Ege Tıp Dergisi 41 (4): 255 - 257, 2002

MAYER-ROKITANSKY-KUSTER-HAUSER SYNDROME WITH A BORDERLINE OVARIAN SEROUS CYSTADENOMA: A CASE REPORT

MAYER-ROKİTANSKY-KUSTER-HAUSER SENDROMLU BİR OLGUDA İZLENEN OVERİN BORDERLİNE SERÖZ KİSTADENOMU: OLGU SUNUMU

Murat ULUKUŞ[1] Mustafa ULUKUŞ[1] Ege Nazan TAVMERGEN GÖKER[2] Mustafa Coşan TEREK[1] Osman ZEKİOĞLU[3]

[1]Ege Üniversitesi Tıp Fakültesi Kadın Hastalıkları ve Doğum Anabilim Dalı, Bornova, İzmir
[2]Ege Üniversitesi Rektörlüğü Aile Planlaması Kısırlık Araştırma ve Uygulama Merkezi, Bornova, İzmir
[3]Ege Üniversitesi Tıp Fakültesi Patoloji Anabilim Dalı, Bornova, İzmir

Anahtar Sözcükler: Mayer-Rokitansky-Kuster-Hauser Sendromu, over tümörü.
Key Words: Mayer-Rokitansky-Kuster-Hauser Syndrome, ovarian tumour.

ÖZET

Mayer-Rokitansky-Kuster-Hauser (MRKH) sendromu, vagina ile uterusun beraberce hipoplazisi olarak bilinmektedir ve 4000-5000 dişi doğumda 1 olarak gözlenmektedir. Primer amenore, sterilite ve pelvik ağrı gibi belli başlı jinekolojik problemlere yol açan bu sendromun tanısı, cerrahi eksplorasyonla olduğu kadar, günümüzde gelişen görüntüleme teknikleri ile de mümkün olabilmektedir.

Bu makalede kliniğimize üç gündür süren alt kadran ağrısı şikayeti ile başvuran ve yapılan tetkikler sonucunda MRKH ile birlikte overde borderline seröz kistadenom saptanan 43 yaşında bir olgu sunulmuştur.

SUMMARY

The Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome comprises the combined hypoplasia of the vagina and the uterus and it occurs in 1 of 4000-5000 female births. It may cause many gynecological problems such as primary amenorrhea, sterility, pelvic pain and can be diagnosed by screening or surgical methods.

We report a 43-year-old women with MRKH syndrome and an ovarian serous borderline cystadenoma. She was admitted to our clinic complaining of lower abdominal pain for three days.

INTRODUCTION

The Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome occurs in 1 of every 4000-5000 female births and is characterized by normal external genitalia, an absent vagina, absent or rudimentary uterus and normal fallopian tubes and ovaries (1). The coelomic epithelium which creates the ovaries develops independently of the Mullerian duct which creates the uterus, cervix and 2/3 of the upper vagina. Therefore, individuals with MRKH syndrome have normal ovaries and to our knowledge they are at normal risk of developing ovarian neoplasms.

Yazışma adresi: Murat Ulukuş, Ege Üniversitesi Tıp Fakültesi Kadın Hastalıkları ve Doğum Anabilim Dalı, Bornova, izmir
Makalenin geliş tarihi: 23. 09. 2002 ; Kabul tarihi: 15. 11. 2002

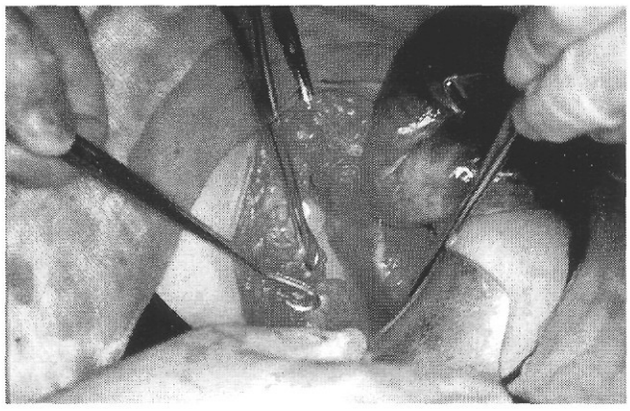
CASE

A 43-year-old widowed woman was admitted to our clinic complaining of lower abdominal pain for three days. She stated that she had neither had a menstrual period nor she had been sexually active. Gynecologic examination

showed a complete vaginal agenesis. Abdominal and rectal examinations revealed a partly cystic, well-shaped mass, extending from above the symphysis to 4 cm below the umbilicus. No uterus or ovaries were palpated by rectal examination. Phenotypic sex was female; breast development, axiller and pubic hair distribution and external genitalia were normal. A hypoplastic uterus was found during the diagnostic laparoscopic examination that was performed 20 years ago.

