

## 자궁내막증이 동반된 Mayer-Rokitansky-Küster-Hauser 증후군의 수술적 치료경험 1례

울산대학교 의과대학 서울아산병원 산부인과학교실

### A Case of Mayer-Rokitansky-Küster-Hauser Syndrome Accompanying Endometriosis

Jeong Yeol Park, Chung Hoon Kim, Cheon Sik Kang, Ji Young Lee, So Youn Park,  
Sung Hoon Kim, Hee Dong Chae, Byung-Moon Kang

Department of Obstetrics and Gynecology, College of Medicine,  
University of Ulsan, Asan Medical Center, Seoul, Korea

The Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome refers to a condition which presents as normal female secondary sex characteristics, normal external genitalia, congenital absence of the internal vagina, usually a rudimentary uterus in the form of bilateral noncanalculated muscular buds, and normal tubes and ovaries with normal cytogenetic and endocrine evaluation, frequent association of renal, skeletal and other congenital anomalies. However, rarely, whole uterus or a segment of uterus may be present, but lacking a conduit to the introitus. If a partial endometrial cavity is present in a segment of uterus, cyclic abdominal pain may be a complaint and furthermore endometriosis can be developed. Recently, we experienced a case of MRKH syndrome with the segments of uterus accompanying endometriosis in young woman. We present this case with a brief review of literatures.

**Key Words:** MRKH Syndrome, Endometriosis

Mayer-Rokitansky-Küster-Hauser (MRKH) . MRKH 가  
4,000~5,000 1  
(Müller) ,  
1927 Sampson  
46,XX , (implantation theory)<sup>1</sup>  
MRKH 1593 Realdus Columbus가  
,<sup>2</sup> 1948 500

가 , 1979 , 가  
 , 30 가  
 . 가 MRKH  
 가 ,

MRKH

1

: O , 17 ,  
 : 0-0-0-0

가 : 1 2

: 1986

(Duhamel's operation)

: 2001 12 28

2002 1 3

: 가 ,  
 . 167 cm,

43 kg ,

:

:

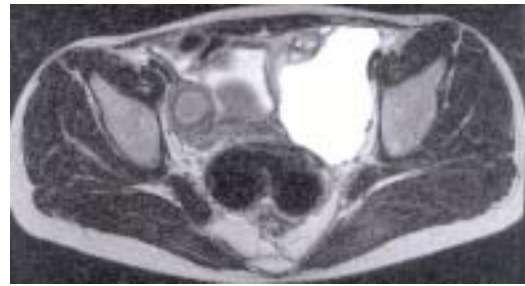
:

7~8 cm

6 cm

: Speroff (1994)

PRL 5.4 ng/ml, TSH 5.6 µIU/ml, LH 5.0 mIU/ml,



**A**



**B**



**C**

**Figure 1a.** A segment of right uterine body with functioning endometrium and hematometra. Normal left ovary and cystic mass are seen.

**1b.** Right ovarian cyst.

**1c.** A segment of left uterine body with functioning endometrium.

FSH 5.04 mIU/ml, E2 64 pg/ml, T <0.1 ng/ml

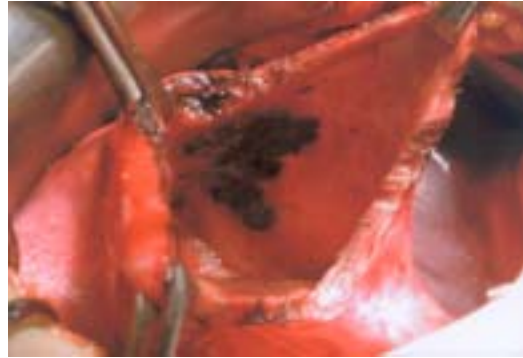
46,XX

(Figure 1a).

(multilo-



**Figure 2.** A segment of right uterine body forming hematometra.



**Figure 3.** Right ovarian endometrial cyst containing chocolate-colored fluid.

culated tubular structure) , (Figure 1b).

(Figure 1c) 가 (Figure 1a). 12 cm (Figure 1a, 1c).

cm , X

: (segmental agenesis of Mullerian duct)

(paraovarian cyst)

, 2003 1 21

: 2003 1 23

7.6×7×6 cm

(Figure 2).

가 , 6×6×5 cm

가

(Figure 3).

