

A surgical case report on primary umbilical endometriosis (Villar's nodule)

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Abstract

Endometriosis is a disease characterized by the implantation of endometrial-like tissue outside the uterine cavity. Common symptoms include cyclical pain, dysmenorrhea, dyspareunia, and infertility. Although endometriosis can spread to various extrauterine locations including the ovaries, fallopian tubes, and peritoneal surfaces, umbilical endometriosis is a rare manifestation of the disease. We report an intriguing clinical case of primary umbilical endometriosis in a 36-year-old female patient admitted to our department due to the notable manifestation of a painful swelling at the umbilicus, accompanied by cyclic episodes of bleeding. Subsequent investigations, incorporating ultrasound and computed tomography, indicated the presence of umbilical endometriosis, a finding that was subsequently confirmed by pathological examination of a mass biopsy. Surgical resection of the umbilical mass was performed, and histopathological analysis definitively confirmed the diagnosis of endometriosis. This case report aims to discuss in depth the diagnosis and management of umbilical endometriosis.

Keywords

Primary umbilical endometriosis, umbilical endometriosis, Villar's nodule, umbilical bleeding, cyclic umbilical swelling, endometriosis

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Introduction

Endometriosis is a chronic pathological condition in the field of gynecology, distinguished by the ectopic presence of endometrial-like tissue with its glandular and stromal components. According to World Health Organization estimates, around 10% or 190 million women of childbearing age worldwide suffer from this condition.¹

Endometriosis can be classified into two types. Primary endometriosis refers to the existence of ectopic endometrial-like tissue outside the uterus, without any prior surgical intervention in that particular region. Secondary endometriosis may result from open or laparoscopic surgery. The most common symptoms include abdominopelvic pain, dysmenorrhea, heavy menstrual bleeding, and infertility, although 20%–25% of women with the condition are asymptomatic.²

Endometriosis mainly affects the pelvic organs, but around 12% of endometriosis cases occur in sites outside the reproductive organs, such as the lungs, diaphragm, or umbilicus.³ Umbilical endometriosis, a rare variant of the disease, was first reported in 1886.² Its incidence is estimated at

0.5%–1% of women with extragenital endometriosis, including secondary and spontaneous primary forms.⁴

This report aims to present a clinical case of primary umbilical endometriosis and to discuss appropriate management approaches for this medical condition.

Case presentation

In this case report, we present the unusual clinical history of a 36-year-old female, primiparous, who presented to our department. The patient's primary complaint was the presence of persistent painful umbilical swelling lasting for

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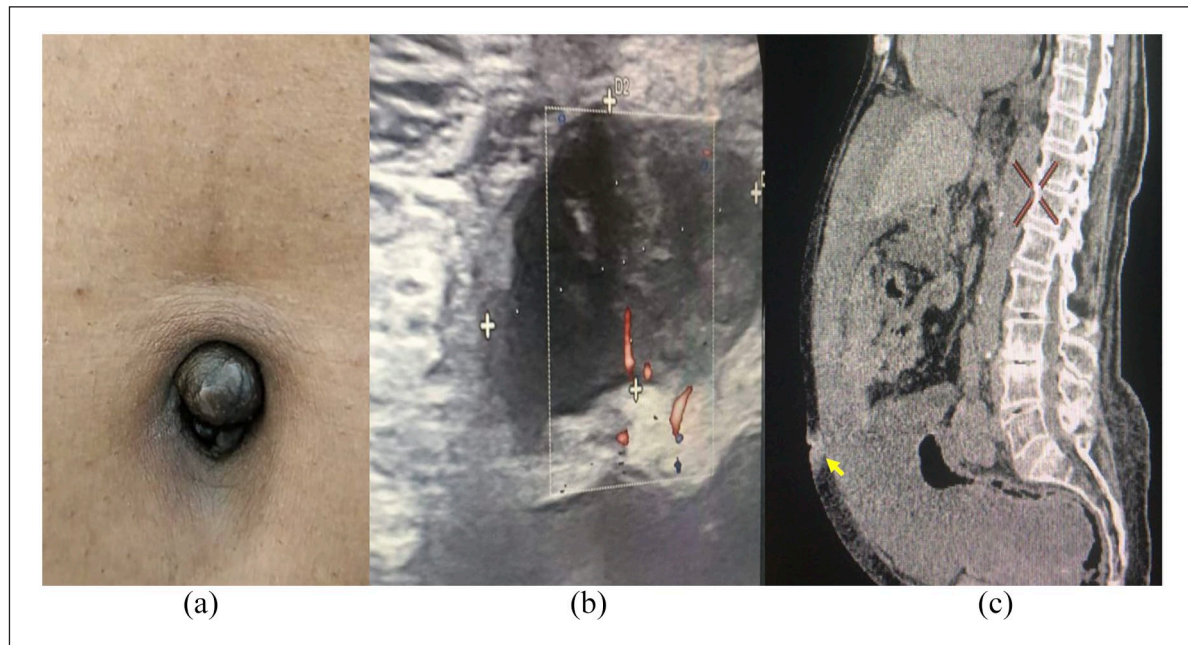


