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Endometriosis beyond the Pelvis: A Case Series of Cutaneous Endometriosis and Literature Review

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Abstract

Introduction: Cutaneous endometriosis is an uncommon but well-known skin disorder that represents about 0.5% to 1% of all endometriosis. The objective of this case series is to report clinical presentation, diagnosis, and management of various forms of cutaneous endometriosis. **Material and Methods:** It was an observational, retrospective and descriptive review of cases presenting with cutaneous endometriosis among Cameroonian women managed at the gynaecological outpatient department of Yaounde Gynaeco-Obstetric and Pediatric Hospital. All the following parameters were analysed: age, parity, previous pelvic surgery, presenting symptoms and duration, associated symptoms, localizations, imaging, size of the lesion, other localization of endometriosis, management and histopathological results. **Results:** we reported 4 cases of cutaneous endometriosis, with 3 umbilical endometriosis and 1 abdominal scar endometriosis. Patient age ranged from 28 to 39 years with an average of 33 years. All patients described infertility (two primary and two secondary) and two had a history of abdominal surgery. All patients presented local cyclical signs such as pain, swelling, color change and bleeding. The duration of symptoms varied from 2 to 3 years and the size of lesions ranged from 2 to 3.5 cm for umbilical lesions and was 9 cm for abdominal scar endometriosis. In all cases, no imaging was required for the diagnosis, which was suspected on the basis of patient's history and the cyclical nature of local signs, followed by wide surgical excision and confirmation on histo-

pathology. **Conclusion:** Cutaneaous endometriosis is a rare benign condition. Umbilical endometriosis seems to be the main cutaneous localization and can be described as primary or secondary. Even if its diagnosis must be confirmed by histopathology, it should be considered in patient with cutaneous cyclic signs such as pain, swelling or bleeding with or without history of abdominal surgery.

Keywords

Endometriosis, Cutaneous Endometriosis, Umbilical Endometriosis, Cyclic Umbilical Swelling, Cameroon

1. Introduction

Endometriosis is a common benign gynecological condition that is characterized by the presence of functioning endometrial tissue outside the uterine cavity. It affects approximately 20% of women hospitalized for pelvic pain and 50% of infertile women [1]. It is not only a disease of the pelvis, thus it can be divided into pelvic and extrapelvic endometriosis. Pelvic endometriosis sites are commonly seen on ovaries, fallopian tubes, uterosacral ligaments, and pelvic walls [2]. Extrapelvic sites include various organs such as the liver, diaphragm, brain, lung, anterior abdominal wall, and skin. Cutaneous endometriosis is uncommon but well-recognized skin disorder that represents about 0.5% to 1% of all endometriosis [3]. Umbilical endometriosis, one of the most seen cutaneous endometriosis, was first described by Villar in 1886 [4]. Depending on the patient's surgical history, it can be classified as primary or secondary. Primary umbilical endometriosis also known as Villar's nodule, occurs spontaneously in patients with no known history of endometriosis nor signs and symptoms related to endometriosis. Its etiopathogenesis is still unclear [5]. Secondary endometriosis mostly occurs after abdominal or pelvic surgery, due to the seeding of pre-existing lesions [6].

Since this pathology can mimic the clinical features of many skin disorders, it can be considered as a diagnostic trap [7], difficult to diagnose even if surgical excision with wide margins is mostly used as the treatment of choice [8]. The majority of publications on cutaneous endometriosis are case reports or case series. The purpose of this report is to present four cases of cutaneous endometriosis managed in Yaounde Gyneco-Obstetric and Pediatric Hospital (YGOPH) in Cameroon.

2. Material and Methods

We conducted an observational, retrospective and descriptive review of cases of cutaneaous endometriosis. We selected patients who came to us with cutaneous lesions, and whose skin excision specimen showed cutaneous endometriosis on anatomopathological analysis, identified from the records of Histopathology

Department database of our hospital between 2018 and 2023. Then, using their names, we searched for their files in the archives, and from their medical records, we extracted and analyzed the following variables, age, parity, history of abdominal surgery, presenting symptoms and duration, associated symptoms, localizations, imaging, size of the lesion, other localization of endometriosis, management, and histopathological results.

The Ethics Committee board of our Hospital approved the study and written informed consent was obtained from all patients for publication of this paper with accompanying image.

