THE TREATMENT OF CYCLIC PELVIC PAIN IN 35 YEARS OLD PATIENT WITH MAYER-ROKITANSKY-KUSTNER-HAUSER SYNDROME

Alpaslan KABAN[1], Ugur ATES[2], Isik KABAN[3], Alim OZCAN[1]

[1] Department of Obstetrics and Gynecology, Arnavutkoy State Hospital, Istanbul, Turkey
[2] Department of Obstetrics and Gynecology, Haseki Education and Research Hospital, Istanbul, Turkey
[3] Department of Obstetrics and Gynecology, Fatih Sultan Mehmet Education and Research Hospital, Istanbul, Turkey

SUMMARY

Mayer-Rokitansky-Kustner-Hauser (MRKH) syndrome comprises the absence or hypoplasia of the uterus and the upper two third of the vagina. Diagnosis of these patients may be delayed until the period of adolescence or even sexualy active period because the external appearance of patients are the same with completely normal female and often do not have any complaints. Renal, cardiac, skeletal abnormalities, pulmonary and hearing defects may be associated with MRKH syndrome. Accuracy of diagnosis of MRKH syndrome is very important for providing psychological support, acquisition of normal sexual function, treatment and to give advice on assisted reproductive techniques. In these cases, a detailed systemic evaluation for the presence of other accompanying system anomalies should not be accuracy of diagnosis. Our presentation is about a patient with 36 years old with MRKH syndrome diagnosed at the age of 16 and had undergone vaginoplasty operation there after she could be sexually active.

Key words: *cyclic pelvic pain, Mayer-Rokitansky-Küstner-Hauser syndrome, uterovaginal agenesia*

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MAYER-ROKİTANSKY-KÜSTNER-HAUSER SENDROMLU 35 YAŞINDA HASTANIN SİKLİK PELVİK AĞRI TEDAVİSİ

ÖZET

Vajenin üst 2/3 kısmının ve uterusun olmayışı yada gelişmemiş olması Mayer-Rokitansky-Küstner-Hauser sendromu (MRKH) olarak bilinir. Bu hastaların tanısı genellikle ergenlik dönemlerine kadar hatta cinsel aktif döneme gelinceye kadar gecikir. Çünkü hastaların dış görünüşü tamamen normal dişi görünümündedir ve çoğunlukla herhangi bir şikayetleri olmaz. Beraberinde renal, kardiyak, iskelet sistemi anomalileri, pulmoner ve işitsel kusurlar bulunabilir. MRKH sendromunda tanının doğruluğu, psikolojik destek sağlanması, normal seksüel fonksiyonların kazanılması, tedavi ve yardımla üreme tekniklerine yönelik danışmanlık verilmesi açısından oldukça önemlidir. Bu olgularda, eşlik eden diğer sistem anomalilerinin varlığına yönelik ayrıntılı bir sistemik değerlendirme ihmal edilmemelidir. Burada 16 yaşındayken tanısı konmuş ve vajinoplasti operasyonu geçirmiş sonrasında cinsel yönden aktif beraberlik yaşayabilen 36 yaşındaki hastamızı sunacağız.

Anahtar kelimeler: *Mayer-Rokitansky-Küstner-Hauser sendromu, siklik pelvik ağrı, uterovajinal agenezi*

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Address for Correspondence: Alpaslan Kaban. Arnavutköy Devlet Hastanesi, Arnavutköy, İstanbul, Turkey
Phone: + 90 (212) 441 69 52
e-mail: dralpaslan2009@gmail.com
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INTRADUCTION

