Vulvar and Periclitoral Endometrioma: Case Report

Vulvar ve Periklitoral Endometrioma Olgusu

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Yazışma Adresi/Correspondence: Rabia YILDIRIM AKBAŞ, MD Ankara Training and Research Hospital, Clinic of Gynecology and Obstetrics, 06340, ANKARA rabiayildirim@hotmail.com **ABSTRACT** Endometriosis is a common benign gynecologic disease characterized by the presence of ectopic endometrial tissue, outside of the uterus. Vulvar endometriosis is a very uncommon disorder. When endometriosis develops as a distinct mass, the condition is classified as an endometrioma. In this article we report a dramatic case of vulvar and periclitoral endometrioma. A 24 years old woman had approximately 3-4 cm size palpable mass and pain in the periclitorial region. The mass was excised surgically under general anesthesia and pathology reveals that it was endometrioma. With this case, we emphasized endometrioma in the differential diagnosis of vulvar tumors and tried to summarize the literature about etiopathogenesis and diagnostic/therapeutic approach of vulvar endometrioma.

Key Words: Clitoris; endometriosis; vulva

ÖZET Endometriozis sık görülen benign bir jinekolojik hastalıktır. Endometrial dokunun uterus dışında bir yerde lokalize olması ile karakterizedir. Vulvar endometriozis ise nadir görülen bir tipidir. Endometriozis sınırlı kitle şeklinde karakterize olduğu zaman endometrioma olarak sınıflandırılır. Bu makalede tipik bir vulvar, periklitoral endometrioma olgusu rapor ettik. Yirmi dört yaşındaki hastanın periklitoral bölgesinde yaklaşık 3-4 cm boyutlarında palpabl ve ağrılı kitlesi mevcuttu. Kitle genel anestezi altında cerrahi eksizyonla çıkarıldı ve patoloji sonucu endometrioma olarak rapor edildi. Bu olgu vesilesi ile vulvar endometriomanın diğer vulvar tümörlerden farkını, etiyopatogenezini, tanısal ve terapötik yaklaşımını vurguladık.

Anahtar Kelimeler: Klitoris; endometriozis; vulva

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Hence the presence of ectopic endometrial tissue, outside of the uterus.¹ Endometriosis is a benign condition affecting 15-20% of women with a child-bearing potential. Most commonly it affects organs such as the ovaries, uterine ligaments, fallopian tubes, rectum and the cervico-vaginal region. Involvement of the urinary tract rare and seen in about 1% of patients.² When endometriosis develops as a distinct mass, this condition is classified as an endometrioma.³ In this article we report a dramatic case of vulvar and periclitoral endometrioma. A 24 years old woman had palpable mass and pain in the periclitorial region. A brown-black palpable mass at approximately 3-4 cm size is detected in pelvic examination of the vulva and the periclitorial area. The laboratory findings were non specific and the pelvic ultrasound was normal. An accessional biopsy was performed. The result of the pathological examination of the total accessed mass: extensive bleeding areas, in the cystic structure lined by the stratified squamous epithelium, macrophages with hemosiderin, endometrium glands and stroma was observed, the patient has been diagnosed with endometriosis.

CASE REPORT

Female patient aged 24 applied due to the lump, palpable mass and pain in the periclitorial region (Figure 1). The patient, who had noticed a small lump (at a size of lentil grain) adjacent to the clitoris one year ago, mentioned that this painful lump which softens during the menstruation and hardens afterwards, has grown bigger slowly. It is understood from the history that Gravida 1, parity 1, she had normal, spontaneous vaginal delivery, has been using intrauterine device as contraception for 4 years, she had regular periods every 28 days, the periods lasted for 7 days and the bleeding amount was much more than normal, she had groin pain and back pain; she had pain in the mentioned lump area. The patient who had pain during urination, frequently urinating and stress inconti-



FIGURE 1: Female patient aged 24 applied due to the lump, palpable mass and pain in the periclitorial region



FIGURE 2: A brown-black palpable mass at approximately 3-4 cm size is detected in pelvic examination of the vulva and the periclitorial area.

nence complaints, didn't have hematuria. The patient didn't have any internal diseases and wasn't using regular medications. A brown-black palpable mass at approximately 3-4 cm size is detected in pelvic examination of the vulva and the periclitorial area (Figure 2). The laboratory findings were non specific and the pelvic US was normal. An accessional biopsy was performed (Figure 3a, 3b) The result of the pathological examination of the total accessed mass: extensive bleeding areas, in the cystic structure lined by the stratified squamous epithelium, macrophages with hemosiderin, endometrium glands and stroma was observed, the patient has been diagnosed with endometriosis. She didn't take any medical treatment after operation, the result of surgical treatment was good.

