Case Report

Alleviating Bothersome Symptoms of Umbilical Endometriosis by En Bloc Resection in a Young, Fertile Patient

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Background: Abdominal wall endometriosis (AWE) is the most common extra-pelvic endometriosis encountered. Between 30% and 40% of AWE cases are umbilical endometriosis (UE), also known as Villar's nodule. This is a rare form of endometriosis, accounting for only 0.5% to 4% of extragenital seedings.

Methods: We present a case of UE in a young, fertile female treated with en-bloc resection of the nodule in accordance with the Surgical Case Report (SCARE) guidelines.

Result: A 35-year-old female presented with a persistent, painful, discolored mass in the umbilical region. An initial physical examination revealed a 2x2x2 cm discolored mass protruding from the umbilicus, accompanied by cyclical bleeding coinciding with her menstrual cycle. The mass was excised along the umbilicus, extending to the *linea alba* and peritoneum. To ensure the complete removal of all endometrial tissue from the nodule margins, a specimen was sent for a frozen section, confirming the presence of endometrial tissue and clear margins. A vertical two-lateral flap *neoumbilicoplasty* was performed. The patient exhibited no recurrent lesions, and the *neoumbilicoplasty* resulted in good cosmetic outcomes.

Conclusion: En-bloc resection and excision of the underlying peritoneum is the best choice for treating UE. A vertical umbilical reconstruction after excision offers excellent cosmetic results.

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Background

Endometriosis is a common benign gynecological disorder in which functional endometrial tissue (glands and stroma) is found outside the normal uterine cavity, and it is commonly associated with chronic pelvic pain and infertility. It affects up to 15% of women of reproductive age.[1][2][3][4][5] Although the pelvic cavity is the most frequently affected area by endometriosis, distant endometrial implants have been documented as primary or secondary lesions in nearly every organ system, including the brain.[6][7][8] Abdominal wall endometriosis (AWE) is the most common form of extra-pelvic endometriosis encountered.[3][4][5][6][9][10][11][12] AWE usually develops due to endometrial seeding at surgical sites from the exposure of raw endometrium after gynecological surgeries.[3][4][5][6][7][9][10] However, 30% - 40% of AWE cases are attributed to umbilical endometriosis (UE), also known as Villar's nodule. Villar first described it in 1889 as endometrial glands or stroma within the umbilicus. This is a rare form of endometriosis and accounts for only 0.4% to 4% of extragenital seedings. [3][4][5][6][9][10][11] We describe a case of umbilical endometriosis in a young, fertile female treated with an en-bloc nodule resection.

Case Illustration

A 35-year-old female was referred to the Division of Digestive Surgery, Department of Surgery, *Cipto Mangunkusumo* Hospital due to a persistent, growing, painful, and discolored mass in the umbilical region following surgical resection seven months prior. The first resected mass was deemed a benign tumor and did not undergo pathological examination. Shortly after the surgery, the patient had no complaints. However, one month before her presentation, she began to experience a painful sensation around her umbilical area that radiated to the right lower quadrant of the abdomen. The patient was diagnosed with appendicitis and underwent an open appendectomy. After surgery, she again felt a painful sensation around the umbilical area, accompanied by the reappearance of a discolored mass in the same region. During the physical examination at her initial presentation, we observed a solid, discolored mass measuring 2x2x2 cm protruding from the umbilicus, with no pain during palpation.

At the initial presentation, she informed us that the second umbilical mass had cyclic bleeding concurrent with her menstrual cycle (Figure 1A). The mass did not shrink in size after the bleeding, and we began to suspect umbilical endometriosis. We ordered a CT scan and scheduled elective surgery for mass resection. The abdominal CT scan results with contrast showed a solid lesion (2.1 x 2.2 x 2.2 cm)

within the cutaneous and subcutaneous tissue of the umbilical region, characterized by distinct borders, regular edges, and homogeneous enhancement with contrast. There was no infiltration into the abdominal cavity, but the lesion had infiltrated the *linea alba.*

The patient has maintained a regular menstrual cycle every 28 to 30 days since the age of 11. The cycle duration lasted 3 to 4 days, accompanied by mild to moderate bleeding. She experienced two vaginal deliveries in 2007 and 2011. Additionally, she has a history of using contraceptive injections and intrauterine devices (IUDs). The patient also reported dysmenorrhea, along with a history of dysuria and dyschezia during menstruation.

