REVIEW

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Laparoscopic Myomectomy in a Patient with a Laparoscopic Vaginoplasty (Modified Davydov) and Review of the Literature

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ABSTRACT

Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome is characterized with the congenital absence of uterus and vagina. Uterus may be rudimentary. Coexistence of a myomauteri with MRKH syndrome is possible. The case presented in this study is the eighteenth report in the literature. Moreover, it is the first one with coexistent skeletal system anomalies, left pelvic renal ectopia, and leiomyoma of the rudimentary uterus. Coexistence of a uterine leiomyoma and MRKH syndrome is very rare. However, if a patient with MRKH syndrome presents with a pelvic mass, the possibility of a leiomyoma should be considered. The review of the english written medical literature revealed that the coexistence of a uterine leiomyoma and MRKH syndrome is very rare. However, if a patient with MRKH syndrome present with a pelvic mass, the possibility of a leiomyoma should be considered.

Key Words: *Mayer-Rokitansky-Kuster-Hauser syndrome, davydov operation, myoma uteri*

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INTRODUCTION

Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome is characterized with the congenital absence of uterus and vagina. While uterus may be rudimentary, fallopian tubes and ovaries are present. Secondary sexual characteristics are normally developed. Anomalies of the urinary tract and skeletal system may accompany this syndrome.

Coexistence of uterine myoma and Rokitansky syndrome was first reported in 1977[(1)]. The case presented in this study is the eighteenth report in the literature. Moreover, it is the first one with coexistent skeletal system anomalies, left pelvic renal ectopia, and leiomyoma of the rudimentary uterus.

CASE REPORT

The 24-year-old, married patient was diagnosed with MRKH syndrome at the age of 18 when she first applied to the hospital with the complaint of never having had menstruation. During her diagnostic laparoscopy, she was found to have a rudimentary uterus, bilateral and normal appearing ovaries and fallopian tubes, and left renal pelvic ectopia. Her gynecologic examination revealed a blind vaginal pouche of 1 cm in length and secondary sexual characteristics were fully developed. The karyotype was 46, XX.

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Her presenting complaint when she applied to our clinic in 2009 was not being able to have sexual intercourse despite two years of marriage. Transabdominal ultrasonography revealed a rudimentary uterus and bilateral normal ovaries. Left renal pelvic ectopia was diagnosed during intravenous pyelography.Posterior arcus fusion defect in sacral vertebras were identified. Davydov operation was planned for the treatment of vaginal agenesis.

During the operation, an eight cm sized mass originating from the rudimentary horn of the uterus was seen. Laparoscopic excision of the mass was carried out. Bilateral ovaries were normal in appearance. Vesicorectal space was dissected untilperitoneum was visualized. Peritoneal edges and perineal epithelium were approximated with sutures. Afterwards, peritoneum, which would be the vaginal cuff,was saturated laparoscopically. At the end of the operation, a rigid stent soaked with vaseline was put into the newly created vagina. The pathological report of the excised mass was in accordance with uterine leiomyoma. Vaginal dilator was taken out on the postoperative fifth day and the patient was discharged on the postoperative sixth day.

DISCUSSION

Diagnosis of MRKH syndrome is generally made in adolescence during the work up of primary amenorrhea. Vaginal reconstructive operations are carried out in patients with this diagnosis to improve penetration during sexual intercourse and help these patients with their psychological conditions. Since bilateral ovaries, fallopian tubes, and two uterine remnants of various sizes exist in patients with this syndrome, pathologies of these structures are possible. In the literature regarding MRKH, there are case reports of leiomyomas originating from rudimentary horns in patients presenting with pelvic pain, adenomyosis, degenerated leiomyomas, ovarian carcinoma in two patients, ovarian dysgerminoma, immature teratoma of the ovary in a four year old girl and ovarian endodermal sinus tumor[2-7]. Since ovarian steroidogenesis continues in these patients, they should be followed up for genital neoplasms showing estrogenic activity. Complications due to pathologies as the degeneration of a leiomyoma or torsion of an ovarian cyst should be kept in mind.

Table 1 shows the characteristics of MRKH cases with leiomyomas and it can be seen that most cases presented many years after vaginal reconstruction. The leiomyoma in this case was diagnosed and excised during laparoscopic davydov operation. Magnetic resonance imaging is very helpful in identifying ovaries, rudimentary uterus as well as accompanying pelvic and skeletal anomalies in related body regions in these patients. The incidence of upper urinary tract anomalies coexisting MRKH syndrome is 30-40%.The most common accompanying anomalies are renal agenesis and pelvic renal ectopia[8]. This case report is the first in the literature presenting leiomyoma originating from a rudimentary uterine horn with skeletal system defect and renal pelvic ectopia in a patient with MRKH syndrome.

