



Case Report

Case Report: Primary Umbilical Endometriosis without Dysmenorrhea or Dyspareunia

Sung-Tack Oh^{1,2*}, Bum-Chae Choi¹

¹Creation and Love Women's Hospital, Gwangju, Korea

²Chonnam University Medical School, Gwangju, Korea

***Corresponding author:** Sung-Tack Oh, Creation and Love Women's Hospital, 409 Sungam Officetel, 62 Jukbongdaero, Seogu, Gwangju 61932, Korea

Citation: Oh ST, Choi BC (2023) Case Report: Primary Umbilical Endometriosis without Dysmenorrhea or Dyspareunia. Ann Case Report. 8: 1140. DOI:10.29011/2574-7754.101140

Received: 19 January 2023, **Accepted:** 23 January 2023, **Published:** 26 January 2023

Abstract

Objective: This report presents a rare case of primary umbilical endometriosis and reviews the literature of it.

Case report: In a 42-year-old woman, umbilical mass with cyclic swelling and tenderness on menstruation was found. It was diagnosed pathologically umbilical endometriosis at small incision biopsy by general surgeon. She had no history of pelvic or abdominal surgery except local small incision biopsy by general surgeon. There was no history of endometriosis or endometriosis-associated symptoms. Therefore, monthly gonadotropin-releasing hormone (GnRH) agonist therapy was done for 6 months. The swelling and pain of umbilical mass were slightly decreased after this therapy. However, these symptoms were reappeared 6 months later again. Therefore, we decided the surgical removal of mass. The mass and adjacent tissue of umbilicus were much resected sparing the umbilicus under general anaesthesia. Although omphalectomy is choice of surgery, umbilicus was saved because of her request to save umbilicus. Histologic examination confirmed the diagnosis of umbilical endometriosis.

Conclusion: Umbilical endometriosis are rare and difficult to recognize, but the possibility of endometriosis must be considered to umbilical mass with cyclic swelling and tenderness on menstruation despite the absence of previous surgery. Complete excision of umbilical mass with adjacent normal tissue or complete surgical excision of umbilicus with mass are highly recommended.

Keywords: Primary Umbilical Endometriosis

Introduction

Endometriosis is the presence of endometrium tissue outside the uterine cavity. It is found at 10–15% of all premenopausal women and 6% of perimenopausal women [1,2]. Endometriosis is mainly found in the vulva, vagina, cervix, ovaries, and pelvic peritoneum. However, it is sometimes found at extra-genital sites including the intestinal tract, urinary tract, lungs, thoracic cage, surgical scars, abdominal wall, inguinal area and umbilicus in lesser than 12%. Secondary umbilical endometriosis can occur on scars of abdominal wall after gynaecologic surgeries such as hysterectomy, caesarean section, and laparoscopic operation [3-

8]. However, primary umbilical endometriosis is not associated with surgical procedure and the prevalence is very rare. The exact prevalence is still not well-known but the incidence is reported to be 0.5% - 1% of all extra-genital endometriosis [9]. Primary umbilical endometriosis was described by Villar for the first time in 1886 [10]. When concurrent pelvic endometriosis is present, possible theory of umbilical endometriosis is explained to haematogenous or lymphatic spread [11]. If pelvic endometriosis is not present, it can be also explained to metaplasia of urachal remnants [11]. The diagnosis is mainly clinical by bluish purple, brown umbilical mass associated with aggravation of expansion and pain on every menstruation. We present an uncommon case of spontaneous umbilical endometriosis without dysmenorrhea, pelvic pain or dyspareunia or pelvic endometriosis.

Presentation of case

A 42-year-old multiparous woman with one son and one daughter suffering from a slowly growing swelling in the umbilicus that she noticed 3 years ago, and she also had umbilical tenderness during only menstruation for the last 2 years. She went to general surgeon, and it was diagnosed to umbilical endometriosis by small incision biopsy. On physical examination, a slightly tender nodule of 10 × 15 mm in size was palpated in the umbilicus without colour change. She had no history of abdominal operations except small incision biopsy of umbilical mass, and she did not receive any medication. She did not suffer from any symptom of pelvic endometriosis such as dysmenorrhea, abdominal pain or dyspareunia. She had regular menstrual cycles and did not use oral contraceptive. The concomitant pelvic endometriosis finding was not found on gynaecological physical examination and ultrasonography. At transvaginal US, the uterus and both ovaries appeared normal. Preoperative CA125 level and HE4 level were within normal range. At the first, monthly gonadotropin - releasing hormone (GnRH) agonist, therapy was done for 6 months. The swelling and pain of umbilical mass were slightly decreased after this therapy. However, these symptoms were reappeared 6 months later again. Therefore, we decided the surgical removal of mass. The umbilical mass with normal adjacent tissue were removed under general anaesthesia [Figure 1 and Figure 2]. Although omphalectomy is choice of surgery, umbilicus was saved because of her request to save umbilicus. Concomitant diagnostic laparoscopy with 5mm telescope via umbilicus was also performed to carefully inspect the abdominopelvic cavity and to exclude any coexisting pelvic endometriosis lesion. There was no endometriosis lesion in abdominal cavity. Histological examination confirmed the diagnosis of umbilical endometriosis. In the histopathological examination of resected tissues, a well-circumscribed endometriosis focus was identified with dilated glands localized with stratified squamous epithelium [Figure 3].