3. Results

We recorded a series of 4 cases during the period of study. The cases are presented as follows.

Case 1:

A 28-year-old Cameroonian woman presented to our department with a two-year history of monthly bleeding from the umbilicus associated with tender swelling. Her medical history revealed a 4-year primary infertility with a polymyomatous uterus, deep dyspareunia, and chronic pelvic pain for 4 years. She underwent both myomectomy by laparotomy and laparoscopy for tubal assessment three weeks later. During the laparotomy, peritoneum and uterosacral ligaments were involved with endometriotic lesions which were excised according to the postoperative report. One year after this surgery, she noticed the progressive appearance of an umbilical mass with local pain and bleeding every time she had her period. On clinical examination, the patient was in good general condition. There was a 3 × 3.5 cm dark-colored soft-firm nodular umbilical tumor surrounded by dried blood (Figure 1). This nodule was not reducible by gentle digital pressure, non-pulsating, and not expansive to coughing. Abdominopelvic Doppler ultrasound scan showed a 3.5 cm, well-vascularized hypoechoic mass around the umbilical region. She underwent an excision of the umbilical nodule

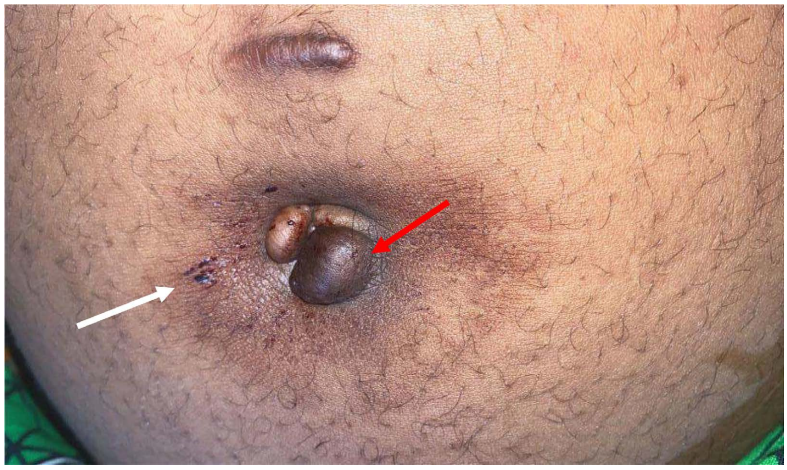

Figure 1. Umbilical nodule slightly pigmented during period (red arrow); dried blood from the nodule (white arrow).

(Figure 2) with umbilicoplasty. The postoperative period was uneventful, and she was discharged one day after the surgery.

At microscopy (Figures 3(a)-(c)), there was acanthosis scattered over the epidermis. The tumor was located in a vitreous stroma with numerous vessels and endometrial glands. This stroma harbored a minimal lymphocytic inflammatory infiltrate of predominantly perivascular topography.

There was no evidence of cutaneous metastasis. The diagnosis of secondary umbilical endometriosis was made on the basis of her history of known pelvic endometriosis. After two weeks, the healing of the skin was well, without signs of local recurrence six months after the excision.

Case 2:

A 35-years-old woman, with 4 years history of chronic pelvic pain, presented

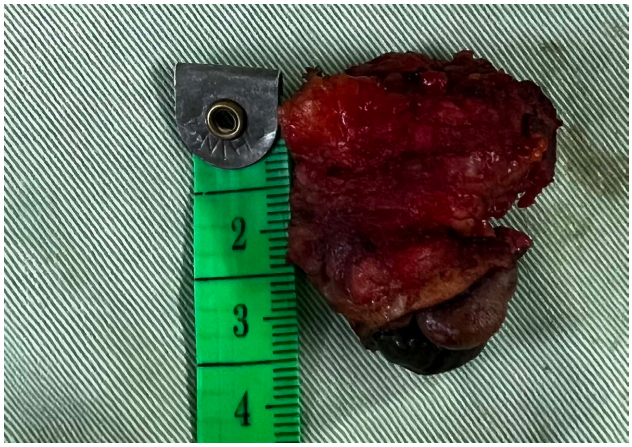

Figure 2. Omphalectomy piece.