Figure 1. (a) Endometriosis presenting as an umbilical mass characterized by swelling and discoloration. (b) Abdominal ultrasound showing a non-vascularized subcutaneous mass with a tissue-like appearance at the umbilicus, measuring $24\text{ mm} \times 19\text{ mm}$. (c) The computed tomography examination reveals the presence of a tissue-based umbilical nodule that does not extend beyond the muscle layer.

2 months. Furthermore, she reported a unique symptom of spontaneous catamenial bleeding originating from the umbilicus during her menstrual period. She reported regular menstrual periods. She had no history of abdominal or uterine surgery or umbilical trauma and had given birth spontaneously by vaginal delivery. The patient did not have a documented history of endometriosis and did not utilize oral contraception. On physical examination, we found a 1.5 cm solid black umbilical swelling that was painful and irreducible on palpation, discharging a bloody fluid (Figure 1(a)). There was no sign of infection.

Abdominal ultrasound revealed a tissue-like, non-vascularized subcutaneous mass at the umbilicus, measuring $24\text{ mm} \times 19\text{ mm}$ (Figure 1(b)). Given the unavailability of magnetic resonance imaging, a computed tomography scan of the abdominopelvic region was performed with contrast injection, revealing a small umbilical tissue nodule measuring $23\text{ mm} \times 19\text{ mm}$, which did not protrude beyond the muscle plane (Figure 1(c)).

Due to the cyclical nature of umbilical pain and bleeding, which correlated with the patient's menstrual cycle, umbilical endometriosis was suspected. The initial differential diagnosis included a simple inclusion cyst, benign/malignant neoplasms of the umbilicus, and umbilical granuloma in consideration. However, following a biopsy of the lesion, the diagnosis of umbilical endometriosis was confirmed, ruling out the possibility of malignancy.

Following a meticulous exploratory laparoscopy aimed at thoroughly examining the abdominopelvic cavity and

identifying any concurrent pelvic endometriotic lesions, no suspicious lesions, whether endometriotic or otherwise, were observed. Consequently, a decision was made to proceed with surgical excision.

During the surgical procedure, an omphalectomy was performed, wherein the entire umbilicus, including the nodule, fascia, and peritoneum, was excised as a single unit. The fascia was sutured to the peritoneum, ensuring proper attachment. The periumbilical skin was then carefully reconnected to the fascia. To complete the procedure, interrupted absorbable sutures were used to close the skin.

The patient had a favorable postoperative course and was discharged on the second day following surgery. The diagnosis of umbilical endometriosis was confirmed through histological examination (Figure 2).

During the follow-up appointments conducted at 6 months and 1 year post-surgery, the patient reported the absence of umbilical pain or discharge.

Discussion

Endometriosis outside the pelvic region is rare, particularly in the umbilicus, which accounts for around 0.5%–1% of all cases of extra-genital endometriosis. Cutaneous endometriosis can sometimes develop in scars, such as those resulting from cesarean sections or episiotomies.⁵

Umbilical endometriosis typically arises as a result of surgical incisions following open or laparoscopic abdominal procedures. However, in rare instances, it can develop

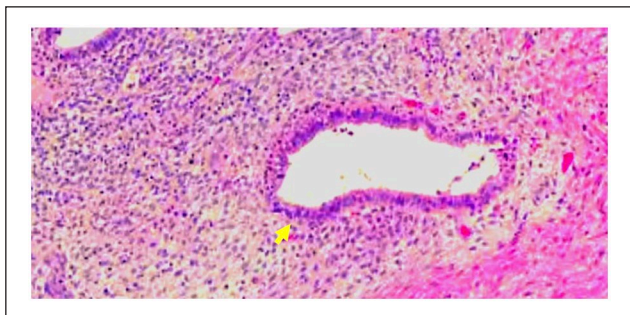


Figure 2. Histological image showing the endometrial gland (yellow arrow) surrounded by cytogenic stroma (endometriosis); hematoxylin–eosin staining, increase $\times 100$.

as primary umbilical endometriosis without any previous surgical intervention.⁶ Our case report presents a rare situation in the medical literature, that of primary umbilical endometriosis that develops without prior surgical intervention. Unlike typical cases involving surgical incisions, our case is exceptional in that it is confined to the umbilical region and manifests as the infrequent symptom of cyclical umbilical bleeding.