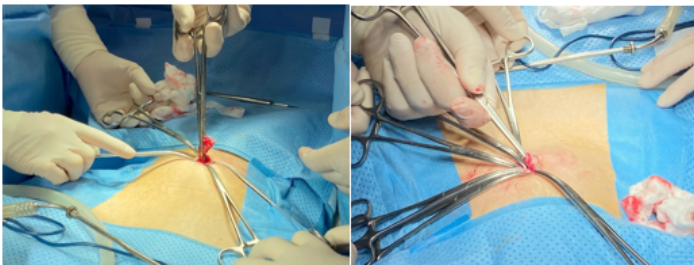


Figure 1: Operative findings of this case.



Figure 2: Removed tissue.

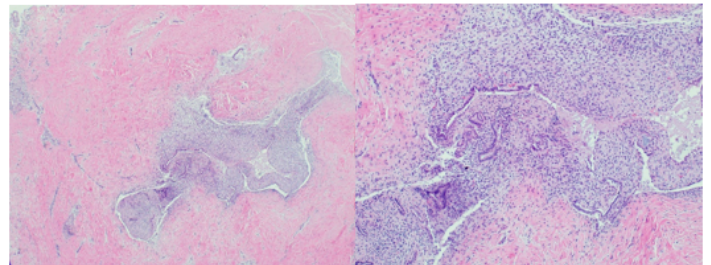


Figure 3: Pathological findings of removed tissue.

Discussion

Endometriosis is classically defined as an estrogen-dependent disorder, with the presence of endometrial tissue outside of the uterus in lesions of varying sizes and appearance. Endometriosis can be broadly classified as pelvic and extra-pelvic endometriosis. Umbilical endometriosis is a form of extra-pelvic endometriosis. The etiology of endometriosis is still debated, there are several theories. The “hypothesis of migratory pathogenesis” is the most widely accepted theory. According to main this hypothesis by Sampson, endometriosis was developed by retrograde menstruation thorough the fallopian tube to the pelvis. However, there are other theories such as caulomic metaplasia, lymphatic spread or haematogenous spread. The hypothesis of origin of extra-genital endometriosis is explained by them, endometrial tissues enter the uterine venous circulation and can then reach the brain, nasal mucosa, spine or other distant sites as endometriosis [12,13]. In metaplastic theory, the peritoneal mesothelial cells of caulomic origin change into endometrial cells by metaplastic transformation and it can be explained the endometriosis in the bladder and prostate of male [14]. The umbilical endometriosis with pelvic endometriosis can be explained by the theory of lymphatic or teratogenic transplantation. Secondary umbilical endometriosis may occur by iatrogenic spread of endometrial

cells after operations such as caesarean section and laparoscopy. However, primary spontaneous umbilical endometriosis without pelvic endometriosis is not explained by this theory. It can be explained by the metaplasia of urachus residues. The possibility of endometriosis must be considered during the evaluation of an umbilical mass despite the absence of previous surgery, and especially it is expanded and tender at only menstruation. Villar first reported primary spontaneous umbilical endometriosis in 1886. The prevalence is known for 0.5-1% of all cases of extra-genital endometriosis [15-17]. In primary umbilical endometriosis, there is an umbilical nodule, which causes periodic pain and may have bluish colour change at menstrual period. Sometimes, there may be a constant pain rather than periodic pain. In differential diagnosis of umbilical nodules, benign diseases such as haemangioma, umbilical hernia, sebaceous cyst, granuloma, lipoma, abscess, keloid, and urachus anomaly, and should always be considered [17]. The treatment of umbilical endometriosis has not been certain due to limited number of cases. The medical treatment to GnRH agonist has not provided sufficient cure results. However, some studies reported success in the reduced size of nodule and improvement of symptoms using this medical therapy [18]. However, almost of patients required surgical treatment [19]. The choice of surgery is total umbilical resection with or without repair of underneath fascia and peritoneum, but it can be performed local excision of endometrial nodule with preserving umbilicus. Total resection of umbilicus is mostly preferred because of recurrence [19, 20]. Local excision of endometrial lesion also should be performed by achieving adequate edge of the surrounding normal tissue in order to avoid local recurrence [18]. It is important with complete excision to perform the diagnostic laparoscopy in order to identify possible pelvic endometriosis lesion and to investigate peritoneal endometriosis. The successive histology is highly recommended for obtaining a definitive diagnosis and optimal treatment.