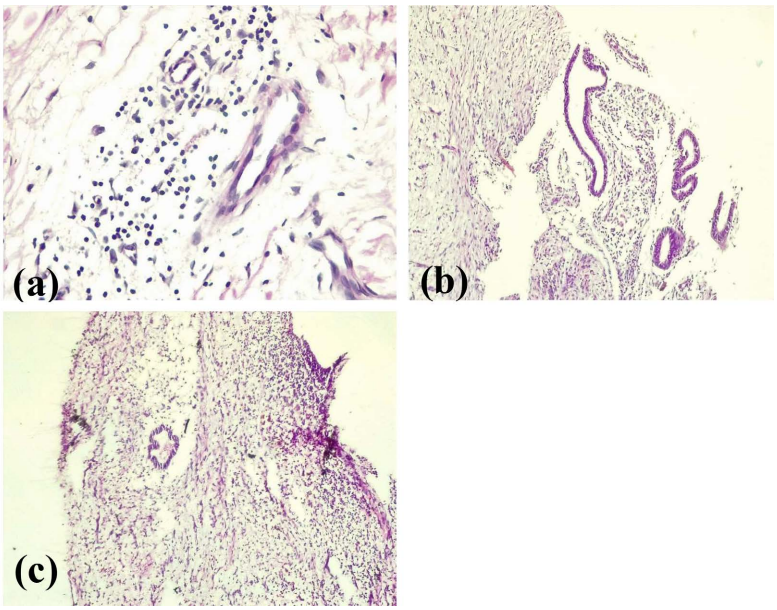

Figure 3. H & E staining, 4× magnificence. (a)-(c) Specimen showed acanthosis scattered over the epidermis. The stroma was vitreous with numerous vessels, endometrial gland and lymphocytic inflammatory infiltrate of predominantly peri vascular topography.

with a markedly tender swelling of a sub-umbilical midline laparotomy scar with cyclic local bleeding. A year ago, she underwent an emergency cesarian section for hemorrhagic placenta previa at 32 weeks gestational age. Unfortunately, the baby did not survive and she was having difficulty conceiving again. Six months after the surgery, the patient noticed bleeding and thickening of the scar, which progressively widened and became increasingly painful, especially during her periods (**Figure 4**). The diagnosis of abdominal scar endometriosis was suspected based on the history and physical examination. We performed a full thickness wide excision of the scar (**Figure 5**) and histological analysis confirmed the diagnosis (**Figure 6**).

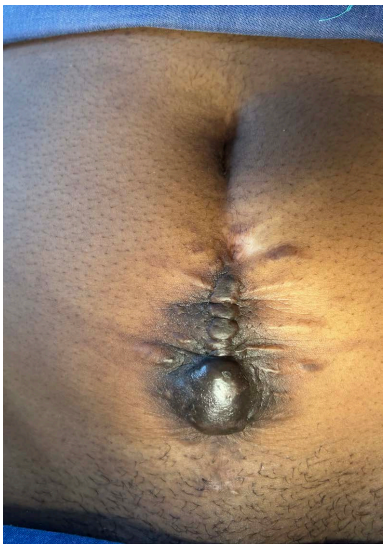

Figure 4. Sub-umbilical midline thickening of the scar.

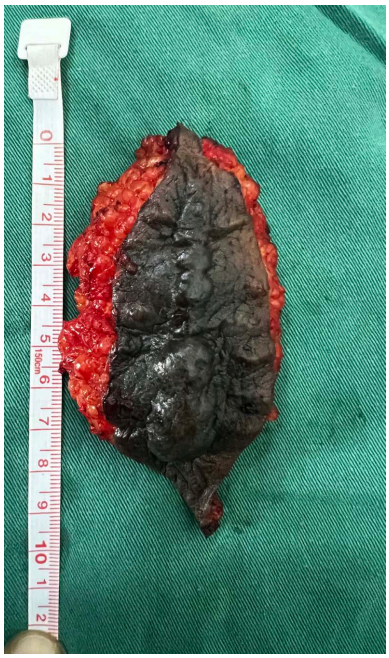

Figure 5. Sub-umbilical midline thickening of the scar.