Mayer-Rokitansky- Küstner-Hauser syndrome-(MRKH) was defined by Mayer in 1829[1].
Mayer-Rokitansky-Küstner-Hauser (MRKH)syndrome, occurs as a result of interruptions during the embryonic development of the mullerian ducts and it is the congenital aplasia of uterus and the upper 2 / 3 vagina.In the MRKH cases external genitalia is normal and at adolescence, secondary sex characters show a normal development. Genetic sex with 46, XX , in the female phenotype and their ovarian functions are normal[2].
Uterus, fallopian tubes &upper 2/3 of vagina developed from the fusion of two Müllerian ducts. Lower third of vagina developed from the ascending Sino vaginal bulb. Nearly the most common presentations of mullerian agenesis are amenorrhea, impossible sexual intercourse at more advenced age or infertility.The rate is 1/4000-5000 of live female births [1]. Usually with congenital absence of the uterus and vagina there are rudimental uterine bulbs and tubes. Mullerian agenesis may be isolated,or may be along with a number of additional anomalies. skeletal abnormalities involving the spinal cord and urinary tract anomalies especially in renal or may accompany congenital heart anomalies [3].
Previously the syndrome can be seen as a sporadic but recently, an increasing number of familial cases supports the idea of a genetic predisposition.
The cases are mostly diagnosed during the adolescant period while researching the primary amonorhea.
In this article a 36 years old sexually active type I MRKH sendrom with a analgesic resistant cyclic pelvic pain passed 3 years is presented and discussed with the current literature. The case was diagnosed and has been opereted with the technique Mc Indo vaginoplasty at the age of 16.

CASE

36 year old patient, was referred to our clinic because of the cyclic lower abdominal pain unresponsive to analgesics.In the gynecological examination, the perineum and vulva have the anatomically normal view,and the development of breast was Tanner Stage 5. The patient has a regular sexual life and the depth of her vagina was approximately 8 cm .The vaginal mucosa of the case compared to normal mucosa and tissue of the vagina was more rigid. In the transvaginal ultrasonography uterine dimensions are 2.5 x 2 x 1.5 cm, right ovary had a normal appearance and size,left ovary had a follicular cyst. Patients' urinary system X-rays were normal. Hormonal examinations of FSH: 7.7 Estradiol: 95 CA-125: 6.0 detected.
The patient was prepared for operation. due to the first operation vaginal manipulation was difficult so laparotomy was preferred instead of laparoscopy laparotomy was made with Pfannenstial incision and two rudimentary uterine horns, the form of normal ovaries were appeared (Figure 1). Rudimentary uterine horns and atrophic corpus were excised (Figure 2). The patient who remained stable in the postoperative period and discharged had no complaint during the first four months.

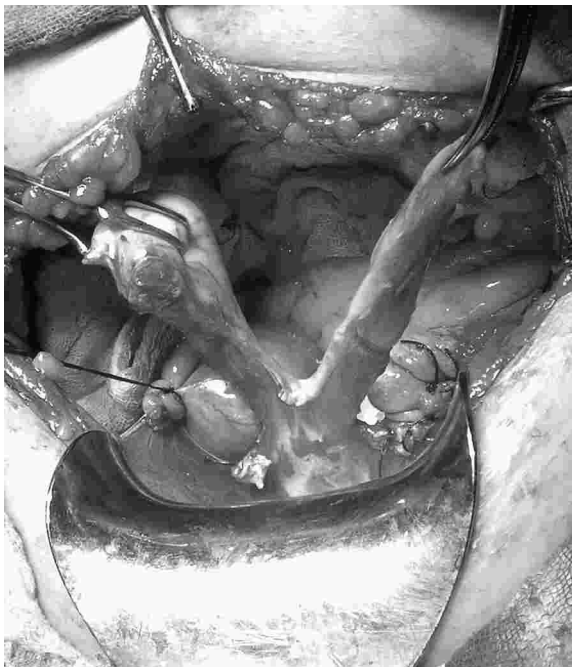

Figure 1:

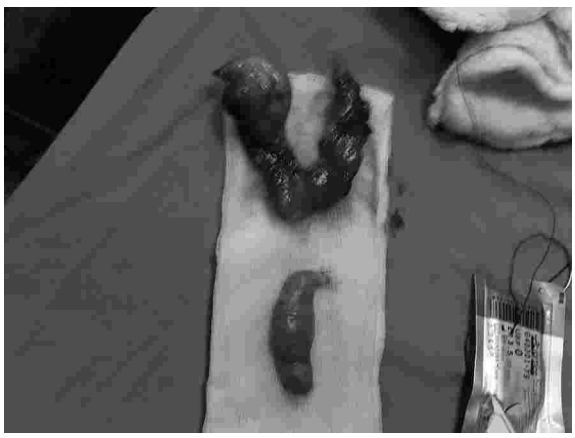

Figure 2:

DISCUSSION

MRKH syndrome is a sydrome thought to be due to degradation of Mullerian duct developmet and differentiation between the 28 to 45 days of embryogenesis[4]. Some factors are thought that may cause this syndrome have been reported which were not certain. Maternal gestational diabetes, teratogens such as thalidomide, increased galactose levels, decreased galactose-1-phosphate üridiltransferaz (GALT) activity, activation of Mullerian inhibiting Substance (MIS), MIS receptor defects, recessive inheritance and aetiological causes of spontaneous mutation but no evidence has been proposed to be enough[5,6].