Conclusion

Primary umbilical endometriosis is a rare disease with a limited number of cases reported in the literature. Primary umbilical endometriosis should be differentially diagnosed without any previous surgical history, if the women present umbilical mass with painful and expanded cyclically on menstruation. Diagnosis is carried out by US, MRI or needle biopsy with the diagnostic laparoscopy in order to identify possible pelvic endometriosis lesion. The successive histology is highly recommended for obtaining a definitive diagnosis. Complete surgical excision of umbilicus with mass is the treatment of choice. If local excision only possible due to various reasons, local excision of endometrial lesion should be performed by achieving adequate edge of the surrounding normal tissue in order to avoid local recurrence

Conflicts of interest: The authors declare that they have no conflicts of interest.

References

1. Spaziani E, Picchio M, Di Filippo A, De Cristofano C, Ceci F, et al (2009) Spontaneous umbilical endometriosis: a case report with one-year follow-up. *Clin Exp Obstet Gynecol* 36: 63-264.
2. Rosina P, Pugliarello S, Colato C, Girolomoni D (2008) Endometriosis of umbilical cicatrix: case report and review of the literature. *Acta Dermatovenerol Croat* 16: 218-221.
3. Catalina-Fernández I, López-Presa D, Sáenz-Santamaria J (2007) Fine needle aspiration cytology in cutaneous and subcutaneous endometriosis. *Acta Cytol* 51: 380-384.
4. Agarwal A, Fond YF (2008) Cutaneous endometriosis. *Singap Med J* 49: 704-709.
5. Arava S, Iyer VK, Mathur SR (2010) Cytological diagnosis of peritoneal endometriosis. *J Cytol* 27: 77-78.
6. Kaushik R, Gulati A (2008) Inguinal endometriosis. A case report. *J Cytol* 25: 73-74.
7. Pathan ZA, Dinesh US, Rao R (2010) Scar endometriosis. *J Cytol* 27: 106-108.
8. Medeiros FC, Cavalcante DI, Medeiros MA, Eleuterio J Jr (2011) Fine needle aspiration cytology of scar endometriosis: study of seven cases and literature review. *Diagn Cytopathol* 39: 18-21.
9. Taniguchi F, Hirakawa E, Azuma Y (2016) Primary umbilical endometriosis: unusual and rare clinical presentation. *Case Rep Obstet Gynecol*. 2016: 9302376.
10. Kjer BD, Kjer JJ (2017) Primary umbilical endometriosis (PUE). *Eur J Obstet Gynecol Reprod Biol* 209: 44-45.
11. Laghzaoui O, Laghzaoui M (2001) Nasal endometriosis: apropos of 1 case. (2001) *J Gynecol Obstet Biol Reprod* 30: 786e8.
12. Barresi V, Cerasoli S, Vitarelli E, Donati R. (2006) Spinal intradural mullerianosis: a case report. *Histol Histopathol* 21: 1111e4.
13. Lagana AS, Sturlese E, Retto G, Sofo V, Triolo O (2013) Interplay between misplaced Müllerian-derived stem cells and peritoneal immune dysregulation in the pathogenesis of endometriosis. *Obstet Gynecol Int* 2013: 527041.
14. Marranci M, Dianda D, Gattai R, Nesi S, Bandettini L (2000) Umbilical endometriosis: report of a case and review of the literature. *Ann Ital Chir* 71: 389e92.
15. The WT, Vollenhoven B, Harris PI (2006) Umbilical endometriosis, a pathology that a gynecologist may encounter when inserting the verres needle. *Fertil Steril* 86: 1764.e1-2.
16. Khaled A, Hammami H, Fazaa B, Zermani, Ben Jilani S, et al (2008) Primary umbilical endometriosis: a rare variant of extra-genital endometriosis. *Pathologica* 100: 473-475.
17. Bimolchandra L, Devi HB, Rameshwar M, Devi RKP, Singh YA, et al (2008) Primary umbilical endometrioma: a case report. *JMS* 22: 85-86.
18. Sidani MS, Khalil AM, Tawil AN, El-Hajj MI, Seoud MA (2002) Primary umbilical endometriosis. *Clin Exp Obstet Gynecol* 29: 40e1.
19. Zollner U, Girschick G, Steck T, Dietl J (2003) Umbilical endometriosis without previous pelvic surgery: a case report. *Arch Gynecol Obstet* 267: 258e60.
20. Frischknecht F, Raio L, Fleischmann A, Dreher E, Luscher KP, et al (2004) Umbilical endometriosis. *Surg Endosc* 18: 347.