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Case Presentation

A Rare Case of Vulvar and Umbilical Endometriosis in a Single Patient: A Case Report

Pathiraja PDM[1]* and Ranaraja SK[2]

[1]Post Graduate Trainee in Obstetrics and Gynecology, Ministry of Health, Sri Lanka

[2]Consultant Obstetrician and Gynecologist, Teaching Hospital Peradeniya, Sri Lanka

***Corresponding author:** Pathiraja PDM, Post Graduate Trainee in Obstetrics and Gynecology, Ministry of Health, Sri Lanka

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Introduction

Endometriosis is a benign disorder and characterized by ectopic endometrium like tissues outside the uterus. We report a case of a 31 year-old woman referred to our clinic due to complaints of a vulvar and umbilical mass and periodic swelling with pain of the mass at the time of menstruation. The cyst was removed totally. Histopathological examination showed findings compatible with endometriosis in both. We have treated them with suppressive treatment with Depot Medroxy Progesterone Acetate (DMPA) and after six months patient was completely asymptomatic.

Case Report

A 31-year-oldmother of a one child presented for a gynecologic consultation with increasing dyspareunia and cyclical vulvar and periumbilical pain for one-year duration. Previous treatment with analgesics did not alleviate the symptoms. The patient had a regular menstrual cycle with periodic swelling of the vulval and umbilical masses could suggest endometriosis. The patient did not have a history of vulvar or abdominal surgeries.

On physical examination, revealed a 2x2 cm size mass in the umbilical region and that was tender to palpation (Figure 1). Vulval examination revealed that 3x2 cm size tender mass in right vulval region and covered by normal skin (Figure 2). Her biochemical markers were normal including CA 125. Patient underwent fine needle aspiration and reports revealed that fairly cohesive sheets of epithelial cells with very mild nuclear plemorphism and background shows foamy macrophages. Aspiration results suspect on endometriosis or neoplastic lesion.

Patient underwent surgery for complete excision of the both lumps. During surgery, the cyst ruptured and a chocolate-colored liquid escaped onto the surgical field which characteristic of endometriosis. Histology confirmed that section includes fibro fatty connective tissue with endometrial type glands and stroma. The hemorrhagic areas showed clusters of hemosiderin laden macrophages. After the surgery, patient had few cycles of DMPA injection and after six months she was completely asymptomatic.

Figure 1: Right vulvar lesion.

Figure 2: Umbilical lesion.

Discussion

Endometriosis is a common benign gynecological problem in reproductive women where presence of endometrial glands and stroma outside the endometrium lining [1]. This condition was first described by Von Rokitansky in mid 19th century. Endometriosis commonly found in ovaries (endometrioma), uterosacral ligaments, pelvic peritoneum and rarely found in previous surgical scar sites, vulva, pleura [2]. There are several theories to describe the involvement of the common and uncommon sites of the body. In this article we describe six different pattern of spread of the disease [3-5].

Retrograde menstruation

This is the commonly accepted theory as it explained how it could affect the fallopian tube, ovaries and other pelvic structures. Reflux of endometrial tissues can adhere and invade the peritoneum and developed its own blood supply. This leads to continuation of growth.

Lymphatic or vascular spread

This theory helps to explain the spread of the disease in unusual sites (perineum, groin, vulva) as according to the lymphatic drainage. Even retroperitoneal involvement could explained from this theory. And also this theory could explain the premenarcheal and post menopausal endometriosis.

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Coelomic metaplasia

The theory suggests that structures with common origin from coelomic epithelium can undergo metaplastic transformation. Since ovary, fallopian tubes and progenitor of the endometrium could develop the endometriosis.

Induction theory

This theory proposes the hormonal and biological factors could induce the differentiation of undifferentiated cells into endometrial tissues.

Hormonal dependence

Endometriosis is a disease of reproductive women and it is estrogen dependent. Usually, most of the estrogen do produce from ovary and some from peripheral tissues as well. Studies found endometriotic implants so express aromatase and 17 betahydroxysteroid dehydrogenase type 1, which are enzymes responsible for the conversion of androstenedione to estrone. With the estrogen dominant environment endometriosis lesions grow fast.