We performed a mass excision along the umbilicus, extending to the *linea alba* and peritoneum, with the patient under general anesthesia. An incision was made around the nodule until normal tissue at the edges of the nodule was observed. We excised the nodule and surrounding normal tissue, including the umbilicus (Figure 1B). To ensure the evacuation of all endometrial tissue from the nodule margins, we sent a specimen for a frozen section. A defect in the fascia and peritoneum measuring 1x1 cm was observed. We performed a vest-over-pants closure with two Mayo stitches in the linea alba to repair the defect and completed a primary peritoneal repair (Figure 1C). We conducted umbilical reconstruction using the two-lateral-flap technique (Figure 1D). The frozen section confirmed the presence of endometrial tissue and a clear margin.

After the surgical procedure, the patient was observed in post-operative care and was immediately transferred to the inpatient ward. The outcome was unremarkable, and she was sent home two days after the procedure. There were no signs of recurrence upon follow-ups, and the umbilicoplasty healed well.

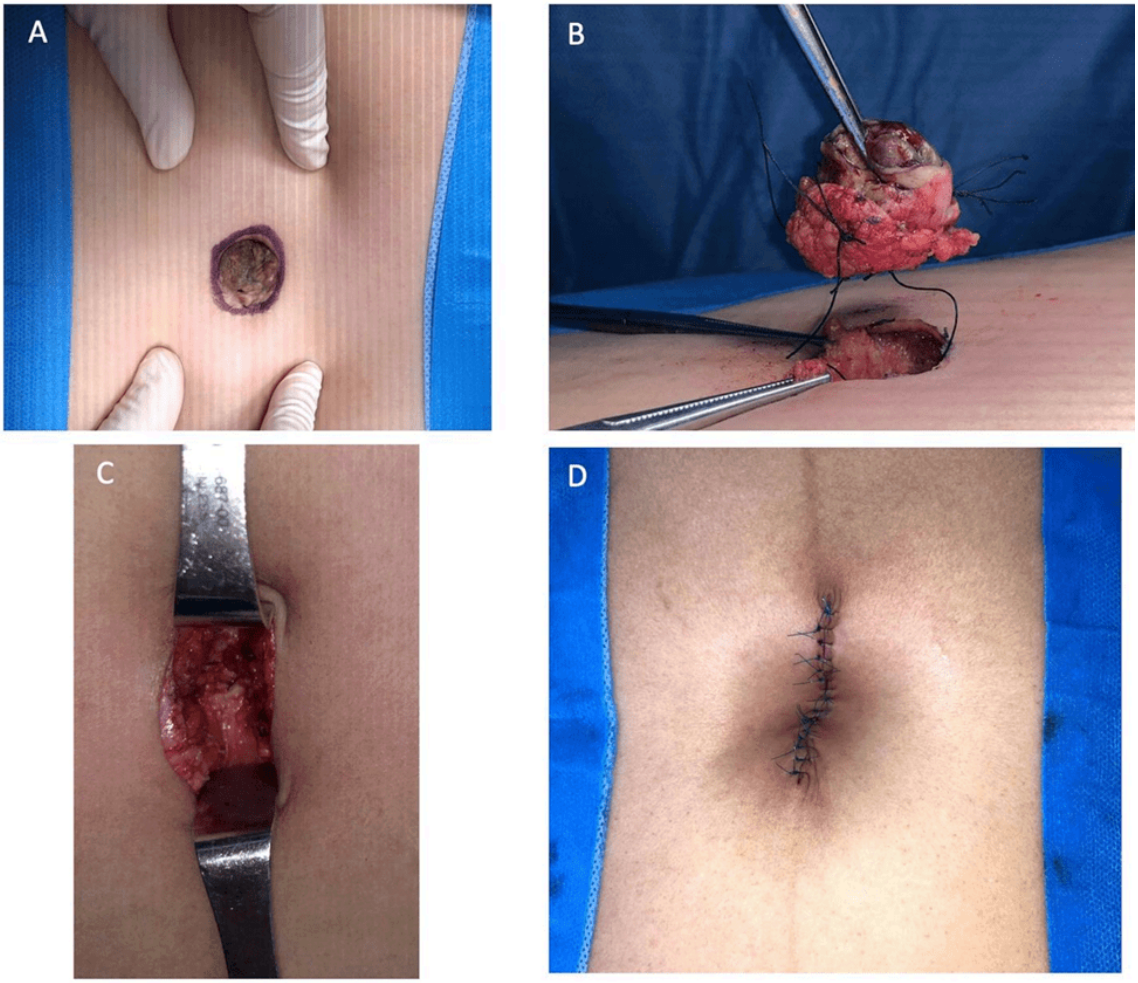

Figure 1. The clinical picture of the case. (A) Protruding discolored umbilical mass; (B) Wide excision of the mass including the underlying peritoneum; (C) Two Mayo stitches for the *Linea Alba*; (D) Two lateral flaps for umbilical reconstruction.

Discussion

Endometriosis was first described by Rokitansky in 1860 as a histopathologic presence of endometrial glands or stroma outside the normal endometrial cavity. It affects 5% to 15% of women of reproductive age. Although endometriotic implants are most frequently observed in the pelvic cavity, forming peritoneal lesions, ovarian lesions, and deep infiltrating endometriosis involving the bowel or bladder, approximately 12% to 15% of lesions are extragenital. AWE is the most prevalent extra-pelvic site, with a reported incidence of up to 3.5%. Umbilical endometriosis (UE), or Villar's nodule, accounts for 30% to

40% of all AWEs, representing only about 0.4% to 4% of extragenital lesions and up to 1% of all endometriosis cases.

