
Symptomatic Primary Umbilical Endometriosis: A Case Report and A Review of the Literature

BACHAR Amine¹, JAMALEDDINE Khalid², MASTAR Hajar³, ESSAIDI Zakaria⁴, ELABBASSI Taoufik⁵, LEFRIYEKH Mohamed Rachid⁶

^{1,2,3,4,5,6}Department of general surgery, IBN ROCHD University hospital of Casablanca, Casablanca, Morocco

ABSTRACT: Endometriosis is a condition defined as extrauterine functional endometrial tissue, typically seen on pelvic peritoneal surfaces, leading to symptoms such as cyclic pain, dysmenorrhea, dyspareunia, and infertility. Extra-pelvic endometriosis is uncommon, especially the umbilical form. We present an interesting case of primary umbilical endometriosis (Villar's nodule) in a patient with no medical or surgical history, who presented with umbilical pain and bleeding. She was found to have a bluish umbilical nodule, which was surgically removed and sent to the pathology to confirm the diagnosis. The report aims to highlight the diagnosis and management of umbilical endometriosis and the possibility of its occurrence even in patients without prior surgical history.

KEY WORDS: Primary Umbilical endometriosis – Umbilical bleeding – Cyclic umbilical swelling – Surgical excision

INTRODUCTION

Endometriosis (EM) is a benign gynecological disorder [1] characterized by the presence of functional endometrial tissue outside the uterus. It frequently affects the pelvic peritoneum, ovaries, or rectovaginal septum. Symptoms vary based on the affected organs and inflammation level but typically include dysmenorrhea and dyspareunia, with about 20–25% of cases being asymptomatic. This chronic condition impacts 6–15% of women of reproductive age and 6% of postmenopausal women, with infertility occurring in 30–50% of affected individuals [2].

Endometriosis in extra-pelvic sites is rare, with less common locations including the diaphragm, lungs, and anterior abdominal wall [3]. Among patients with endometriosis, 0.5–1% develop umbilical endometriosis, which can be primary or secondary. Secondary umbilical endometriosis arises post-laparoscopic procedures, while primary umbilical endometriosis (PUE), first identified by Villar in 1886, is particularly rare [2].

Here, we present the case of a 36-year-old nulliparous woman with primary umbilical endometriosis.

PRESENTATION OF CASE

A 36-year-old nulliparous woman with no comorbidities or history of abdominal or pelvic surgeries presented to the emergency room with dysmenorrhea and a sharp, throbbing pain originating from a swelling, bleeding, bluish nodule in the umbilicus (figure 1). The pain began five days prior to admission, and the bleeding started on the day of admission. Her last menstrual period began four days before admission, and she reported that this condition had been recurring for four years, with symptoms worsening during menstruation. She did not take oral contraceptive pills and found that analgesics only provided temporary relief. The patient denied other symptoms such as umbilical/abdominal trauma, fever, chills, vomiting, diarrhea, nor constipation.

Upon admission, her vital signs were within normal limits. Clinical examination revealed multiple ill-defined bluish papules surrounded by erythematous skin in the inferior aspect of the umbilicus. A gynecological evaluation was normal. Laboratory tests showed a hemoglobin level of 11.2 g/dL, a platelet count of 264,000/mm³, and a negative blood β -hCG.

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Figure 1: initial physical examination showing 1.5-cm size endometrial implant on the inferior aspect of the umbilicus.

An abdominal wall ultrasound revealed a small collection in the umbilical region with central necrosis, extending 27x10x9 mm, with a rounded edge and a fistulous path measuring 2.5 mm in diameter and 10 mm in length, limited to the parietal fatty layer. The diagnosis of endometriosis was suggested based on the cyclical nature of her symptoms.

A pelvic MRI showed a normal-sized uterus, ovaries, and vaginal tract with no endometrial or cervical thickening and no laterouterine mass. It revealed an oblong lesion in the pouch of Douglas, measuring 7.4x6 mm, with T2 hypersignal and gadolinium enhancement, attaching the uterus to the rectum. Additionally, subcutaneous thickening in the umbilical area with gadolinium enhancement measured 14.4x24 mm. These findings suggested endometriosis (figure 2).

