Uterus Unicornis and Unilateral Ovarian and Renal Agenesis in A Case with Primary Amenorrhoea✍

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The purpose of this study is to report a case of primary amenorrhoea, uterus unicornis and unilateral ovarian and renal agenesis. A 17-year-old girl was referred to our university hospital because of primary amenorrhoea. Secondary sex characters were concordant with her age, hormonal tests were normal and the rectal examination identified a small uterus. An abdominopelvic ultrasound and pelvic magnetic resonance imaging were performed but could not detect uterus, endometrial line and both of the two ovaries. A chromosome karyotype analysis showed a normal 46, XX. A laparoscopy had confirmed the diagnosis of a unicornuate uterus with no evidence of a rudimentary horn and one tube, one round ligament and one normal ovary on the left side. Agenesis of the right kidney was found on postoperative intra venouse pyelography. Estradiol hemihydrate 2 mg estrogen orally once per day 21 days and after 10 mg medroxyprogesterone acetate was initiated for 3 months. Although their causes are controversial, they can be created by a localized defect at the caudal section of the Mullerian canal and genital ridge area or an adnexal torsion during intra-uterine period.

Key Words: Primary amenorrhoea, Uterus unicornis, Ovarian and renal agenesis

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Introduction

Primary amenorrhea can be diagnosed if a patient has normal secondary sexual characteristics but no menarche by 16 years of age. If a patient has no secondary sexual characteristics and no menarche, primary amenorrhea can be diagnosed as early as 14 years of age and its insidence ranges between 0.1% and 2.5%. One of the most important reasons for primary amenorrhoea is mullerian agenesis. It is seen in approximately 16% of cases with primary amenorrhoea.[1,2] During the embryonic development of the female fetus, with the joining of mullerian canals inside the abdomen, the uterus, tubes and the upper vagina are formed. Because congenital abnormalities of the uterus are asymptomatic, they are usually diagnosed late. The anomalies of the uterus are seen at a rate of 2-4 % at all reproductive ages.[3] Uterus unicornis is seen at a rate of 10 % among all uterovesical anomalies.[3] Cases with uterus unicornis accompanied by unilateral ovarian agenesis are very rare in the literature.[4-6] Due to the same embryonic origin, mullerian system defects usually occur together with urinary system anomalities. Thus, in cases with uterus unicornis accompanied by unilateral ovarian agenesis, pelvic kidney or renal agenesis are also observed.[4,5]

We report here a case with primary amenorrhoea and uterus unicornis with unilateral ovarian agenesis under the literature support.

Case Report

A-17 year-old girl was referred to our university hospital because of primary amenorrhoea. The patient had had a 3-months history of using oral contraceptive (Gynera tb, 1x1) without evidence of menses. There were no further specifications in the background or family history. Secondary sex characters were concordant with her age and hormonal tests were normal. Gynecological examination shows that her breasts and pubic and axillary hair are Tanner stage 4 with no galactorrhea. The hymen was intact, the major and minor labia, urethral meatus and vaginal introitus were normal and the rectal examination identified a small uterus. Laboratory tests included FSH levels: 1.94 mIU/ml, LH levels: 1.65 mIU/ml ve E2 levels: 50.46 pg/ml. The other biochemical laboratory tests were within normal limits. An abdominopelvic ultrasound and pelvic magnetic resonance imaging were performed but could not detect uterus, endometrial line and both of the two ovaries. A chromosome karyotype analysis showed a normal 46, XX. At laparoscopy, she was found to have unicornuate uterus

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without any rudimentary horn. The right fallopian tube, round ligament, and ovary were all absent The left fallopian tube, round ligament, and ovary were all normal. An implantation focus belonging to intra-abdominal endometriosis was not detected. Any trace of an old bleeding of the menses inside the abdomen was not detected (Figure 1). Agenesis of the right kidney was found on postoperative intra venouse pyelography (Figure 2). Estradiol hemihydrate 2 mg estrogen (Estrofem, Novo Nordisk) orally once per day 21 days and after 10 mg medroxyprogesterone acetate (Farlutal, Deva) was initiated, through 3 months. Later it was planned to evaluate the patient under hysteroscopy in terms of vaginal and/or cervical agenesis and she was discharged from hospital.

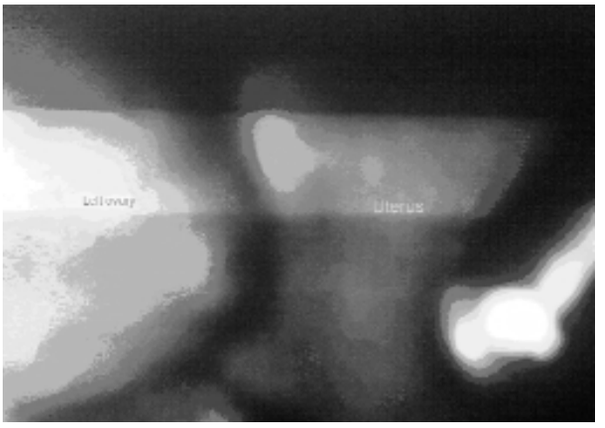

Figure 1: A laparoscopic view of the left ovary and uterus.