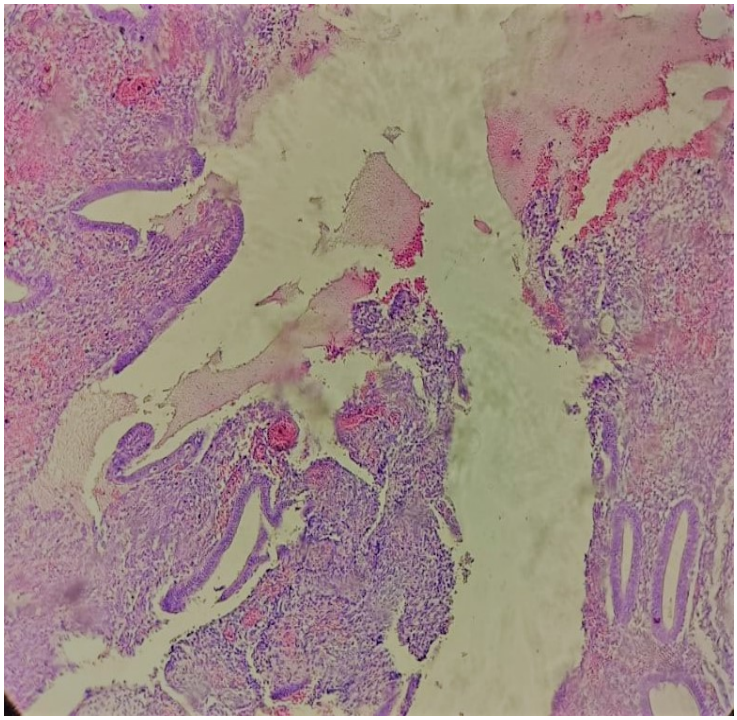

Figure 6. H & E staining, 4× magnificence. Specimen showed Endometrial glands and stroma, fat and fibrotic tissue.

Case 3:

A 31-year-old woman, with unremarkable medical condition, presented with a painful, black-colored mass in her umbilicus that has gradually developed over the past 3 years. During her menses, she had periodic umbilical pain and bleeding associated with local swelling.

Seven years ago, she had one previous normal vaginal delivery. She never experienced chronic pelvic pain and never had any abdominal or pelvic surgeries. She has a history of heavy menses. Clinical examination revealeda painful, well-defined dark coloured soft-firm nodule, 3 × 2 cm in size in the umbilical region with bleeding (Figure 7). The uterus was midly enlarged like 12 weeks with irregular surface. Abdominopelvic ultrasound showed a 2.5 × 2 × 1.5 cm heterogenous hypoechoic lesion at the umbilicus with posterior shadowing, suggestive of umbilical endometriosis due to cyclic local signs. Ultrasound also found two subserosal myomas, one measuring 9 cm and the other 4 cm, located respectively in the posterior part of the body of the uterus and at the level of uterine fundus. Unfortunately, the ultrasound images are not available.

The patient underwent wide local excision with about 1 cm of free margins with umbilicoplasty (Figure 8). During laparoscopic that was also offered to the patient for removal of her 2 myomas, no signs of endometriosic lesions was described. Histopathological exam revealed endometrial gland and stroma, confirming the diagnosis of primary umbilical endometriosis (Figure 9). The postoperative period was unremarkable with good wound healing, and disappearance of its initial symptoms.

Figure 7. Dark coloured umbilical mass with bleeding.

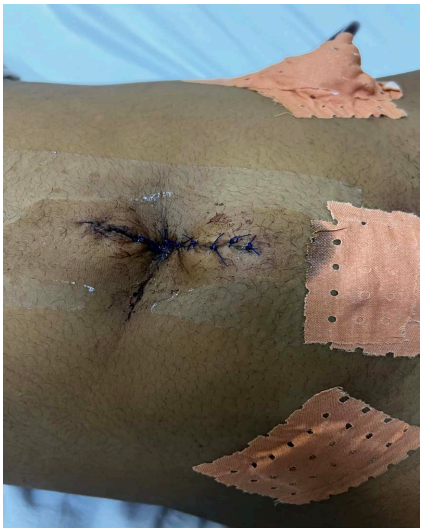

Figure 8. Umbilicoplasty according to Kokuba.