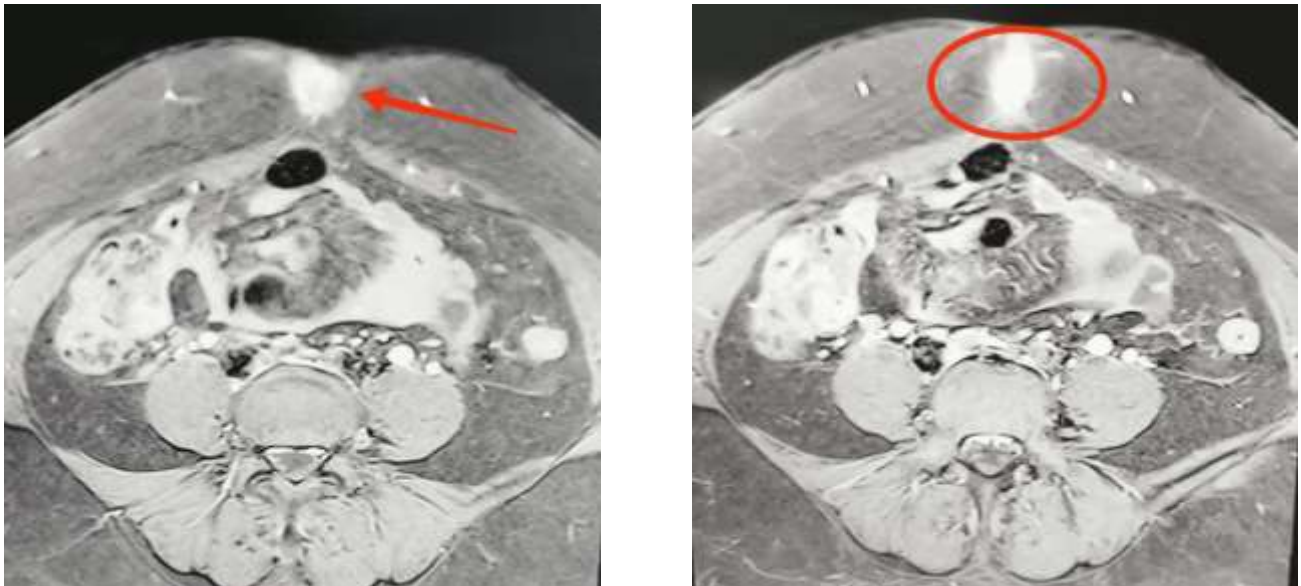


Figure 2: Pelvic MRI images of the patient showing a subcutaneous hypersignal T2 (red arrow and red circle), referring to endometriosis in the umbilical area.

The decision was made to treat the affected tissue with radical local excision and umbilical reconstruction. The surgery, performed at the end of the menstrual cycle, involved excising the nodule with a 1-cm safety margin up to the rectus fascia and performing an umbilicoplasty to close the umbilical defect. Pathology confirmed the presence of endometrial implants within the umbilical mass. At the postoperative visit, the patient reported satisfaction with the results. She was applying a topical antibiotic and healing cream to the umbilicus as instructed. Clinical examination showed excellent healing of the umbilicoplasty.

DISCUSSION

Endometriosis is a benign gynecological disorder characterized by the presence of endometrial tissue outside the uterus, often on pelvic organs such as the ovaries, fallopian tubes, pelvic peritoneum, uterosacral ligaments, and broad ligaments. Approximately 12% of endometriosis lesions are extragenital, occurring in locations such as the brain, lungs, gastrointestinal tract, urinary system, and musculature. Among extragenital sites, the abdominal wall (AWE) is the most common location for endometriosis.

Umbilical endometriosis (UE) involves the presence of endometrial glands and/or stroma within the umbilicus. It is a rare form of endometriosis, accounting for 30-40% of AWE cases, 0.4-4% of extragenital lesions, and 0.5-1% of all endometriosis cases [1], [4], [5].

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There are two types of UE:

- Primary UE: First described by Villar in 1886, it is also known as Villar's nodule [6]. Primary UE occurs without a history of surgery. Its pathogenesis is unclear, but theories include coelomic metaplasia, congenital presence of displaced endometrial tissue, direct extension through the round ligament or the patent omphalo-mesenteric duct, and mechanical seeding of endometrial tissues via the lymphatic or venous system.
- Secondary UE: This type develops on scar tissue following abdominal procedures such as laparoscopy.

Understanding the distinction between primary and secondary UE is important for comprehending the disease's pathogenic mechanisms. According to Hirata et al. (2020), the risk of malignant transformation of UE is about 3% [2], [5].

Due to its low frequency, limited data are available on the prevalence of primary and secondary UE and the associated symptoms [1]. Diagnosis of umbilical endometriosis is primarily clinical, with histopathology confirming the diagnosis [3]. Common clinical symptoms include cyclic, menstrual-related focal abdominal pain, swelling, and bleeding [2]. UE typically presents as a red, purple, or black umbilical nodule, measuring between 0.5 and 3 cm in diameter [5].

Imaging techniques can improve diagnostic accuracy:

- Ultrasound: Although not specific for an abdominal endometrioma, it may show a mass in the abdominal wall that appears solid, hypoechoic, and may contain internal vascularity and cystic areas. The differential diagnosis includes neoplasms (sarcoma or lymphoma), suture granuloma, ventral hernia, abscess, or hematoma.
- CT and MRI: These modalities also show a solid mass in the abdominal wall and are useful for characterizing the extent of the disease/mass preoperatively, though they cannot definitively diagnose endometriosis.

While ultrasound-guided fine needle aspiration can provide a definitive diagnosis, wide surgical excision is recommended for both diagnosis and treatment of an abdominal wall mass [4].

Surgery may involve resection of the umbilical mass with wide local excision (en-bloc omphalectomy) and, if possible, evaluation for pelvic endometriosis via laparoscopy. Surgical treatment is strongly recommended despite limited evidence on long-term efficacy and complications.

Medical treatment, such as combined oral contraceptives or progestins to reduce the stimulation and inflammatory effects of endometriotic implants, is weakly recommended due to limited supporting data and a lack of comparative studies between medical and surgical treatments for umbilical endometriosis [1], [3].

CONCLUSION

The umbilical endometriosis remains a very uncommon condition. However, it should be considered in the differential diagnosis of umbilical lesions. Our case report is an opportunity to highlight the importance of recognizing UE whenever a case of umbilical swelling or bleeding and cyclic pain occurs. It is not mandatory, as depicted in our patient, to have a surgical history.

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