Recurrent Vulvar-Perineal Endometriosis

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Abstract

Endometriosis, which is defined as the presence of endometrial glands and stroma outside the endometrium, most commonly affects pelvic peritoneal surfaces, ovaries, and uterine ligaments. Even it is quite rare, endometriosis may affect the vulva, vagina, or perineal region, generally secondary to obstetric or surgical trauma. In this paper, we described a virgin patient with perineal endometriosis, who did not have any history of perineal or vulvar surgery, and had recurrence of the lesion despite surgical and medical therapies.

Key words: endometriosis, vulva, perineum, recurrence, surgery

Özet

Rekürren Vulvar-Perineal Endometriozis

Endometriosis, endometrial gland ve stromanın endometrium dışında bulunmasıdır. En sık yerleşim yeri peritoneal yüzeyler, overler ve uterin ligamanlar olmakla birlikte nadiren ve genelikle de obstetrik ya da cerrahi bir travma sonrası, vulvar, vaginal ve perineal yüzeylerde de yerleşebilir. Bu yazıda, geçirilmiş obstetrik ve/veya cerrahi travma öyküsü bulunmayan bir genç kızda rekürrens gösteren bir vulvar-perineal endometriozis olgusu sunulmuştur.

Anahtar sözcükler: endometriosis, vulva, perine, rekürrens, cerrahi

Introduction

Endometriosis is defined as the presence of endometrial glands and stroma outside the endometrium, including myometrium.1 It is more frequently observed in pelvic structures such as pelvic peritoneum, ovaries, broad ligaments, round ligaments, whereas uncommonly it occurs as cutaneous lesions. Among the common cutaneous lesions of endometriosis are those occurring on anterior abdominal wall, especially on the umbilicus. Endometriotic lesions may also be encountered on the scar tissue due to previous surgery.2 Rarely, these lesions occur on the vulvar and perineal area. An outstanding shared feature of the reported cases of perineal and vulvar endometriosis in the literature is previous history of surgical and/or obstetrical trauma. 3-5

Case

A 19-year-old virgin patient presented to our clinic with the complaints of a hyperemic, burning, itchy, papullar lesion that had been lasting for one year, the symptoms of which aggravated during menstruation. On visual inspection a pale red, papullar cutaneous lesion of 3x2 cm in size located on the posterior confluence of major labias, lying over the perineum was detected (Fig. 1). On gynecological and ultrasonographic examination internal pelvic organs were detected normal. Histopathologic examination of the lesion revealed *"endometriosis externa"* (Fig. 2). To confirm the diagnosis, 5Fm slides were immunostained for estrogen and progesterone receptors using "s*treptavidin biotin peroxidase"* technique, and the lesion was demonstrated to be strong positive for estrogen and progesterone receptors.

The endometriotic lesion was completely excised with a 5-mm lesion free margin. Postoperatively oral contraceptives were started on to suppress ovarian functions. On the 3rd postoperative month the patient applied to our clinic for the recurrence of the symptoms and reappearance of the lesion in the same area. On physical examination a 2x1 cm dark white-pink colored lesion located on the scar tissue of the previously excised lesion was detected. The lesion including scar tissue was excised as a whole and submitted for histopathologic evaluation, which was later demonstrated to be *"endometriosis externa"*.

Discussion

Most of the endometriotic lesions in the vulvar and perineal area occur secondary to obstetrical and surgical trauma. Furthermore, a big proportion of these lesions are observed on episiotomy scars.4,5 In addition, vulvar-perineal endometriotic lesions after radical vulvectomy and vulvar LEEP (loop electrosurgical excision procedure) have also been reported.2,6 In a case series of 6 patients by Liang *et al.,* all endometriotic lesions occurred on the episiotomy scars after vaginal delivery. Cases of clear cell adenocarcinoma and endometrial stromal sarcoma, which

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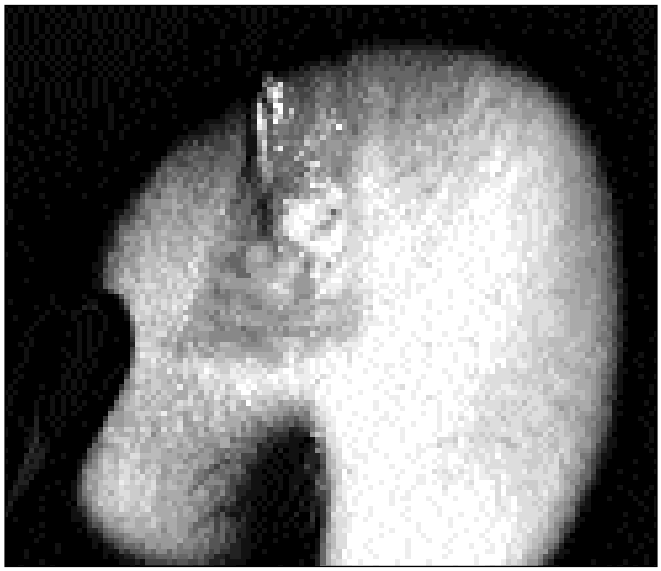

Figure 1. Vulvar-perineal endometriosis.