Whether there is a familial predisposition to fully is unknown. However, an increasing number of cases of familial notification of the transition in question may be a genetic suggest.In recent studies Chromosomal analysis of patients with MRKH syndrome were made, and in some patients with chromosomal microdeletions determining some of the chromosomal structure was normal[7-10].

MRKH Syndrome is the secondary most common cause of amonerhea after gonadal disgenesis. It is the %15-20 of all primary amenorrhea causes. MRKH syndrome may be isolated (type I MRKH) or may be with renal, cardiac, skeletal system abnormalities, and visual defects (Type II MRKH)[11,12].

In isolated MRKH type I syndrome, bilateral ovaries, the development of fallopian tube, and the renal system were normal. Uterin aplasia and there are peritoneum associated two rudimentary horns. Vagina is atretic or terminates in the form of a blind pocket.

Oppelt et al. their 53 cases of MRKH a study including 25 (47%) cases of type I MRKH as shown. Type II MRKH syndrome, includes accompanied other system malformations. Mullerian, renal and cervical spine (MURCS) syndrome and GREASE (Genital Renal Ear Syndrome), are evaluated in this group. In Type II MRKH syndrome, asymmetry of uterin horns and hypoplasia of the fallopian tube can be seen[13,14].

Raudrant et al used the method of, laparoscopic Excision of the rudimentary uterine horn and uterovajinal anastomosis in the treatment of cyclic pelvic pain for MRKH Syndrome patients. They reported that this may be an alternative method of laparotomy and classical radical excision procedure of horns[15].

Lamarca and colleagues, reported a case that a MRKH Syndrome patient developed fibroids on rudimentary uterine horns[16].

There are some reviews in literature about endometriotic masses and foci despite the absence of uterus. This hypothesis about the formation of endometriosis supports the idea of cholemic metaplasia[17,18].

Cyclic pelvic pain in patients with MRKH syndrome laparotomy or laparoscopic treatment of uterine Horn removal has been the classic approach[19,20].

Today, patients with MRKH, have no chance except for the surrogate mother to have children. IVF programs applied in patients with MRKH ovarian responses to gonadotrophins are normal and carrier rate of congenital anomalies in infants born to surrogate mothers does not increase[21,22].

REFERANCES

1. Kapoor R, Sharma DK, Singh KJ, Suri A, Sıngh P, Chaudhary H, Dubey D, Mandhani A. Sigmoid vaginoplasty: Long-Term results. Urology 2006; 67: 1212- 5.
2. Folch M, Pigem I, Konje JC. Müllerian agenesis: etiology, diagnosis, and management. Obstet Gynecol Surv 2000; 55: 644- 9.
3. Şatıroğlu H, Berker B. Jinekolojik ve Obstetrikal Cerrahi. Güner H, Editör: Müllerien Anomaliler ve Cerrahi Tedavileri. 1. Baskı. Ankara: Güneş Kitabevi; 2005, p: 701-6.
4. Folch M, Pigem I, Konje JC. Müllerian agenesis: etiology, diagnosis, and management. Obstet Gynecol Surv 2000; 55: 644- 9.
5. Edmonds DK. Congenital malformations of the genital tract and their management. Best Pract Res Clin Obstet Gynaecol 2003; 17: 19- 40.
6. Zenteno JC, Carranza-Lira S, Kofman-Alfaro S. Molecular analysis of the anti-Müllerian hormone, the anti-Müllerian hormone receptor, and galactose-1-phosphate uridyl transferase genes in patients with the Mayer- Rokitansky-Küstner-Hauser syndrome. Arch Gynecol Obstet 2004; 269: 270- 3.
7. Guerrier D, Mouchel Th, Pasquier L, Pellerin I: The Mayer-Rokitansky- Küstner-Hauser syndrome (congenital absence of uterus and vagina)- phenotypic manifestations and genetic approaches. J Negat Results Biomed 2006, 5: 1.
8. Moerman P, Fryns JP, Sastrowijoto SH, Vandenberghe K, Lauweryns JM: Hereditary renal adysplasia: new observations and hypothesis. Pediatr Pathol 1994; 14: 405- 10.
9. Cheroki C, Krepischi-Santos ACV, Szuhai K, Brenner V, Kim

CA, Otto PA, Rosemberg C: Genomic imbalances associated with müllerian aplasia. J Med Genet 2008; 45: 228- 32.

