

Primary Umbilical Endometriosis: A Case Report and Literature Review

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Introduction: Endometriosis is a disease condition characterized by the presence of endometrial glands and stroma outside the uterine cavity and musculature. It affects 7–10% of women of reproductive age and usually involves pelvic organs. The common symptoms include dysmenorrhea, menorrhagia, pelvic pain, dyspareunia, and infertility. Umbilical endometriosis is the rarest form of extra-pelvic endometriosis.

Case Presentation: A 32-year-old Para II mother presented with umbilical swelling and pain that worsened during the menses of 2 months. She had two caesarean scars, both transverse Pfannenstien incisions. There was a dark blue, firm to hard 2×3 cm umbilical mass on physical examination. Laboratory results are normal. Ultrasound of the abdomen showed umbilical mass with homogeneous echotexture measuring 2 cm × 3 cm with an irregular border and hypoechoic texture. An excisional biopsy was performed and a biopsy showed umbilical endometriosis.

Discussion: Primary umbilical endometriosis is a disorder first described by Villar in 1886, and among all cases of extra-genital involvement of endometriosis, primary umbilical endometriosis accounts for 0.5–1%. The usual presentation of primary umbilical endometriosis is typically the presence of a discrete bluish-purple mass in the umbilicus, which becomes swollen, painful, and bleeds concomitantly with the menstrual cycle. Ultrasound and other imaging techniques help in making diagnoses, and the diagnosis is confirmed by cytological examination. The management of umbilical endometriosis involves surgical excision and reconstruction of the umbilicus.

Conclusion: Umbilical endometriosis is an uncommon form of extra-pelvic endometriosis. There is a wide differential diagnosis for swellings of the abdominal wall; however, it should be suspected in any female patient of reproductive age with an umbilical lesion that becomes more painful and swollen during her menstrual period. Appropriate clinical examination and workup are helpful to make a diagnosis and do a surgical excision.

Keywords: endometriosis, umbilical nodules, umbilical hernia, abdominal wall scar, caesareans section

Introduction

Endometriosis is a term first used by Sampson for a gynecologic disease condition that is characterized by the presence of endometrial glands and stroma outside the uterine cavity and musculature.¹ This problem occurs in 7–10% of women of reproductive age, and usually it affects the pelvic organs.^{1,2} It can also involve extra pelvic sites such as, the diaphragm, pulmonary, urinary tract, gastrointestinal tract, brain, and cutaneous endometriosis. Of all extra pelvic endometriosis, umbilical endometriosis, a form of cutaneous endometriosis, comprises 0.5–1%. Umbilical endometriosis can be primary, without any previous surgical scar at the site, or secondary, when it occurs following either laparoscopic or open procedure. The common symptoms of endometriosis include dysmenorrhea, menorrhagia, pelvic pain, infertility, and dyspareunia.^{1,2} However, endometriosis may involve non-pelvic organs, and particularly in patients who had gynecologic procedures, the occurrence of abdominal wall endometriosis is 0.03%–1.08%.² Primary umbilical endometriosis is also called Villar's nodule because it was first described by Villar in 1886. The pathogenesis of endometriosis is barely understood. One of the possible pathogeneses is postulated by Sampson, the so-called Sampson's theory of retrograde menstruation; others include coelomic metaplasia, induction theory, embryonic Mullerian rests, bone marrow stem cell theory, and hematogenous and lymphatic spread. In cases of umbilical endometriosis, hematogenesis or lymphatic theory is favoured where there is co-existing pelvic

endometriosis. However, isolated umbilical endometriosis is explained by metaplasia of urachal remnants.³ Here, we are presenting a case of primary umbilical endometriosis, which is a rare occurrence in the absence of a previous surgical scar.

