Vulvar Endometriosis in a Postmenopausal Woman: A Case Report

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Abstract

Endometriosis is a benign gynecological disorder that is characterized by the presence of ectopic endometrium outside the uterus. Endometriosis can be found in atypical locations both within and outside the pelvis. This disease mainly occurs during the reproductive years and is rarely diagnosed after menopause. Vulvar involvement of endometriosis is a rare condition, especially when not associated with local previous trauma (e.g. episiotomies) and in postmenopausal women. We present a case of a 46-years old woman who presented with vulvar lump that was surgically removed and later diagnosed with vulvar endometriosis.

Introduction

Endometriosis is characterized by the presence of endometrial tissue outside the endometrium, which induces a chronic inflammatory reaction. This condition is predominantly found in women of reproductive age [1], because of the estrogen dependence of the ectopic tissue, however it can affect women from pre-menarche until postmenopause [2], regardless the race or ethnicity. Depending on the area affected, endometriosis is characterized as endopelvic or extrapelvic disease. Most commonly, endometriosis spreads into the pelvis affecting the ovaries, pelvic peritoneum, uterosacral ligaments, fallopian tubes and broad ligaments. Although more unusual, it can be also found in abnormal locations both within and outside the pelvis. Endometriosis, in fact, can affect different and multiple organs outside the genital tract like abdominal wall, scars of perineum, urinary and gastrointestinal tract, lungs and brain [3]. Cutaneous endometriosis is a rare, but still a possible endometriosis location. It is often secondary to skin trauma occurring on scar tissue as after a Caesarean section or myomectomy. The pathologic mechanism of intrapelvic endometriosis is generally accepted as being largely due to retrograde menstruation. However, the mechanisms by which extrapelvic endometriosis forms still does not find a univocal explanation and various theories have been developed. The mechanism thought to be responsible of cutaneous endometriosis is thought to be the seeding of endometrial tissue in the surgical incision after the entry into the endometrial cavity or after an episiotomy. Endometriosis of the perineum and the vulva has been reported in literature, with the most common site being the episiotomy scar [4]. However, primary vulvar endometriosis, not associated with episiotomies or trauma, is extremely rare. Extragenital or extrapelvic endometriosis represents a rare condition that is still challenging to diagnose as long as it comes with a wide variety of symptoms.

Since it is an estrogen dependent pathology, it is generally believed that endometriosis is related to ovarian function and that it tends to reverse after menopause. However, it is becoming increasingly clear that endometriosis is a problem affecting not only reproductive age, but also postmenopausal women. The physiopathology of endometriosis in this particular group of patients is even more complex since it is not known whether it is a continuation of a previously existing disease or it develops de novo. Postmenopausal endometriosis is rare, because of the reduction or absence of estrogen hormone production. During menopause the source it thought to be mainly peripheral, especially in adipose tissue, adrenal glands and eventually exogenous sources (Hormone Replacement Therapy- HRT) [5].

Case Presentation

A 46-years old woman gravida 0, para 0, came into our gynecological emergency department...
complaining painful vulvar lump, on the right side of the labia majora. The patient reported a story of dysmenorrhea 10/10 (using the Visual Analogue Scale), heavy menstrual periods, previous unexplained infertility and a prior history of inguinal tuberculosis medically treated in 2015. No mention about previous diagnosis of endometriosis during her life. No other medical known conditions or current pharmacological therapy. At the moment she came to our department she referred menopausal status started two years before and no HRT was previously assumed by the patient.

The gynecological inspection showed a tense lesion measuring about 3.5 cm within the lower internal fold of the right labia majora. The lesion was tender and painful on palpation and had a hard-elastic consistency. The overlying skin was not reddened or warm.

At the translabial ultrasound the mass appeared uniloculated, with regular margins and wall, ground glass echogenicity and poor vascularization on Color Doppler (Color Score 2). It measured 39 mm in length, 34 mm in height and 38 mm in width (Figure 1). Uterus and adnexa showed regular appearance at transvaginal ultrasound evaluation.

Due to the macroscopic and ultrasonographic characteristics of the lesion, the suspect of an abscess or even a recurrence of tuberculosis was arisen and patient was given antibiotics and painkillers for 6 days as first line therapy. Despite the therapy, after a week, the patient was still complaining pain and swelling and the lesion appeared unchanged at the clinical and ultrasonographic evaluation. With the patient consent, it was decided to perform surgery in order to remove the lesion. After counseling for surgical management to remove the lesion, the patient accepted for surgical treatment, and the written consent was recorded. Surgical approach was included in the routine procedure of our department, and no IRB approval was required.

