Case Report

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Distal Vaginal Atresia Combined with Unicornoate Uterus: A Case Report

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Running Title Distal Vaginal Atresia Combined with Unicornoate Uterus

ABSTRACT

Congenital absence of the vagina with variable uterine development known as Mullerian agenesis. We presented the case of a patient with distal vagina agenesis with presence of proximal vagina and functioning unicornoate uterus.

A 13-year-old patient was referred to pelvic floor service due to primary amenorrhea and pelvic pain. She was diagnosed with the distal vaginal agenesis, functional unicornoate uterus and the ectopic right kidney.

Rectoabdominal examination revealed segmental vaginal agenesis and a likely atretic cervix with a huge abdominopelvic mass of about 10 cm in size. On exploratory laparotomy, the bladder was completely dilated and the right kidney was ectopic and both kidneys had moderate hydronephrosis. On the left, a unicorn uterus containing blood and clot, and on the right, a non-functional rudimentary horn with a normal fallopian tube was seen. A neovagina was created by dissection of the space between the urethra hyatus and rectum in laparatomy.

It is important to note that the patient may initially present with urinary symptoms and renal signs. Therefore, in the symptoms of urinary tract obstruction, mullerian anomalies should be considered.

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Introduction

Vaginal agenesis, also known as Mullerian malformations, refers to the congenital absence of the vagina with variable uterine formations [1]. These anomalies occur with the prevalence of 6.7% in the general population, 7.3% in the infertile population and 16.7% in the recurrent miscarriage population [2]. Patients with vaginal agenesis can have a normal karyotype (46, XX) with female phenotype, normal ovaries, and normal secondary sexual development other than menarche [3]. The case described in this paper is a new female genital anomaly with a combination of vaginal atresia and unicorn uterus with rudimentary horn.

Case presentation

A 13-year-old girl was seen at the gynecology outpatient pelvic floor clinic of Taleghani Hospital,

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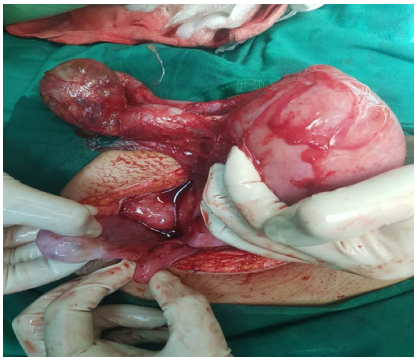