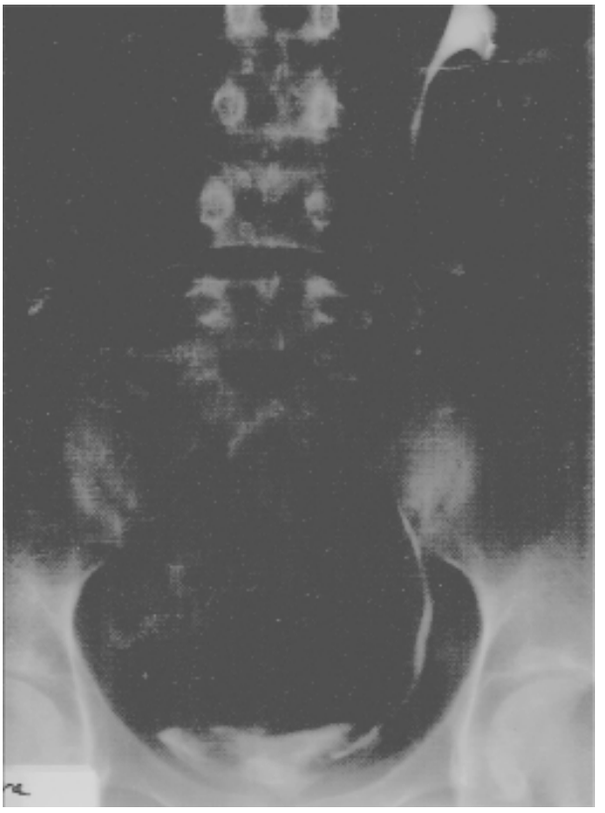

Figure 2: Postoperative intravenous pyelography showed

Discussion

A classification proposed by Buttram and Gibbons in 1979 based on the degree of normal development disorder of the female genital system was modified by the American Reproductive Medicine Association in 1988. According to this classification, with the absence of contra-lateral rudimentary horn and because of the existence of uterus unicornis, the case was evaluated as Class II. The laboratory results including FSH and LH were found to be lowered in this case. However, we didn't consider as hypogonadotopic hypogonadism because secondary sexual characteristics such as pubic, facial, and underarm hair, normally development at puberty and ability to smell were present in this case.

There are many publications indicating that mullerian anomalities are observed together with urinary system anomalities. However, cases with Mullerian anomalities observed together with unilateral ovarian agenesis are rare. A review of the literature about the causes reveals three different mechanisms. The first is non ascended ovary[7] and with these kinds of patients, it has been reported that when over stimulation is carried out with clomiphene citrate, the visualization of the non ascended ovary can be achieved. However with our patient 5-9 days after clomiphene citrate stimulation (clomiphene citrate: 50 mg. 1x1) and later at the 12th day of siclus MR did not indicate any findings about non ascended. Although it was reported that MR is more efficient than laparoscopy following over stimulation with clomiphene citrate or the detection of ectopic ovary for it is cheaper, non-invasive and more sensitive,[7] for our case both ovaries and the uterus were not seen in MR imaging. For this reason, we believe that examination of more cases and more research would be beneficial for the development of an efficient diagnosis method. According to the second mechanism, it is argued that this is caused by a localized defect at the caudal section of the Mullerian canal and at the genital ridge area or because Mullerian and mesonephric canals are not developed totally at one side.[8] This mechanism can be valid for our case too. According to the final mechanism, intrauterine can be caused by asymptomatic adnexal torsion which happens during childhood or adulthood.[9] Considering that our case is 17 years old, the adnexal torsion might only have happened during intauterine period.

In conclusion, although their causes are controversial due to their rarity, they can be caused by a localized defect at the caudal section of the Mullerian canal and genital ridge area or an as a result of an adnexal torsion during intrauterine period.

Primer Amenoreli Bir Olguda Uterus Unikornus ve Unilateral Ovarian ve Renal Agenezis

Nadir görülmesi nedeniyle primer amenoresi ve uterus unikornus ile unilateral ovarian ve renal agenezisi olan bir olguyu literatür desteği altında yayınlamak.

17 yaşında, bekar olarak ve primer amenoresi nedeniyle başvurdu. Anamnezinde 3 ay boyunca oral kontraseptif (Gynera tb, 1x1) kullandığı ancak menses olmadığı saptandı. Büyümesinin normal olduğu ve meme ve pubik kıllanma gelişiminin Tanner evre 4 olarak değerlendirildi. Yapılan kromozomal analiz sonucu 46, XX olarak rapor edildi. Laboratuar değerlerinden FSH, LH ve E2 normal olarak saptandı. Hastada müllerian agenezis ön tanısıyla yapılan transabdominal ultrasonografisinde ve pelvik magnetik rezonanas görüntülemede her iki over ve uterus izlenmedi şeklinde rapor edildi. Laparoskopi sırasında uterus orta hatta görülmekle birlikte unikornus yapıda idi. Rudimenter horn saptanmadı. Sol round ligament, tuba ve overler normal olarak izlendi. Sağ round ligament, tuba ve over izlenmedi. İntraabdominal endometriozise ait herhangi bir implantasyon odağı saptanmadı. Batın içinde mensese ait eski kanamaya ait bulgu izlenmedi. Postoperatif intra venöz pyelografi çekildi ve sağ böbrek izlenmedi, sol böbrek normalden büyük olarak rapor edildi. Hastaya oral kontraseptif başlanarak sonrasında histeroskopi altında vajinal ve/veya servikal agenezis açısından değerlendirilmesi planlanarak taburcu edildi.

Nadir görülmeleri nedeniyle sebepleri tartışmalı da olsa müllerian kanalın kaudal kısmında ve genital kabartı bölgesinde lokalize defekt veya intrauterin dönemde adneksal torsiyon sonucunda oluşabilir.

Anahtar Kelimeler: Primer amenore, Uterus unikornis, Yumurtalık ve böbrek agenezisi

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