DISCUSSION

Endometriosis is a common disorder of women of reproductive age, yet diagnosis of this condition is often problematic. The most frequent clinical presentations of endometriosis include dysmenorrhea, pelvic pain, dyspareunia, infertility, and pelvic mass. However, the correlation between these symptoms and the stage of endometriosis is poor., Endometriosis is the presence of endometrial-like tissue outside the uterine cavity, which induces a

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chronic inflammatory reaction. It can occur in various pelvic sites such as on the ovaries, fallopian tubes, vagina, cervix, or uterosacral ligaments or in the rectovaginal septum. It can also occur in distant sites including laparotomy scars, pleura, lung, diaphragm, kidney, spleen, gallbladder, nasal mucosa, spinal canal, stomach, and breast. The exact cause and pathogenesis of endometriosis is unclear. Several theories exist that attempt to explain this disease though none have been entirely proven.

Previous theories suggest that endometriosis results from the transport of viable endometrial cells through retrograde menstruation. Cells flow backwards through the fallopian tubes and deposit on the pelvic organs where they seed and grow. A population of cells reside in the endometrium, which retain stem cell properties. It may be these properties that allow these cells to survive in ectopic locations.

Retrograde menstruation is a common physiologic event. Diagnostic laparoscopy during the perimenstrual period has shown that as many as 90% of women with patent fallopian tubes have bloody peritoneal fluid. Since most women do not have endometriosis, perhaps immunologic or hormonal dysfunction leaves some women predisposed. Recent research has suggested involvement of the immune system in the pathogenesis of endometriosis. Women with this disorder appear to exhibit increased humoral immune responsiveness and macrophage activation while showing diminished cell-mediated immunity with decreased Tcell and natural killer cell responsiveness.

Transtubal dissemination is the most common route, although other routes have been observed. These include lymphatic and vascular channels. This may explain how endometrial tissue can be found at distant locations in the body. The most common locations for endometrial growth are in the:

- cul-de-sac, or pouch between the uterus and rectum
- uterosacral ligaments, or ligaments that attach the uterus and cervix to the base of the spine
- ovaries (egg producing glands)
- the surface of the uterus
- bowel
- rectum
- bladder
- peritoneum (lining of the abdominal cavity).

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Metaplasia, or the changing from one normal type of tissue to another normal type of tissue, is another theory. The endometrium and the peritoneum are derivatives of the same coelomic wall epithelium. Peritoneal mesothelium has been postulated to retain its embryologic ability to transform into reproductive tissue. Such transformation may occur spontaneously, or it may be facilitated by exposure to chronic irritation by retrograde menstrual fluid.

Another theory states that remnant mullerian cells may remain in the pelvic tissues during development of the mullerian system. Under situations of estrogen stimulation, they may be induced to differentiate into functioning endometrial glands and stroma.

Finally, iatrogenic deposition of endometrial tissue has been found in some cases following gyne-cologic procedures and cesarean sections.

Some women may have a genetic predisposition to endometriosis. Studies have shown that firstdegree relatives of women with this disease are more likely to develop it as well. The search for an endometriosis gene is currently underway. Many theories exist as to why endometriosis occurs, and it is likely a combination of these factors that cause and determine severity of disease.

Currently available laboratory markers are of limited value. At present, the best marker, serum CA-125, is usually elevated only in advanced stages and therefore not suitable for routine screening⁴ Ectopic endometrium has been described in almost every location of the female body but is most commonly located in the pelvic organs⁵ The term endometrioma is used when endometriosis appears as a circumscribed mass.⁶

In this article we reported a dramatic case of vulvar and periclitoral endometrioma. A 24 year old woman had palpable mass and pain in the periclitorial region. The mass was excised surgically under general anesthesia and pathology reveals that it is endometrioma. With this case, we emphasized endometrioma in the differential diagnosis of vulvar tumors and tried to summarize the literature about etiopathogenesis and diagnostic/therapeutic approach of vulvar endometrioma.

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