Surgery was performed under spinal anesthesia, with the patient in gynecological position and a urethral Foley’s catheter in situ. A longitudinal incision at the medial face of the minora right labia was made and the skin flaps were reflected. The lesion was exposed by blunt dissection and entirely excised. After bleeding control, the tissue was repaired in two layers. Cut section of the lesion showed uniloculated cyst with chocolate fluid content (Figure 2). The lesion was sent to histopathological examination and the definitive diagnosis referred to glandular endometriosis.

Postoperative recovery was fast and the patient was discharged the day after the surgery.

**Discussion**

Despite the fact that up to 12% of patients with endometriosis have extrapelvic disease [6], it still remains a misdiagnosed entity.

We report a case of a vulvar location of endometriotic lesion in a 46-years old nulliparous postmenopausal woman who presented with a history of vulvar lump and pain in the right side of the labia majora. Vulvar location of endometriosis is a rare phenomenon and very few cases are reported in literature, in most cases near or at the site of surgical scars [4]. The mechanisms that lead to the development of extrapelvic lesions are still unclear. It can occur via iatrogenic mechanical transplantation of endometrial tissue at surgical sites or accidental trauma, and this mechanism can explain the presence of endometriosis in previous episiotomy sites or abdominal wall scars after a Caesarean section. In contrast, the lymphovascular dissemination of endometriomal tissue could be responsible for the pathogenesis of primary vulvar endometriotic lesion. The occurrence of endometriosis in the vulva, in fact, could be explained through the extension of endometriomal tissue from the pelvis through the round ligament to the vulva. In our patient, the lesion involved the right side of labia majora. Although very rare, the occurrence of endometriosis in the round ligament has been reported in literature, involving the right side in 90% of cases [7]. A possible explanation is the presence of atypical lymphatic flows from the peritoneal cavity and pelvis to the right groin, or non-obliteration of the parietal peritoneum accompanying the round ligament in the canal of Nuck [8].

Vulvar endometriosis is a rare condition and its diagnosis can be difficult. The differential diagnoses should include Bartholin’s cysts, abscesses, Skene’s duct cysts, lipoma’s as well as vascular or squamous cell tumors. Pain is the most common presenting symptom; although a wide range of clinical manifestation can occur, like superficial dyspareunia, occasional discharge or cyclical swelling [9].

The other interesting characteristic of this case is that it involved a postmenopausal woman who was never diagnosed with endometriosis during her reproductive age, although she reported some of the most typical endometriotic symptoms such as story of dysmenorrhea, heavy menstrual periods and previous unexplained infertility. In the majority of cases, the hypo-estrogenic status induced by the cessation of ovarian activity leads to regression of endometriotic lesion. However, some ectopic endometrial location can remain active after menopause and also the occurrence of de novo lesion has been reported in literature, although it remains difficult to ascertain whether they truly are de novo lesions or pre-existent lesions that became symptomatic after menopause. Postmenopausal endometriosis may be more likely a progression of previously existing disease. In fact, since endometriosis does not always present with classic symptoms, its prevalence could be underestimated and some
women without symptoms during reproductive age could progress into the disease and become symptomatic during menopause [10]. Even after menopause, endometriosis still remains an estrogen related disease and the source of estrogen results mainly from the peripheral conversion of androgens or eventually exogenous sources (HRT). The pathogenic mechanism in post-menopause may involve the estrogen threshold theory; according to this theory some undetected or transient foci of endometriosis may be activated post-menopaussally when estrogens are above their threshold level [5].

An intrinsic mechanism that could enhance the production of estrogens it thought to take place into the endometriotic lesions themselves. The leading estrogen is Estrone (E1); it can be converted to Estradiol (E2), the active form of estrogens, in endometriotic tissue. E2 is not metabolized in the ectopic endometriotic tissue due to the lack of the 17-β hydroxyl steroid dehydrogenase type 2. Increased local E2 promotes local production of prostaglandin E2 that is a potent stimulator of aromatase enzyme. So, a positive feedback loop is established and this could explain how endometriosis lesion may persist and become symptomatic after menopause.

Our patient reported a personal history of symptomatic disease that was never promptly diagnosed during her reproductive age. Maybe, in this case, the vulvar lesion found in menopause could have represented a continuation of a previous endometriotic disease, even in the absence of a specific diagnosis.

Conclusion

Endometriosis is a benign gynecological disease that can involve different organs and tissues as well as women of all age, from promenarche to post-menopause. Although rarely, vulva can also be affected. Endometriosis of the vulva must be taken into account when facing a lesion in a patient with typical clinical signs (dysmenorrhea, infertility, ultrasonographic features). After menopause, detailed anamnestic data relative to gynecological history can be helpful. Clinical diagnosis of skin endometriosis remains challenging and malignant conditions must be ruled out. Radical surgical excision represents the treatment of choice, since it allows definitive diagnosis and a fast recovery.

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IRB

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References