UE is classified into primary or secondary UE depending on its development. While primary UE occurs without a surgical history, secondary UE arises on scar tissue following procedures such as laparoscopy or conventional laparotomy. [3][4][6][7][10][11] Primary UE is more common than the other, with almost 75% of UE cases being primary. [3][4][10][11] Retrograde menstruation, which is the outflow of the endometrial lining through patent fallopian tubes into the pelvic space, is a feature of the menstrual cycle and has been hypothesized as one of the etiologies of endometriosis. This retrograde flow, along with potential hematogenous or lymphatic circulation, may result in the seeding of endometrial tissue in ectopic sites. [1][2] Thus, the pathophysiology of primary UE has been hypothesized to result from the migration of endometrial cells through the abdominal cavity, the lymphatic system, or embryonic remnants in the umbilical fold (e.g., the urachus and umbilical vessels) [4][6][10][11] Several other theories include genetic predisposition, immunologic defects, and prolonged exposure to metaplastic and environmental factors. [1][2][4][6][7][10][11] Endometriosis may arise from Müllerian remnants that did not correctly differentiate or migrate during fetal development or from circulating blood cells that transdifferentiate into endometriosis. [1][2] While isolated UE may arise from metaplastic changes of urachal remnants, pelvic lesions coexist in 20% - 35% of patients with extragenital endometriosis. [5][6][7][10] Interestingly, most UE cases do not report a history of pelvic endometriosis. [5][6][9] Primary UE accounts for 30-40% of all cutaneous forms of endometriosis. [4][5][13] Secondary endometriosis is mainly iatrogenic due to the exposure of raw endometrial tissue to other tissues, including surgical scars. In secondary UE, presumably exposed endometrial tissue binds to the raw, fibrin-rich surface of the surgical scar around the umbilicus, especially after laparoscopic surgery. [7][9][10][11]

We cannot confirm whether our patient had primary or secondary UE, as the first umbilical mass resected was not sent for a histopathology report. However, because the previous umbilical mass excision did not penetrate the peritoneum and no endometrial implants were found on her open appendectomy scar, we suspect this is a recurrent primary UE. Additionally, a pelvic lesion in this patient was ruled out by physical examination and transvaginal ultrasound, further strengthening the hypothesis that this is a primary UE.