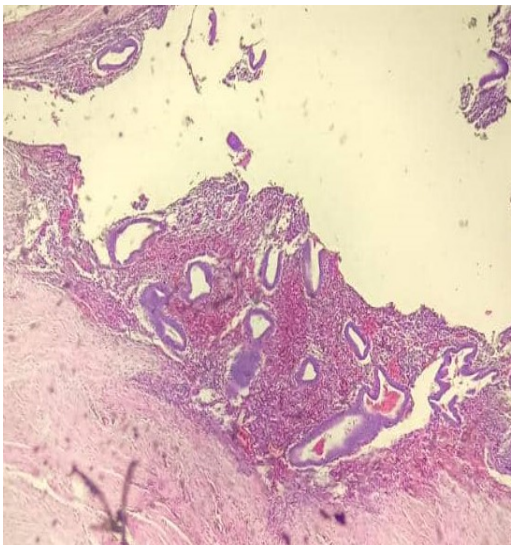

Figure 9. H & E staining, 4× magnificence. Endometrial glands and stroma.

Case 4:

A 39-year-old woman came to the gynecology outpatient department with chief complaint of a painful, palpable, dark coloured mass of the umbilical area (Figure 10) since three years ago. The mass was associated with pain that had a cyclic characteristic, with local swelling and bleeding during the menstrual period. The patient has no previous abdominopelvic surgery, but she described 6-year primary infertility, and cyclic chronic pelvic pain and deep dyspareunia.

On clinical examination, a well-defined dark coloured nodule was palpated, measuring 1 × 2 cm with woody consistency. A biopsy of the lesion was taken and the specimen sent for histological examination. We did not find the histopathological image, but the pathologists' report showed the presence of endometrial tissue in favour of umbilical endometriosis. The patient underwent local surgical excision with concomitant diagnostic laparoscopy, which revealed endometriosis of both uterosacral ligaments (Figure 11), which were excised.

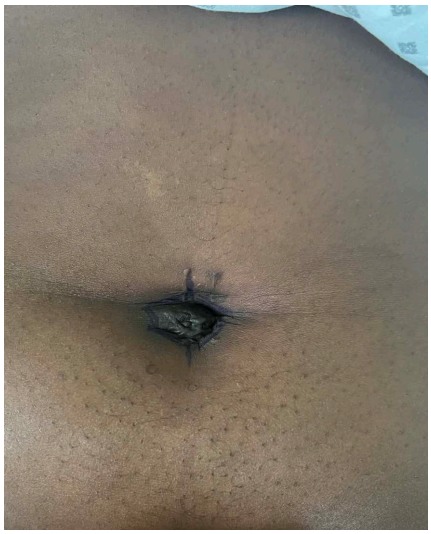

Figure 10. Dark coloured umbilical nodule.

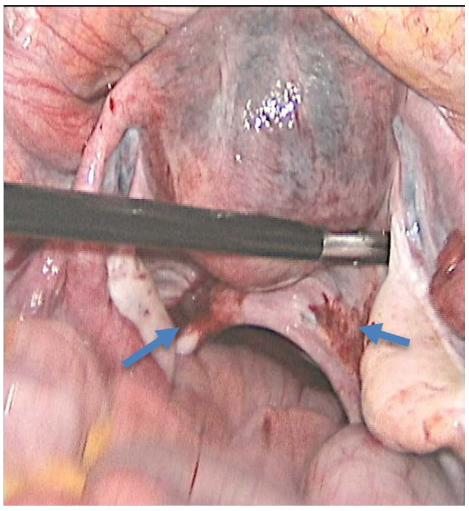

Figure 11. Laparoscopic view of endometriosis of both uterosacral ligaments (blue arrow).

According to these findings, the diagnostic of secondary umbilical endometriosis was retained. Postoperative period was uneventfull and the patient was followed up for 6 month without recurrence.

4. Discussion

Cutaneous endometriosis is a rare condition that occurs when endometrial tissue, which normally lines the uterus, grows outside the uterus and implants into the skin. Although there are large series, most of the cases are published as isolated cases, making it difficult to estimate its true prevalence. According to Victory *et al.* [4], this localization represents less than 1% of all cases, while other studies [9] [10] [11] estimate this prevalence at less than 5.5%. Classically, it affects women of reproductive age. In our series, patient's age ranged from 28 to 39.

