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

MAYER-ROKITANSKY-KUSTER-HAUSER SENDROMU İLE İLGİLİ ÖLGÜNÜ İZLENEN OVERİN BORDERLINE SERÖZ KİSTADENOMU: OLGU SUNUMU

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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.

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, axillary 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.

Erythrocyte 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 hyperecogenenous 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.

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 lymphadenopaties in the abdomen or in the pelvis.

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 eosinophytic 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 cells.

**Figure 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 literature; it is essential to keep in mind that women with such anomalies should still 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**