Role of immune theory

The menstrual tissues and endometrium which shed into the peritoneum usually get cleared by the macrophages and Natural Killer cells (NK cells). This theory explained, when the impaired regulation of the cellular and humoral immunity would leads to growth of endometriosis [6].

Although the typical complaints of patients with pelvic endometriosis include dysmenorrheal, menstrual irregularities, dyspareunia, and infertility. Patients with extra pelvic endometriosis like inguinal endometriosis and umbilical endometriosis present with unusual symptoms. Sometimes it makes a diagnostic dilemma. Periodic menstrual pain at the mass is sometimes the primary complaint. A history of a previous gynecologic procedure or surgical trauma is also contributory. Some patients may have no history of dysmenorrheal, pelvic pain, or dyspareunia as in our case. Although our patient had not undergone any vulvar or abdominal surgery that might have made implantation easier, endometriosis had occurred. Pruritus or trauma may have led to this implantation.

The underlying cause of this pain is unclear, but proinflammatory cytokines and prostaglandins released by endometriotic implants may be the source [7]. Additionally, there is also evidence to suggest that pain from endometriosis correlates with depth of invasion and that the site of pain may indicate lesion location [8]. Recent data suggest that endometriosis pain may result from neuronal invasion of endometriotic implants that subsequently develop a sensory and sympathetic nerve supply, which may undergo central sensitization.

National Institute for Clinical Excellence (NICE) guidelines suggest do not use CA 125 as a marker of severity or diagnose the endometriosis. Raised CA 125 may consistent with endometriosis and on the other hand endometriosis can be present with normal CA 125 level [9]. Diagnostic modalities such as radiological studies, ultrasound, and computed tomography scan have not been specifically helpful in the diagnosis [10]. The primary method of diagnose the endometriosis *via* visual assessment through laparoscopy. This may even without histological diagnosis. Since endometriosis varied from person to person it need a standard classification system. American Society Of Reproductive Medicine (ASRM) has develop a classification system in 1996 based on the laparoscopic appearance of the lesions. According to this endometriosis can be categorized in to minimal (stage 1), mild (stage 11), moderate (stage 111), and severe (stage 1V).

The appearance of these lesions by laparoscopy is varied and colors may include red (red, red-pink, or clear), white (white or yellow-brown), and black (black or black-blue) Dark lesions are pigmented by hemosiderin deposition from trapped menstrual debris. The gross appearance of endometriotic lesions often suggests certain microscopic findings. For example, when examined microscopically, red lesions are frequently vascularized, whereas white lesions more often display fibrosis and few vessels. The histopathology is helpful in diagnose the difficult cases. The histopathology requires the presence of both stoma and glands outside the uterus to confirm as endometriosis. Further, haemosiderin deposition and fibro muscular metaplasia can be observed.

The management of endometriosis depends on the location of the lesion, severity, pain score, fertility wishes and responds to medical treatments. Pelvic endometriosis can treat with medication which can create a pseudo pregnancy or pseudomenapausal state [11]. The ectopic lesion can be treated with suppressive medication like GnRH analogues (Gonadotropin Releasing Hormone analogue) or surgical evacuation [12]. In our case as patient not respond to standard treatment we have performed the excision and she was completely asymptomatic after the treatment. Use of Combined Oral Contraceptives (COCP) as a primary prevention of endometriosis is uncertain. Endometriosis itself does not increase the risk of cancer. However, some cancers (Ovarian and Non Hodgkin Lymphoma) are slightly common with patient with endometriosis.

Conclusion

The causation of an extra pelvic endometriosis remains unknown. As symptoms and signs of extra pelvic endometriosis are quite difficult in diagnosing, it is better to assess by histological analysis. The main stay of treatment for endometriosis by medication (Analgesia and Hormonal treatment), however, some need surgical clearance.

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