Cutaneous endometriosis can be classified as primary if it occurs spontaneously without any history of a local surgery, or as secondary also called scar endometriosis if associated with prior abdominal or pelvic surgery [4] [12]. The less common of the two is primary form with 30% of cases [4]. Two of our patients (case 1 and case 2) had a history of pelvic surgery and therefore had secondary cutaneous endometriosis, one localized in the umbilical level and the other at the level of the median subumbilical scar. If the pathogenesis of secondary cutaneous endometriosis can easily be conceptualized by iatrogenic seeding of endometrial cells into the surgical site and peri-incisional tissue during surgery, the pathogenesis of primary cutaneous endometriosis remains unclear. The most common localization of primary cutaneous endometriosis is the umbilicus, which comprises 30% to 40% of all cutaneous endometriosis [4] [13]. It was first, described in 1886 by a physician named Villar and it is why this is also known as Villar's nodule [4] [12] [13]. Three of our patients (Case 1, Case 3, and Case 4) had a Villar's nodule with particularity of Case 1 who had also pelvic endometriosis without incision over the umbilicus. Other hypotheses, including metaplasia, venous and lymphatic metastasis, have been put forward to explain this pathogenesis [10]. Due to the fact that the umbilicus acts as a physiological scar, some authors have postulated that primary and secondary umbilical endometriosis have similar pathogenesis [4].

Clinically, the manifestation of cutaneous endometriosis presents as a painful nodule of variable color and aspect fluctuating with the hormonal environment and is associated sometimes with catamenial bleeding. According to one systematic review, pain was the leading symptom, found in 77.93% of patients, along with bleeding and swelling [4]. These findings are consistent with our series, where the diagnosis of cutaneous endometriosis was made exclusively on the basis of the cyclical characteristics of the symptoms, confirmed by anatomopathological examinations. However, in up to 80% of cases, patients are asymptomatic [7] [14] [15] [16]. The differential diagnosis of umbilical endometriosis is very wide, including umbilical lesions like hernia, Sister Mary Joseph nodule, keloid,

melanoma, melanocytic nevus, urachal duct cyst, pyogenic granuloma dermatofibroma and dermatosarcoma protuberans [5] [11] [14] [15]. These differential diagnoses differ from umbilical endometriosis in that they do not have cyclical manifestations coinciding with menstruation, as is the case with umbilical endometriosis, and a biopsy is needed to assess the diagnosis.

The size of the umbilical lesion in our series (2 - 3.5 cm) coincides with that of the literature, which is usually 2 - 3 cm [14] [17] [18], but it can enlarge to even more enormous sizes [19]. Imaging studies such as ultrasonography with or without Doppler or CT scan or Magnetic resonance imaging are also helpful for the diagnosis and to precise the size and extension of lesion before the surgery. however, none of our patients benefited from MRI due to the non availability of this examination and its financial inaccessibility.

However, diagnostic confirmation is always based on the histology of the surgical specimen, which is the standard diagnostic test [4] [11] [14] [15] [16]. The histological findings are characterized by irregular glandular cavities embedded in a stroma with a high cellular and vascular content, similar to the stroma of a functional endometrium.

Wide margins excision of the lesion is the treatment of choice [10] [11] [14] [15] [16] [20]. It was realized in all of our patients. Omphalectomy was performed in all 3 cases of umbilical endometriosis, and omphaloplasty was realized according to the technique described by Kokuba *et al.* [21] who used 2 semicircular defatted flaps. This technique is efficient in creating a new umbilicus with a natural appearance while leaving a minimal scar [21]. Simultaneous laparoscopic surgery has been performed in one of our patients for pelvic symptomatic endometriosis and infertility, but the routine exploratory laparoscopy is debatable [14].

In our series, patients were informed of the possibility of non-surgical treatment, but all underwent surgical treatment. The non-surgical approach includes the use of hormonal agents alone, such as gonadotropin-releasing hormone agonists, oral contraceptives, and danazol. They can help improve symptoms and reduce the size of lesions for subsequent surgery [22]. Patients must be counseled that the lesion may recur when they discontinue their hormonal treatment. Generally, the prognosis of cutaneous endometriosis is favorable, with a low risk of recurrence [13] and a very low risk of malignant transformation with only 2 described cases [23] [24].

5. Conclusion

Cutaneous endometriosis is a rare benign condition. Umbilical endometriosis seems to be the main cutaneous localization and can be described as primary or secondary. Even if its diagnosis must be confirmed by histopathology, it should be considered in patients with cutaneous cyclic signs such as pain, swelling, or bleeding with or without a history of abdominal surgery. Surgery is the cornerstone of treatment with a low risk of recurrence and malignant transformation.

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Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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