Case Presentation

A 32-year-old female patient presented with umbilical swelling and pain of 2 months duration. The swelling was gradually increasing in size, and the pain worsens when she is on menses. Her menses are regular and come every 28 days. She has had no pelvic pain or other complaints. She has two children, one female, 11 years old, and the other male, 9 years old, both delivered through a caesarean section. She did not conceive for nine years, despite not taking any contraceptives. On physical examination, she appears healthy with vital sign parameters of blood pressure of 120/80 mmHg, a respiratory rate of 20 breaths/m, a pulse rate of 82 beats/m, and a temperature of 36.5.

Abdominal examination revealed a 3×2 cm firm irregular dark blue umbilical mass. The mass is fixed to the underlying structures and the overlying skin. Laboratory investigations show a normal range of complete blood counts and organ function tests. Her urine HCG (Human Chorionic Gonadotrophin) is negative. The abdominopelvic ultrasound finding was described as an umbilical mass with homogeneous echotexture measuring 2 cm × 3 cm with an irregular border and hypoechoic texture. Excisional biopsy was performed after an informed consent was obtained. Under local anesthesia and aseptic procedures, an elliptical incision was made, the mass was firm to hard and fixed to the skin, and the fascia was excised. The fascia was closed with Vicryl number two interrupted stitches, and the wound was finally closed in layer. The specimen was subjected to pathological analysis, and microscopic sections show skin-covered tissue with underlying endometrial glands and stroma embedded in dense fibrous stroma (Figure 1). At the time, this case report was written, the patient had a follow-up visit the second month after the surgery. She has no complaints, and on examination, the wound is healed, but there is a dark discoloration of the skin around the scar (Figure 2).

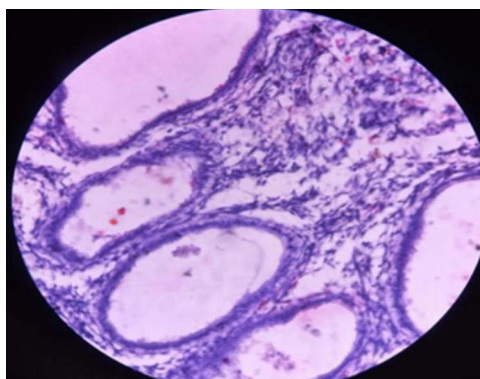


Figure 1 Microscopic tissue biopsy showing, skin-covered tissue with underlying endometrial gland and stroma embedded in dense fibrous stroma.

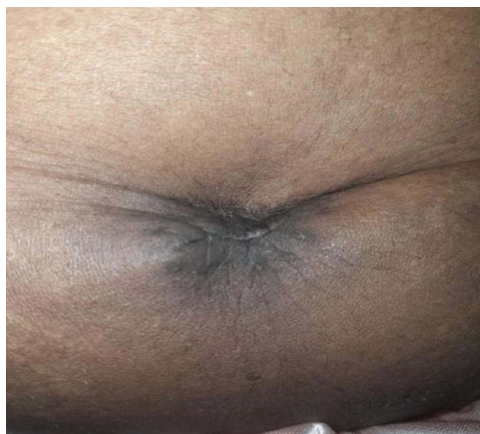


Figure 2 A post excision and reconstruction picture showing a scar with dark discoloration.

Discussion

Primary umbilical endometriosis is a disorder first described by Villar in 1886, and among all cases of extra-genital involvement of endometriosis, primary umbilical endometriosis accounts for 0.5–1%. The condition affects 6% of premenopausal women.^{4,5} The presence of extra-pelvic endometriosis can affect multiple organs and organ systems. The sites of involvement, from most frequent to least frequent, include the intestine, skin (including the umbilical and abdominal scars), inguinal region and thigh, lungs and pleura, pancreas, meninges, and vertebrae. Over all, cutaneous and subcutaneous endometriosis is believed to be caused by a cicatricial process following abdominal and/or pelvic surgical procedures, such as laparoscopy/laparotomy, caesarean section, hysterectomy, myomectomy, episiotomy, and appendectomy, removal of Bartholin's gland cyst, amniocentesis, and intrauterine injections for abortion.⁶

