Thai Journal of Obstetrics and Gynaecology
September 1998, Vol. 10, pp. 179-181

GYNAECOLOGY

Laparoscopy for monthly pelvic pain in the Mayer-Rokitansky-Kuster-Hauser syndrome. A case report.

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ABSTRACT

A case of Mayer-Rokitansky-Kuster-Hauser syndrome in which the patient developed monthly pelvic pain was detected. Removal of the remnant of Mullerian duct was performed via laparoscopy. The patient had complete resolution of her pain.

Key word : MRKH syndrome , mullerian duct , laparoscopic , amenorrhea

Primary amenorrhea is a common reason for medical consultation , defined as the absence of menstruation by 16 years of age in the presence of normal secondary sexual characteristics or by 14 years of age when there is no visible secondary sexual characteristic development.[1,2] In order to detect the cause of amenorrhea , it is useful to determine whether secondary sexual characteristics are present. The presence of secondary sexual characteristics indicate that a women has been exposed to estrogen stimulation.

Mullerian agenesis (Mayer-Rokitansky-Kuster-Hauser syndrome) is the second most common cause of primary amenorrhea. (Gonadal dysgenesis is the most common cause)[2,3,4] MRKH syndrome is frequently associated with renal and skeletal anomalies , then chromosomal analysis , intravenous pyelogram and adequate survey for anomalies of the spine should be done.[1,2,4,5] The aim of treatment is psychological support and r eassures the patient about her abnormalities. Operation should be done in the symptomatic patient such as cyclic pelvic pain , pelvic mass. Laparoscopy is not only useful for diagnosis but can also be valuable for any treatment required.[6]

Case report.

A 22 year-old single woman had absence of menstruation , had monthly pelvic pain for 10 years.

Physical examination revealed that she was mild hypersthenic build. Breast and pubic hair were V Tanner staging. Abdomen was flat , soft and no abnormal mass. Extremities and spines were no deformity.

Pelvic examination revealed normal external genitalia with blind pouch vagina (about 2 cm.) , unseen cervix. Rectal examination was revealed firm mass at tip of the finger about 3 x 5 centimeters.

Chromosomal study was 46,XX. Intravenous pyelography and plain spine X-ray were within normal limit. Transabdominal ultrasound revealed a

solid mild hyperechoic mass about 3 x 5 centimeters at left adnexa , unseen uterus and no ectopic kidney.

Treatment.

After complete history taking , physical examination and investigation , counselling the patient about her abnormalities and planning with her about the operation.

Diagnostic laparoscopy was done and revealed abnormal Mullerian duct (as figuer 1,2) with normal both uterine tubes , and ovaries. No ectopic kidney was seen. Resection of remnant of Mullerian duct was done by laparoscopic surgery. The patient was good recovery and discharged from the hospital on the 4th day after operation.

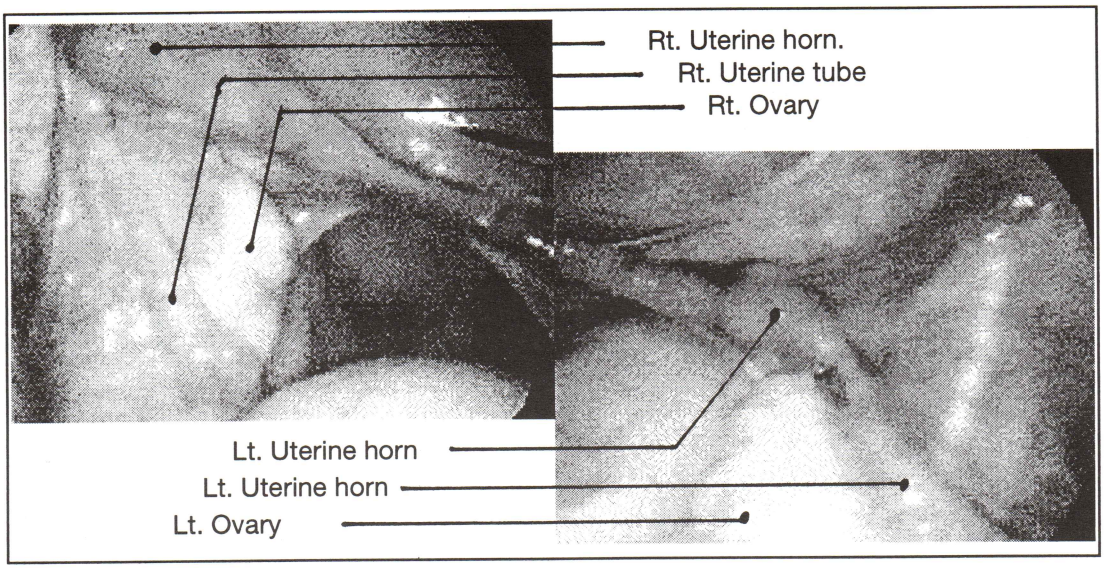

Figure 1. Finding from laparoscopic view.

Pathological finding. (B39-8683-8684) : Tissue from pelvic cavity consistent with Mullerian duct remnant. (The tissue is lined with tubal like epithelium.)

Discussion.