In cases of leiomyoma arising from a rudimentary horn, myomectomy with excision of the horn is indicated[9,10]. However, since the leiomyoma was pedunculated in this case, only myomectomy was performed. Myomectomy can be done with laparotomy or laparoscopy[11-20]. Since laparoscopy had initially been used in this case, myomectomy was also laparoscopically carried out.

Various vaginal reconstruction methods are defined for patients with MRKH syndrome. The most popular of these are the Frank nonsurgical technique, the Williams vaginoplasty and its Creatsas modification, the Vechietti operation, and laparoscopic davydov operation used in this case. On the postoperative sixth week, the vaginal length in this case was 7-8 cm. Two years after the operation, the vaginal length and width was 6 and 2 cm, respectively. The patient reported to be able to have sexual intercourse.

Coexistence of uterine leiomyoma and MRKH syndrome is very rare. Therefore, it may not be possible to identify myoma before the operation. However, if a patient with MRKH syndrome presents with a pelvic mass, the possibility of a leiomyoma should be considered.

Table 1. Characteristics of patients with MRKH syndrome and coexisting leiomyoma

Case number	Detecting age of myoma uteri	Complaint	Type of operative procedure for vaginal agenesis	Dimension of myoma uteri
Case I 1977 (1)	Data not available			
Case II 1978 (10)			McIndoe's procedure	Large leiomyoma
Case III 1988 (15)	32	Lower abdominal pain and bloating	McIndoe's procedure	100 x 8.5 mm
Case IV 1988 (21)			Vaginal dilatation	4 cm
Case V 1999 (16)		Episodic pelvic pain and urinary frequency	No operation	62*62*63 mitotically active myoma
Case VI 2000 (2)	27	Pelvic pain	McIndoe's procedure	49 x 46 x 44 mm
Case VII 2000 (13)	36	Pelvic pain	vaginopoiesis	85 mm
Case VIII 2000 (22)	Data not available			
Case IX 2002 (17)	52	Low abdominal pain	No operation	12 x 12 x 8.5
Case X 2003 (12)	42	Deep dyspareunia	Vaginoplasty using amniotic membranes	100 mm
Case XI 2003 (18)	42	Lower abdominal pain	Creatsas modification of Williams' vaginoplasty	59 x 55 mm
Case XII 2003 (18)	38	A mass in the area of left adnexa	Creatsas modification of Williams' vaginoplasty	48 x 36 mm
Case XIII 2003 (23)	55	The first one torsion of myoma uteri, second one episodic pelvic pain	Deepen of the natural recess between bladder and rectum	The first one 11 cm, the second one 10 x 7.5 cm
Case XV 2006 (24)	41	Cyclic lower abdominal pain	Vaginal reconstructive procedure	56 x 40 mm
Case XVI 2008 (25)	47	Lower abdominal pain, pelvic tumescence		50 mm
Case XVII 2009 (26)	39	Asymptomatic	No vaginal reconstruction	90 mm
Case XIX 2009 -our case	24	Failure of sexual relationship	Laparoscopic approach of davydov	80 x 70 mm
Case XX 2012 (20)	28	Mass and acute abdominal pain	Unknown	10 x 15 cm
Case XXI 2012 (20)	34	Primary amenorrhea, infertility	Unknown	6 cm
Case XXII 2013 (19)	35	Mass and pain, primary amenorrhea		25 x 18 x 12 cm

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DERLEME-ÖZET

Laparoskopik Yaklaşımlı Vajinoplasti Yapılan Hastada Laparoskopik Miyomektomi ve Literatürün Taranması

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Mayer-Rokitansky-Kuster-Hauser (MRKH) sendromu uterus ve vajinanın konjenital yokluğuyla karakterizedir. Uterus rudimenter olabilir. MRKH sendromuyla miyoma uteri birlikteliği olabilir. Bu olgu literatürde bildirilen 18. olgudur. Dahası olguya iskelet sistemi anomalisi, sol pelvik renal ektopi ve miyoma uteri eşlik ettiği için literatürde ilktir. Uterin leiomiyoma ve MRKH sendromu birlikteliği çok nadirdir. Bununla birlikte MRKH sendromu olan bir hastada pelvik kitle saptanırsa leiomiyoma olasılığı düşünülmelidir. İngilizce literatürün taranması uterin leiomiyoma ve MRKH sendromu birlikteliğinin çok nadir olduğunu gösterdi.

Anahtar Kelimeler: *Mayer-Rokitansky-Kuster-Hauser sendromu, davidov operasyonu, miyoma uteri*

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