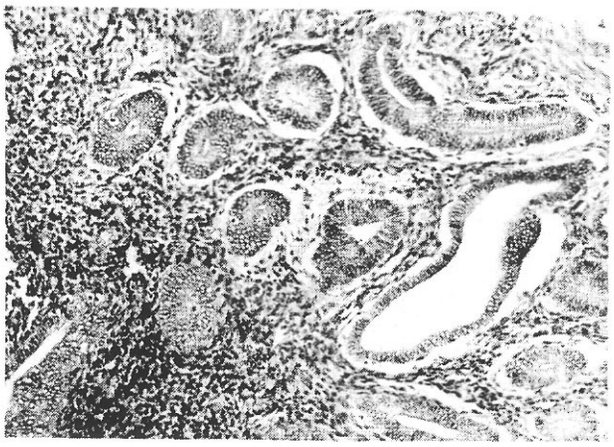
Erytrocyte sedimentation rate (30 mm/h) and serum CA-125 levels (54 U/mL) were slightly increased. The hormone profile showed a postmenopausal status; FSH was increased (54 IU/L), estradiol was decreased (20 pg/mL). Chromosomal analysis of leucocytes revealed a normal karyotype (46,XX).

Abdominopelvic ultrasound showed a 12 x 9 cm cystic mass originating most probably from the left adnexa including heterogeneous and hyperecogeneous solid components. The uterus was hypoplastic and both ovaries were not distinguishable. Computerized tomography (CT) and magnetic resonance imaging (MRI) were performed and were consistent with Mayer-Rokitansky-Kuster-Hauser syndrome and left ovarian mass. Intravenous urographic examination (IVU) showed completely normal urinary tract.

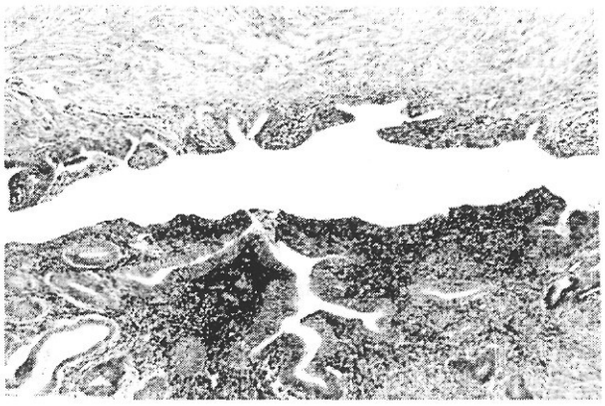


Figüre 1. The view of the pelvis at the time of operation. The lower forceps at the picture (the right side of the patient) holds the round ligament of the hypoplastic uterus. At the upper right side of the picture the left ovarian mass is seen.

At laparatomy, a 10 x 12 x 15 cm, partly cystic, well-shaped mass originating from the left adnexa was found (Figüre 1). The uterus was formed of bilaterally rudimentary uterine bulbs joined by a band behind the bladder. The left uterine bulb was smaller than the other and looked like a fibrous band. While the uterine vessels, round ligament and the fallopian tube could be demonstrated on the right uterus, these structures were missing on the contralateral left uterus. The right ovary was atrophic and there was no sign of ovulation . There was no evidence of metastasis. First the left ovarian mass was extirpated and sent for frozen section pathologic examination. Examination of frozen section revealed a borderline ovarian tumor. Then, both rudimentary uterus and right adnexa were extirpated, partial omentectomy and appendectomy was also performed. Multiple peritoneal biopsies were also obtained. There were no detectable lymphadenopathies in the abdomen or in the pelvis.



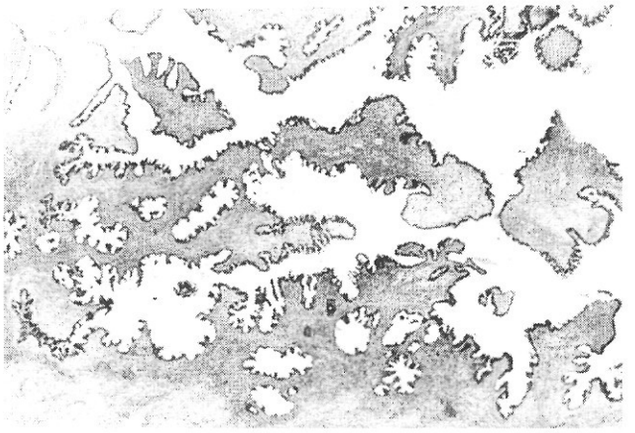
Figüre 2. Proliferative appearance of the endometrium in hypoplastic uterus (H&E, x 100)



Figüre 3. Low power view of the borderiine serous cystadenoma with microinvasion (H&E, x 100)

The histopathological evaluation of the right uterine bulb revealed a rudimentary uterus with proliferative endometrium (Figüre 2). Microscopic evaluation of the tumor revealed a borderline serous cystadenoma of the left ovary (Figüre 3). There was moderate to marked epithelial proliferation. The stratification of the epithelial lining of the papillae, epithelial budding and tufting were prominent. The single enlarged cells with abundant eosinophylic cytoplasm were detected within stromal desmoplasia in only one focus (Figüre 4). The left uterine

bulb was formed of disorganized smooth muscles with a few endometrial glands. The right ovary and appendix showed no pathological findings. Cytologic examination of the peritoneal washing was negative for malignant celis.