1 cm

가 (Blue-black spot)

, 12 cm

paraovarian cyst

:

,

,

:

,

,

,

(hemosiderin-laden

macrophage)가

:

, GnRH agonist

MRKH	1593	Realdus Columbus <sup>1</sup> 가	(vaginal vestibule)	1~2 cm
		Mayer <sup>3</sup> 가	가	
		Rokitansky <sup>4</sup> 가	1/3	
Küster <sup>5</sup>				12%
Hauser <sup>6</sup>		MRKH	4,5	
	1948	500	6	가
		가 1979	MRKH	가
가	80	(1983),	MRKH	<sup>11</sup> Kar-
(1984),	(1994)		yotype	46,XX Barr body
30	가			XX/XO
		4,000~5,000	mosaicism, 47,XXX/46,XX mosaicism	가
1 <sup>7,8</sup>		2,000	<sup>12</sup>	Bryan <sup>8</sup> 1:100, Turunen
			<sup>13</sup> 200	1
				<sup>10</sup>
		(Mullerian duct)		
(urogenital sinus)			가	
(Wolffian duct)				
가				

**Table 1.** Principal clinical features of the Mayer-Rokitansky-Küster-Hauser syndrome

1. Primary amenorrhea associated with congenital absence of the vagina.
2. 46,XX karyotype.
3. Uterus that varies from anatomically complete to rudimentary bicornuate cords to complete absence.
4. Normal ovarian function and normal ovulation.
5. Normal female breast development, body proportions, and body hair.
6. Frequent association of renal, skeletal, and other congenital anomalies.

				가 3~4 cm
				가 3 cm
	Table 1			, Frank, <sup>17</sup>
			Warton, McIndoe, Williams	가
			Deen, <sup>18</sup> Golditch <sup>19</sup>	가
	X-			가 MRKH
dysgenesis), (testicular feminiza- tion)		(gonadal (testicular feminiza- tion)		1
가				
MRKH				
				1. Sampson JA. Peritoneal endometriosis due to the menstrual dissemination of endometrial tissue into the peritoneal cavity. Am J Obstet Gynecol 1927; 14: 422-69.
				2. Shute WB. Congenital absence of the vagina. Obstet Gynecol 1958; 12: 310.
가				3. Mayer. Über verdoppleungen des uterus und ihre arten nebst bomerkungen uber hausenscharte und wolfsrachen. J Chir Auger 1829; 13: 525-64.
. Farber	Marchant <sup>14</sup>		가	4. Rokitansky. Über die sogenannten verdoppleungen des uterus, Med Jb Ost Staat 1838; 26: 39-77.
가	Feary Weed <sup>15</sup>		가	5. Küster H. Uterus bipartitus solidus rudimentarius cum vagina dolida. Z Gob Gyn 1910; 67: 692-728.
4				6. Hauser GA, Schreiner WE. Das Mayer-Rokitansky-Küster syndrome. Schweiz Med Wochenschr 1961; 91: 381-4.
	, Niver <sup>16</sup>		가	7. Engstad JE. Artificial vagina. J. Lancet 1917; 37: 329-31.
3				8. Bryan AL, Nigro JA, Counseller VS. One hundred cases of congenital absence of the vagina. Surg Gynecol Obstet 1949; 88: 79-86.
	가			9. Owens N. Simplified method for formation of an artificial vagina by split skin graft. Report of a case. Surgery 1942; 12: 139-50.
가				10. Grover S, Solanki BR, Banerjee M. A clinicopathologic study on Mullerian duct aplasia specific reference to cytogenetic studies. Am J Obstet Gynecol
			, Bryan <sup>8</sup>	
가				

- 1970; 107: 133-8.
11. Schmid-Tannwald I, Hauser GA. Deutung der, atypischen, formen des Mayer-Rokitansky-Küster syndroms. *Geburtsh u Frauenheilk* 1977; 37: 386-92.
  12. Azoury RS, Jones HW JR. Cytogenetic findings in patients with congenital absence of the vagina. *Am J Obstet Gynecol* 1966; 94: 178-80.
  13. Turnuen A, Unnerus CE. Spinal changes in patients with congenital aplasia of the vagina. *Acta Obstet Gynecol Scand* 1967; 46(1): 99-106.
  14. Shokeir MH. Aplasia of the mullerian system. *Birth Defect Orig Artic Ser* 1978; 14 (6C): 147-65.
  15. Geary WL, Weed JC. Congenital atresia of the uterine cervix. *Obstet Gynecol* 1973 Aug; 42(2): 213-7.
  16. Niver DH, Barrette G, Jewelewicz R. Congenital atresia of the uterine cervix and vagina: Three cases. *Fertil Steril* 1980 Jan; 33(1): 25-9.
  17. Williams EA. Congenital absence of vagina. *J Obstet Gynaecol Br Commonw* 1972 Dec; 79(12): 1147-8.
  18. Deen DC. Vaginal reconstruction with silicone foam molds. *Am J Obstet Gynecol* 1969 Apr 1; 103(7): 930-4.
  19. Golditch IM. A modified mold in vaginal aplasia. *Am J Obstet Gynecol* 1968 Aug 15; 101(8): 1135-6.
-