Studies have reported that the mean demographic age of UE is between 31 and 40 years old, which reflects premenopausal, ovarian-steroid-dependent susceptibility. It also suggests that the disease likely

occurs only after prolonged exposure to menstrual, metaplastic, or environmental factors that catalyze the development of UE.[4][6][7] Our patient is a premenopausal fertile female who falls into the mean demographic age reported by previous studies. She had all the telltale signs of UE reported in previous studies: a 2 cm intermittent painful and discolored umbilical nodule with cyclical bleeding and pain coinciding with her period (Figure 1A). UE usually presents as a loculated cystic mass described as an umbilical endometrioma. Previous studies have reported the average diameter of UE to be between 0.5 and 3.5 cm, and most reported skin color changes around the umbilicus are accompanied by swelling and bleeding, exacerbated during menstruation. Studies have shown that the most common symptoms of UE are intermittent pain in the umbilical area (66%) and cyclical bleeding (43%).

The management of endometriosis involves a multidisciplinary approach that includes surgical excision or debulking, hormonal treatment to suppress and delay the recurrence and progression of the disease, pain management, and pelvic therapy[1][2][7]. The suggested primary treatment for UE is radical local excision[3][4][5][6][9][10][11]. Symptomatic endometriosis is typically managed with surgical or medical treatment, which are equally effective. However, medical treatment is weakly recommended for UE due to limited supporting data and a lack of studies comparing medical and surgical treatments for UE[1][3][10]. Studies advocate for wide excision as the primary treatment of UE to suppress recurrence and reduce the risk of malignancy. Approximately 3% of UE cases are reported to undergo malignant transformation[3][6][7]. In a systematic review, Dridi et al. reported a very low post-operative recurrence rate after en-bloc resection, suggesting surgery is an effective treatment for UE[10] . Additionally, in a national survey article, Hirata et al. described no recurrences in UE patients who underwent wide peritoneal and mass excision[3]. These findings further reinforce the recommendation of wide excision, including the peritoneum, as the first choice of therapy for UE[3][5][7]. We performed en-bloc resection of the nodule and the underlying peritoneum with an intraoperative frozen section to confirm the diagnosis and ensure a negative resection margin. The umbilicus, fascia, and peritoneum were excised beneath (Figure 1B). The defect in *the linea alba* was closed with two craniocaudal mayo stitches (Figure 1C).

An en-bloc resection of UE results in a cosmetic issue where the abdomen lacks an umbilicus, a specific feature of UE excision[3][4] Several surgical techniques are available for umbilicus reconstruction or creating a new umbilicus (*neoumbilicoplasty*), but there is no specific algorithm for choosing and applying any of these techniques[14] The choice of umbilicus reconstruction technique rests with the surgeon based on each case. Achieving a perfect result in *neoumbilicoplasty* is almost impossible[4][14] No

absolute standards exist to define an aesthetically pleasing umbilicus; however, a vertically oriented umbilicus with superior hooding tends to be more attractive than a horizontal one[14] We opted for two lateral pedicled flaps, where the two lateral skin flaps are sutured with 4.0 nonabsorbable monofilament sutures to the abdominal fascia and to each other (Figure 1D).

Our patient showed no signs of recurrence upon follow-up one month after the surgery; she received the good news of her pregnancy. The *neoumbilicoplasty* healed well, leaving a vertical dimple in place of the umbilicus, which created an aesthetically pleasing appearance. Based on our results and supported by previous studies, we conclude that en-bloc resection and excision of the underlying peritoneum is the best treatment choice for UE. A vertical umbilical reconstruction after excision provides excellent cosmetic outcomes.

Conclusion

Umbilical endometriosis is a clinical diagnosis made based on a patient's medical history and physical examination. It should be considered in fertile female patients who present with a discolored umbilical mass, especially when cyclical swelling and bleeding accompany their menstruation. The optimal treatment choice for UE is wide excision, encompassing the underlying fascia and peritoneum to prevent recurrence and malignant transformation. Umbilical reconstruction can be performed for *neoumbilicoplasty* following the wide excision of UE to achieve a better cosmetic result.

Statements and Declarations

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

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Declarations

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