The 22 year-old woman came to the hospital with primary amenorrhea , she had fully development of secondary sexual characteristic and chromosomal study was 46,XX. The differential diagnosis is outflow track obstruction including imperforate hymen , transverse vaginal septum and hypoplasia or absence of the uterus , cervix , and/ or vagina. (Mayer-Rokitansky-Kuster-Hauser syndrome)[1-5 ,7]

Pelvic examination , rectal examination and ultrasound finding could not identify the uterus then MRKH syndrome is the most likely cause of amenorrhea in this patient. Nevertheless , diagnostic laparoscope should be considered because of monthly pelvic pain[2,8] , and to identify type of MRKH syndrome.[10]

Many reports[8,10,11] have to discriminate typical (type A) from atypical (type B) MRKH syndrome and determine their association with renal and spinal anomalies. The typical form is characterized by symmetrical nonfunctioning muscular bud (the Mullerian duct remnants) and normal fallopian tubes, and the atypical form by aplasia of one or both bud, one bud smaller than the contralateral one, with or without dysplasia of one or both fallopian tubes. Associated anomalies are most common in the group with the atypical form of the MRKH syndrome , renal anomalies are found 37-47

percents [5,10] and spinal abnormalities are found 14/52 patients.[9] Laparoscopy is still needed to discriminate between these two form.[10,12]

For this patient laparoscopic finding was compatible with atypical form (type B) MRKH syndrome , sonography and spinal survey were done and associated anomaly was not found.

About monthly pelvic pain, this pain may be ovulatory or possibly a result of dysmenorrhea originate in well-developed rudimentary uterine bulbs. Differentiation between the two by asking the patient to keep a basal body temperature chart and marking the day when pelvic pain is present.[5] For this patient , the pathological finding could not identify endometrium epithelium, then the cyclic pelvic pain may be a result of ovulatory pain.

Psychologic preparation of the patient is the most important problem.[5] The physician should counselling the patient about her abnormalities and plan of management about fertility function and operating to create vagina for coital function. This patient was single and did not plan for marriage in a recent , then creation of an artificial vagina should be performed in a second operation.

Summary

A 22 years old women presented with primary amenorrhea and monthly pelvic pain. Physical examination, sonography, intravenous pyelography, and spinal survey revealed no associated anomaly. The chromosomal study was 46,XX. Laparoscopic finding was compatible with the atypical form of MRKH syndrome. Removal of the structure was performed via laparoscoy. The patient had complete resolution of her cyclic pain

References

1. Scherzer WJ , McClamrock HD. Amenorrhea. In : Berek JS , Adashi EY , Hillard PA , editors. Novak 's gynecology. 12th ed. Baltimore : Williams & Wilkins , 1996 : 809-32.
2. Brumsted JR , Riddick DH. Menstruation and disorders of menstrual function. In : Scott JR , DiSaia PJ , Hammond CB , Spellacy WN , editors. Danforth 's obstetrics and gynecology. 7th ed.Phiadelphia : J.B. Lippincott , 1994 : 665-79.
3. Muto MG , Friedman AJ. The uterine corpus. In : Ryan KJ , Berkowitz RS , Barbieri RL ,editors. Kistner 's gynecology : principles and practice. 6th ed. St. Louis : Mosby – Year Book , 1995 : 137-65.
4. Speroff L , Glass RH , Kase NG. Clinical gynecologic endocrinology and infertility. 5th ed. Baltimore : Williams & Wilkins , 1994 : 401-56.
5. Rock JA. Surgery for anomaries of the Mullerian ducts. In : Thompson JD , Rock JA , editors. Te Linde 's operative gynecology. 7th ed. Philadelphia : J.B. Lippincott , 1992 : 603-46.
6. Chapron C , Morice P , La Tour MD , Charet X , Dubuisson JB. Laparoscopic management of asymmetric Mayer-Rokitansky-Kuster-Hauser syndrome. Hum Reprod 1995 ; 10 : 369-71.
7. Cunningham FG , MacDonald PC , Gant NF , Gilstrap LC III , Hankins GDV. Williams obstetrics. 20th ed. Stamford : Appleton & Lange , 1997 : 635-55.
8. Giatras K , Licciardi F , Grifo JA. Laparoscopy for pelvic pain in the Mayer-Rokitansky-Kuster-Hauser syndrome. A case report. J Reprod Med 1998 ; 43 : 203-5.
9. Strbbe EH , Lemmens JA , Rolland R, Willemsen WN, van Toor BS. Spinal abnormalities and the atypical form of the Mayer-Rokitansky-Kuster-Hauser syndrome. Skeletal Radiol 1992 ; 21 : 459-62.
10. Strbbe EH , Willemsen WN , Lemmens JA , Thiji CJ , Rolland R. Mayer-Rokitansky-Kuster-Hauser syndrome : distinction between two form base on excretory urigraphic , sonographic and laparoscopic findings. Am J Roentgenol 1993 ; 160 : 331-4.
11. Strbbe EH , Cremers CW , Willemsen WN , Rolland R, Thiji CJ. The Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome without and with associated features : two separate entities ? Clin Dysmorphol 1994 ; 3 : 192-9.
12. Malik E , Mangold R , Rossmanith WG. Laparoscopic extirpation of an aplastic ectopic uterus in a patient with Mayer-Rokitansky-Kuster-Hauser syndrome. Hum Reprod 1997 ; 12 : 1298-9.