is the most serious complication of controlled ovarian hyperstimulation. Exogenous human chorionic gonadotropin (hCG) administration plays an important role in the pathogenesis of OHSS. Regardless of the degree of ovarian response to gonadotropin stimulation, OHSS does not occur unless an ovulatory dose of hCG is administered [7,8]. Exogenous hCG induces intense luteinization of granulosa cells leading to the production of vasoactive substances such as VEGF that increase vascular permeability. Such massive luteinization is not usually observed when the final steps of oocyte maturation are achieved with drugs other than hCG (eg, gonadotropin-releasing hormone (GnRH) agonists) [9].

The diagnosis of OHSS is made by clinical history and transvaginal ultrasound. There should be a history of ovarian stimulation after hCG administration. Early OHSS is usually mild to moderate and begins four to seven days after a human hCG dose to induce ovulation [10]. To the best of our research, we could not find a case in the literature who had undergone neovagen surgery due to mullerian agenesis and was diagnosed with OHSS while waiting for the uterine transplant. We think that this case report, which is a first in this sense, is important to draw attention to uterine transplantation. The effect of the presence of the uterus or the presence of a rudimentary horn on the occurrence or course of OHSS is unknown. With the increase in transplant success in these patient groups, IVF cycles may increase over time and clearer data can be generated.

As a result of problems such as inability to have sexual intercourse and absolute uterine infertility seen in patients with MRKH syndrome; It is stated that psychosocial problems such as low self-esteem, lack of self-confidence, feeling of worthlessness, and fear of rejection by the opposite sex occur [11, 12]. Studies have shown that patients diagnosed with MRKH Syndrome experience negative emotions such as reactive depression, shock, and suicidal thoughts [13], and they feel different and incomplete than other women who experience sexual identity threat and loss of sexual and social roles in the future [14]. Therefore, the psychological and medical approach to patients with Müllerian agenesis has become more important in recent years. Thanks to assisted reproductive techniques before uterine transplantation that can be performed in patients with Müllerian agenesis, with the developing technology and new treatment methods, oocyte collection and embryo freezing has become a new hope for the fertility of these patients.

It is a very difficult process for these patients with absolute uterine factor infertility to decide on uterine transplantation [15]. These patients, on the one hand, come under a very serious financial burden with this decision, and on the other hand, they take the decision to create their own families by taking serious personal risks. In this sense, they should receive serious training and know the risks that may occur in detail [15]. Recent studies have shown that despite all the difficulties, women in this patient group will want to prefer uterine transplantation as an alternative to surrogacy or adoption [15, 16].

This case report reveals that OHSS may be one of the difficulties experienced by all Müllerian agenesis patients who decide to become a real family. These patients should be given up-to-date information about neovagen production, IVF processes and uterine transplantation by all obstetricians. Considering all of these approaches will raise awareness in this patient population, which faces a variety of lifelong challeng-

es in starting a family.

CONCLUSION

Authors' Contribution

Study Conception: NNY,; Study Design: NNY,; Supervision: NNY,; Materials: EK,; Data Collection and/or Processing: YSG,; Statistical Analysis and/or Data Interpretation: EYB,; Literature Review: YSG,; Manuscript Preparation: EYB and Critical Review: EK

Conflict of interest

No potential conflicts of interest relevant to this article were reported.

Financing

There is no source of financial support or funding.

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