Fig. 1. Left unicornoate uterus and cervix

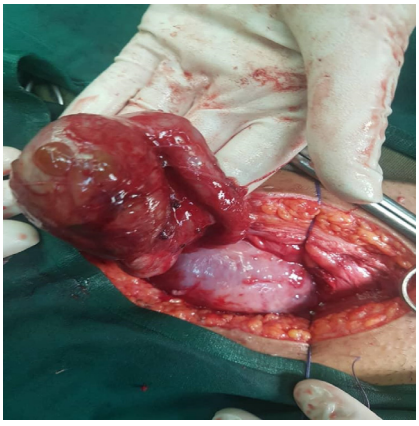

Fig. 2. Right fallopian tube contains hemorrhagic products

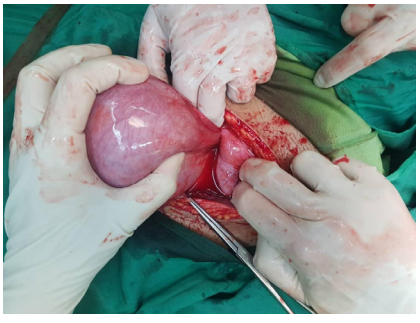

Fig. 3. View of Uterus, both fallopian tubes and non-functional rudimentary horn

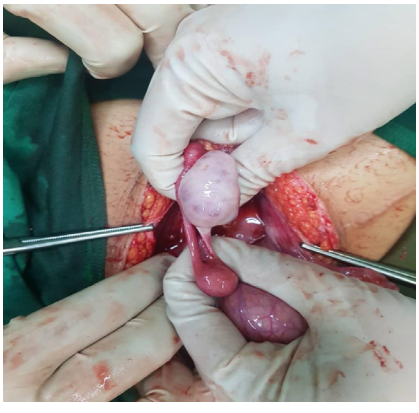

Fig. 4. Right non-functional rudimentary horn and normal ovary

due to primary amenorrhea and pelvic pain. The patient had been referred to the urology service with the same complaint previously. Ultrasound reported a thick-walled cystic mass with a dense accumulation in the midline of the pelvis; also, severe hydronephrosis in both of the kidneys and distention of bladder had been reported. She had previously been tested for kidney function and renal santigraphy for GFR had reported normal perfusion and function of the left kidney but moderately decreased perfusion of the right ectopic kidney with dilated PCS. The MRI showed the proximal segment of the vagina, the uterus and the right fallopian tube were severely distended and contained hemorrhagic products. The bladder was distended and its lower segment was compressed with distended vagina. The ovaries showed normal appearance. The available findings indicated the presence of hematometrosalpynx and cross ectopic right kidney with obstructive uropathy. On physical examination, the patient had adequate secondary sexual characters development and normal external genitalia. Rectoabdominal examination under general anesthesia revealed segmental vaginal agenesis and a likely atretic cervix with a huge abdominopelvic mass of about 10 cm in size. On exploratory laparotomy, the bladder was completely dilated and extended to the umbilical cord. The right kidney was ectopic and both kidneys had moderate hydronephrosis. On the left, a unicorn uterus containing blood and clots measuring 10 cm and obstructed fimberia were seen, and on the right, a non-functional rudimentary horn with a normal fallopian tube was detected. There was no adhesions or endometriosis inside the abdomen. The cervix and the 2/3 of the proximal vagina were severely dilated. A neovagina was created by the dissection of the space between the urethra hyatus and rectum in laparatomy. Neovagina surgery was performed by inserting a vaginal mould. The uterus was larger than normal, so the hematometra was discharged. Laparatomy findings indicated the presence of a unicorn uterus, agenesis of one-third of the distal vagina and non-functional rudimentary horn on the right side. Salpingectomy was performed due to hydrosalpinx and severe adhesion of the left fallopian tube to prevent ectopic pregnancy. The left unicornoate uterus was functional and the ovaries were normal.

Discussion

Paramesonephric ducts shape the female genital tract and any deviation from the normal is Mullerian anomalies[2]. We reported a case of the distal one-third of vagina agenesis and unicornoate uterus, presented as pelvic pain even though the patient had normal growth and secondary sexual characteristics. Distal vagina atresia can cause hematometra, hematosalpinx, endometriosis as well as pelvic infection if diagnosis is delayed [4]. We described a case of a genital anomaly that had not been reported in the literature before. The classification systems of the American Fertility Society do not mention this condition [5]. In this case, a combination of several anomalies was observed that have not been categorized in the Müllerian anomaly classifications. That patient had distal vagina atresia, unicornoate uterus with non-communicating rudimentary horn and ectopic right kidney. The unicornoate uterus with non-communicating rudimentary horn can be classified as class IIc [6]. The uterus, proximal part of the vagina and the fallopian tubes originated from the paramesonephric ducts, and the origin of the distal vagina is the urogenital sinus [7].

The accompanied anomalies, which usually consist of renal and skeletal malformations, were reported 40% and 20%, respectively [1]. The current case had a normal female genotype and phenotype. MRI revealed right ectopic kidney, which is fused to the left kidney, but there were no skeletal abnormalities. The best treatment for vaginal agenesis is still controversial [8]. In the USA and UK, vaginal dilation therapy is considered the first-line treatment and surgery is suggested for the cases who are not appropriate, or when vaginal dilation therapy fails. In most of the European countries, surgical vaginoplasty is the first line treatment, and vaginal dilation is only used post-operatively [9]. In this case, a neovagina was created by the dissection of the space between the bladder and the rectum. This approach is based on the McIndoe procedure. The procedure involves a perineal approach to create a space between the rectum and urethra, followed by the use of a split-thickness skin graft to form the epithelium of the neovagina. The patient is then required to dilate the vagina postoperatively to prevent stenosis. Good anatomical results have been reported [9]. It is important to note that the patient may initially present urinary symptoms and renal signs. Therefore, in the symptoms of urinary tract obstruction, Mullerian anomalies should be considered in order to prevent kidney damage due to the compressive effect of hematometers, hematosalpynx, etc. through appropriate and timely action. In conclusion, the present case is important for clinical management and emerges as a novel anomaly that supports one embryologic concept. It may be necessary to determine the anomalies during operation. Furthermore, it is important for us to observe the patient well, regarding reproductive and obstetric outcomes in future.

Ethical Considerations

Compliance with ethical guidelines

There were no ethical considerations to be considered in this article.

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Conflict of Interests

The authors have no conflict of interest to declare.

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