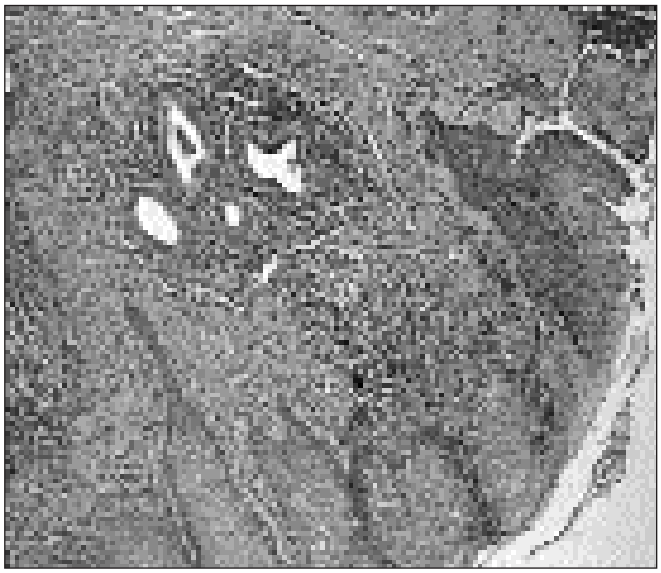

Figure 2. Endometriotic lesions in vulvar tissue (H&E, X100).

were originated from the vulvar endometriotic lesions, were also reported.7-9

Many theories were put forward to explain the etiopathogenesis of endometriosis, however none of them is able to clarify the etiopathogenesis of all endometriotic lesions. Vulvar and perineal endometriotic lesions can be explained by *"implantation theory"* which suggests that viable endometriotic cells shed during menstruation might implant in these areas.5 Eventhough all of these cases could not be explained by this theory, especially the lesions on the episiotomy scars, which are associated with disruption of the epithelial integrity can be explained by this theory. However as in the present case, in which the epithelial integrity is intact, and in theory implantation of the endometrial cells through an intact epithelial lining is out of concern, possibility of occurrence of an unnoticed microtrauma disrupting the epithelial integrity should be kept in mind. On the other hand the fact, that pelvic endometriosis is not observed in every women who has retrograde menstruation, and endometriotic lesions on episiotomy scars are not observed in every women with episiotomy scars, brings the other factors to mind such as immunologic and angiogenic factors that might take role in the etiopathogenesis of endometriosis.10

Angio-lymphatic metastasis theory and mechanical transplantation of endometrial cells during a surgical procedure (transplant theory) are among the others, which were put forward to explain the etiopathogenesis of other ectopic and vulvar-perineal endometriotic lesions. Besides, it was suggested that these lesions could occur by differentiation of multipotent coelemic cells in to endometrial cells.1,2 Eventhough with this last theory especially lesions observed in the lungs and eyes which have no direct contacts with the endometrial tissue could be explained, it has not been proved yet.

One of the shared characteristics of these lesions in the perineal and vulvar area is the aggravation of the symptoms such as pain, itching and burning during menstruation. The cyclical occurrence of the symptoms is an important point, which can be used to differentiate these lesions from the other ones. Infrequently, these lesions might enlarge during menstruation due to internal hemorrhage and appear as an endometrioma.5 Histopathologic examination of the specimen constitutes the basis of the diagnosis that can be done by insicional, excisional or needle biopsy.11

Primary treatment modality is the surgical excision of the lesion. In addition to surgery, postoperative suppression of the ovarian functions might be offered. Since recurrence is likely, and there are some reports of malignant transformation, margins of the lesion should be taken account during surgical excision. The recurrence of endometriosis after surgical removal of the initial lesion is an extremely rare. Liang *et al.,* observed recurrence in a patient treated with only surgery, however none of the patients treated with surgical and medical combination had recurrence of disease.5 When symptoms of vulvar endometriosis reappear several months after treatment, it is difficult to distinguish between recurrence and persistence of the disease, an surgery to remove the recurrent lesions used to be the mainstay of treatment.

Vulvar endometriosis is an infrequent clinical condition, in which to confirm the diagnosis immunohistochemistry studies for estrogen and progesterone receptors should be done. Eventhough primary treatment includes surgery and in some cases medical suppression of ovarian functions should be necessary, recurrence is likely to occur as in our case.

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