In 1920, Sampson proposed the theory that endometriosis is caused by retrograde menstruation passing through the fallopian tube in the pelvis. Other proposed mechanisms include.⁶ The ectopic presence of endometrial stem cells or immune system defects. Investigations into the genetic risk factors for endometriosis revealed that alterations to p53, PTEN, Cytochrome P450 1A1, and Peroxisome Proliferator-Activated Receptor γ 2 Pro-12-Ala have been implicated in endometriosis.⁷ Implantation of endometrial tissue into a surgical incision has been proposed as an explanation of the condition's pathophysiology. It is thought that during a cesarean delivery, endometrial tissue is inoculated into the incision.⁸

Umbilical endometriosis may be mistaken for a differential diagnosis like abdominal wall abscess, lipoma, hematoma, granuloma, neuroma, sebaceous cyst, incisional hernia, lymphoma, sarcoma, etc.⁸ Ectopic endometriosis is a rare event; it manifests as a painful subcutaneous mass that worsens during menses and usually bothers the patient for several years. In addition, endometriosis associated with scars may cause malignant changes.⁹

Our patient had a cesarean section 11 years ago and 9 years ago, both indicated due to her pelvic diameter. In both cases, the incision was a transverse pfannenstiell incision C-section, and there was no surgical procedure involving the umbilicus during both procedures. Her umbilical nodule and symptoms appeared after 9 years of non-scarred umbilicus. She had a 3 × 2 cm, firm, irregular, dark blue umbilical mass. In another similar case reported by Yahaya et al, typical symptoms of umbilical endometriosis were described as the presence of a discrete bluish-purple mass in the umbilicus that became swollen, painful, and bled concomitantly with the menstrual cycle. The diagnosis of umbilical endometriosis involves FNAC, biopsy, and immunohistochemistry.¹⁰

In the case, we present, the diagnosis was made by clinical examination, ultrasound, and excisional biopsy. According to several previous reports, the treatment of umbilical endometriosis is wide local excision including the peritoneum, and the subsequent reconstruction of the umbilicus guarantees the radicality of the procedure.¹¹ In most of these cases of umbilical endometriosis that have been reported in the literature, surgical treatment was performed successfully with a very low rate of recurrence. However, there is little experience concerning medical treatments. Some suggest long-term treatment with progesterone, danazol, and norethisterone, all of which showed conflicting results in very few patients. Gonadotropin-releasing hormone analogs and continuous treatment with a monophasic OC have never been applied to patients with umbilical endometriosis.¹² In the case of the present patient, surgical excision and reconstruction were done, and until the time this case was written, she only had dark skin discoloration. The patient was unable to conceive for nine years. This may strengthen the evidence that umbilical endometriosis may contribute to infertility.¹³

Conclusion

Umbilical endometriosis is an uncommon form of extra-pelvic endometriosis. There is a wide differential diagnosis for swellings of the abdominal wall; however, it should be suspected in any female patient with a lesion that becomes more painful and swollen during her menstrual period. Appropriate clinical examination and workup are helpful to make a diagnosis and do a surgical excision. In addition, attention should be given to ruling out the coexistence of pelvic endometriosis and infertility.

Abbreviations

CBC, Complete Blood Count; OFT, Organ Function Test; FNAC, Fine Needle Aspiration Cytology; HCG, Human Chorionic Gonadotrophin; OC, Oral contraceptive.

Ethical Approval

For publication of a case report at our institution, Jigjiga University, Shek Hassen Yabare Referral Hospital, ethical approval is not necessary. This is because the patient is not involved directly in the study with interviews or any other intervention. The identity of the patient is also protected and not mentioned in the case report.

Informed Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent form is available for review by the Editor-in-Chief of this journal upon request.

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Disclosure

The authors have no conflict of interest to declare.

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