10. Bernardini L, Gimelli S, Gervasini C, Carella M, Baban A, Frontino G, Barbano G, Divizia MT, Fedele L, Novelli A, Bèna F, Lalatta F, Miozzo M, Dallapiccola B: Recurrent microdeletion at 17q12 as a cause of Mayer- Rokitansky-Kuster-Hauser (MRKH) syndrome: two case reports. Orphanet J Rare Dis 2009; 4(1): 25. 11- 2. Morcel K, Camborieux L; Programme de Recherches sur les Aplasies Müllériennes, Guerrier D. Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome. Orphanet J Rare Dis 2007; 2: 13.
12. Duncan PA, Shapiro LR, Stangel JJ, Klein RM, Addonizio JC. The MURCS association: Müllerian duct aplasia, renal aplasia, and cervicothoracic somite dysplasia. J Pediatr 1979; 95: 399- 402.
13. 7. Oppelt P, Renner SP, Kellermann A, Brucker S, Hauser GA, Ludwig KS. et al. Clinical aspects of Mayer-Rokitansky-Küstner-Hauser syndrome: recommendations for clinical diagnosis and staging. Hum Reprod 2006; 21: 792- 7.
14. Strübbe EH, Willemsen WN, Lemmens JA, Thijn CJ, Rolland R. Mayer-Rokitansky-Küstner-Hauser syndrome: distinction between two forms based on excretory urographic, sonographic, and laparoscopic findings. AJR Am J Roentgenol 1993; 160: 331- 4.
15. Raudrant D, Chalouhi G, Dubuisson J, Golfier F, Laparoscopic uterovaginal anastomosis in Mayer-Rokitansky-Küstner-Hauser syndrome with functioning horn. Fertil Steril. 2008 Dec; 90(6): 2416- 8.
16. Lamarca M, Navarro R, Ballesteros ME, García-Aguirre S, Conte MP, Duque JA Leiomyomas in both uterine remnants in a woman with the Mayer-Rokitansky-Küstner-Hauser syndrome. Fertil Steril. 2009 Mar; 91(3): 931. e13- 5.
17. Cho MK, Kim CH, Endometriosis in a patient with Rokitansky-Küstner-Hauser syndrome Oh ST J Obstet Gynaecol Res. 2009 Oct; 35(5): 994- 6.
18. Mok-Lin EY, Wolfberg A, Hollinquist H, Laufer MR. Endometriosis in a patient with Mayer-Rokitansky-Küstner-Hauser syndrome and complete uterine agenesis: evidence to support the theory of coelomic metaplasia. J Pediatr Adolesc Gynecol. 2010 Feb; 23(1): e35- 7.
19. Giatras K, Licciardi F, Grifo JA, Laparoscopy for pelvic pain in the Mayer-Rokitansky-Kuster-Hauser syndrome. A case report. J Reprod Med. 1998 Mar; 43(3): 203- 5.
20. Chapron C, Morice P, La Tour MD, Chavet X, Dubuisson JB, Laparoscopic management of asymmetric Mayer-Rokitansky-Küstner-Hauser syndrome Hum Reprod. 1995 Feb; 10(2): 369- 71.
21. Petrozza JC, Gray MR, Davis AJ, Reindollar RH. Congenital absence of the uterus and vagina is not commonly transmitted as a dominant genetic trait: outcomes of surrogate pregnancies. Fertil Steril 1997; 67: 387- 9.
22. Beski S, Gorgy A, Venkat G, Craft IL, Edmonds K. Gestational surrogacy: a feasible option for patients with Rokitansky syndrome. Hum Reprod 2000; 15: 2326- 8.