Figüre 4. Microinvasive focus of the tumor (H&E, x 400)

DISCUSSION

Primary amenorrhea, sterility and pelvic pain are the main expected gynecological problems in MRKH syndrome. Having ovarian neoplasms together with this syndrome is an unexpected state. Although very few cases of this association are reported in the literatüre; it is essential to keep in mind that women with such anomalies should stili be followed gynecologically because of the normal risk of developing ovarian neoplasm (2-4).

As described by some authors, by using various imaging techniques (intravenous urography, ultrasonography, radiographies of the vertebral column) and diagnostic laparoscopy, MRKH syndrome may be divided into two forms: symmetric muscular bulbs and fallopian tubes are diagnostic of type A (typical form), asymmetric muscular bulbs or abnormally developed fallopian tubes are diagnostic of type B (atypical form) (5). Discrimination between these two types of MRKH syndrome is important, because associated renal, skeletal, ear and ovarian abnormalities occur only in type B (5-6). According to this grouping system, because this case had asymmetric bilateral rudimentary buds, with abnormally developed fallopian tubes and an ovarian neoplasm, it was evaluated as type B.

All patients with vaginal agenesis must be regarded primarily as a MRKH syndrome and they must be investigated to establish whether there were any associated congenital anomalies. In recent years, various radiologic techniques especially magnetic resonance imaging (MRI) became a confident, noninvasive technique in diagnosis of MRKH syndrome (7) and it is believed that it can be more precise than laparoscopy and sonography in defining the anatomical characteristics of this syndrome and it also is less expensive than laparoscopy (8). Especially in such case with pelvic mass, MRI can also predict the features and the origin of the mass, therefore MRI should be one of the first diagnostic evaluation methods to avoid excess intervention.

Treatment choice of this rare syndrome is limited and depends on the symptoms, associated anomalies and the age of the patient. If the patient suffers of pelvic pain due to a rudimentary horn, resection of this structure by laparotomy or laparoscopy may resolve the problem (1). If diagnosis is made on an amenorrheic pubertal girl, a vaginoplasty operation could be applied. Our patient refused such an operation. Estrogen replacement therapy was given to avoid postmenopausal problems.

REFERENCES

1. Giatras K, Licciarddi F, Grifo JA: Laparoscopy for pelvic pain in the Mayer-Rokitansky-Kuster-Hauser syndrome. J Reprod Med 1998;43:203-205.
2. Koonings PP, Al-Marayati L, Schlaert JB, Lobo RA: Mayer-Rokitansky-Kuster-Hauser syndrome associated with endodermal sinüs tumor of the ovary. Fertil Steril 1991;56:577-578.
3. Fisher K, Esham RH, Thorneycroft I: Scoliosis associated with typical Mayer-Rokitansky-Kuster-Hauser syndrome. South Med J 2000;93:243-246.
4. Tsaur GT, Lee MH, Su SL, Wu, MJ, Huang TW: Mayer-Rokitansky-Kuster-Hauser syndrome with immature teratoma of the ovary at age 4 years. Gynecol Oncol 1995;56: 456-459.
5. Strubbe EH, Willemsen WN, Lemmens JA, Thijn CJ, Rolland R: Mayer-Rokitansky-Kuster-Hauser syndrome: distinction between two forms based on excretory urographic, sonographic and laparoscopic findings. Am J Roentgenol 1993;160: 331-334.
6. Strubbe EH, Cremers CW, Willemsen WN, Rolland R, Thijn CJ: The Mayer-Rokitansky-Kuster-Hauser syndrome without and with associated features: two separate entities? Clin Dysmorphol 1994;3:192-199.
7. Russ PD, Allen-Davis JT, Weingardt JP, Anderson MS, Köyle MA: Mayer-Rokitansky-Kuster-Hauser syndrome diagnosed by magnetic resonance imaging in a 15-year-old girl. J Pediatr Adolesc Gynecol 1997; 10: 89-92.
8. Fedele L, Dorta M, Brioschi D, Giudici MN, Candiani GB: Magnetic resonance imaging in Mayer-Rokitansky-Kuster-Hauser syndrome. Obstet Gynecol 1990; 76: 593-596.