One of the hypotheses put forward as to the cause of primary umbilical endometriosis is the migration of endometrial cells via the lymphatic and vascular system toward the umbilicus.⁷

The clinical presentation of umbilical endometriosis is characterized by the emergence of a colored, raised, or protuberant lesion in the umbilical depression, with symptoms that can fluctuate based on the menstrual cycle. This lesion may be accompanied by cyclical pain and intermittent bleeding from the umbilicus.⁸

A systematic review demonstrated that 73.1% of patients diagnosed with umbilical endometriosis were not previously identified as having endometriosis and that pain was the most prevalent symptom in these patients, affecting 77.93%, followed by hemorrhage and edema, corresponds to the clinical presentation observed in our patient.⁹

Ultrasound, computed tomography, and magnetic resonance imaging are essential in the preoperative evaluation of tumor size, extent of abdominal involvement, and identification of additional intraperitoneal lesions that may indicate endometriosis.^{10,11}

In the presence of cyclical umbilical bleeding and/or pain, along with umbilical swelling, healthcare providers should consider the potential diagnosis of umbilical endometriosis. Nevertheless, it is crucial to consider several other differential diagnoses, including secondary metastatic nodules (known as Sister Marie-Joseph's sign),¹⁰ melanocytic nevi, umbilical polyps, umbilical hernias, and lipomas.¹¹

Instances of malignant transformation of the umbilical nodule have been recorded, with an approximate transformation risk estimated to be less than 5%. These cases include endometrioid adenocarcinomas and clear-cell adenocarcinomas.¹²

The initial diagnosis of umbilical endometriosis relies primarily on clinical criteria, but confirmation is achieved through histopathological analysis. Microscopic examination revealing the presence of endometrial stroma and glands serves as the cornerstone for definitive diagnosis.^{5,12}

In our case, the diagnosis of umbilical endometriosis was suspected based on the clinical presentation and imaging findings and confirmed by pathological analysis.

Surgical management is mainly based on an extensive resection that includes the peritoneum, with the possibility of umbilical reconstruction, taking great care to pass through a healthy zone to minimize the risk of recurrence. In addition, it is recommended to perform a pelvic laparoscopic exploration to investigate for potential additional sites of endometriosis, whenever feasible, to complement the surgical procedure.⁹

Different treatment options are available for the management of endometriosis. These include the use of oral contraceptives, progestins, and gonadotropin-releasing hormone antagonists or agonists. However, it should be noted that the evidence regarding the efficacy of these treatments is limited.⁵ In addition, non-radical treatment modalities, such as thermocoagulation, are generally discouraged due to the high probability of disease relapse. In our specific case, the patient expressed a preference for surgery over medical treatment.¹¹

The risk of recurrence of umbilical endometriosis after surgical resection varies from 5.4% to 27%.¹¹ However, encouraging results have been observed with extended resection including the peritoneum and umbilical reconstruction, considerably reducing the risk of disease recurrence.¹³

Conclusion

Primary umbilical endometriosis is a rare entity that requires special attention in the differential diagnosis of umbilical lesions. The clinical diagnosis of umbilical endometriosis can be challenging, as it can mimic other benign or malignant conditions affecting the umbilicus. Surgical excision continues to be the preferred treatment option, providing favorable long-term outcomes. However, due to the rarity of this condition, an individualized approach to treatment is required, taking into account the specific characteristics of each patient. Careful monitoring and anticipation of recurrence are recommended.

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Author contributions

A.B. wrote the article, designed the project, analyzed the data, and contributed to the discussion of the article; A.B., Y.B., S.B., and J.K. treated the patient, gathered the patient's information, and supervised the project and research. All authors have read and approved the article. All the authors have accepted responsibility for the entire content of this submitted article and approved the submission.

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Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

Informed consent

Written informed consent was obtained from the patient for their anonymized information to be published in this article.

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