

# Menstruating From the Umbilical Nodule with Pain as a Rare Case of Primary Umbilical Endometriosis: A Case Report and Discussion on Management Options

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**Keywords** Umbilical Nodule; Endometriosis; Gynecological disorder; Umbilical endometriosis

**Abbreviations** MRI: Magnetic Resonance Imaging; GnRH: Gonadotropin Releasing Hormone; UE: Umbilical Endometriosis

## Abstract

**Introduction:** Endometriosis is a common gynecological condition and presents mainly with involvement of the pelvic organs. Extrapelvic presentations in almost all parts of the body have been reported in the literature. However, umbilical endometriosis that is spontaneous or secondary to surgery is uncommon and accounts for only 0.5% to 1% of all endometriosis cases. We report a recently observed case of primary Umbilical Endometriosis (UE), with the main aim to discuss the management of this rare condition.

**Presentation of case:** A 35-year-old woman P2L2, NFVD complained of a painful nodule on her umbilical region, bleeding with her menstrual cycle. Ultrasonography showed a hypoechoic lesion with cystic area 2.1 x 2.8 x 1.8 cm in the umbilicus and no signs of intra-abdominal endometriosis. Wide Excision with 3cm margin of the nodule under spinal anesthesia was performed. Histopathological analysis confirmed the diagnoses that the patient was asymptomatic at follow-up, but nevertheless warned of the risk of recurrence of umbilical endometriosis.

**Discussion:** Pelvic endometriosis is a common condition, but the diagnosis of primary umbilical endometriosis is difficult and differentials should be considered. Although there is a substantial agreement about the necessity of surgery, treatment options are either local excision of the lesion or removal of the whole umbilicus with or without laparoscopic exploration of the peritoneal cavity. The decision should be tailored for the individual patient, taking into consideration the size of the lesion, the duration of symptoms and the presence of possible pelvic endometriosis.

**Conclusion:** Wide local excision may be the treatment of choice in patients with UE lesions and differential diagnosis of endometriosis should be considered when an umbilical swelling presents in a woman of reproductive age. No recurrence noted in our in this case after follow up for 1 year.

## Introduction

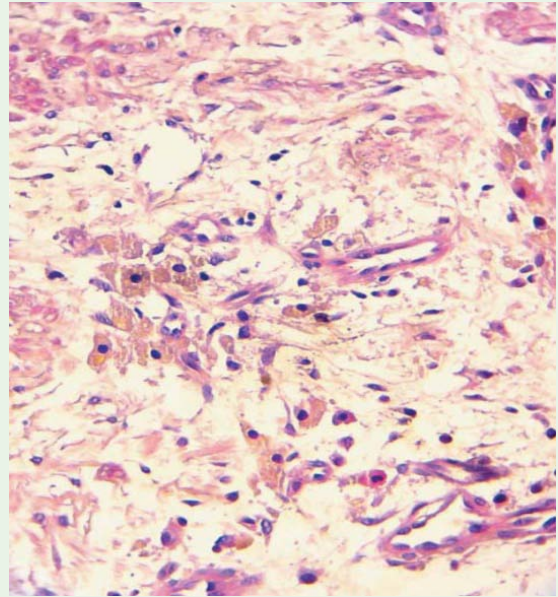
Endometriosis is defined as the presence of endometrial glands and stroma abnormally located outside the uterine cavity. It is a benign gynecological disorder affecting 10-15 % of all women of reproductive age and represents an important cause of infertility [1,2]. Although different theories have been postulated in order to elucidate the patho-physiology of this condition, to date none of them has been proven to be to be completely exhaustive. Common locations of endometriosis are the pelvic organs, mostly the ovaries, the Fallopian tubes, the utero-sacral ligaments, the recto-vaginal septum and the pelvic peritoneum. Extragenital endometriosis is less common, but has been described in almost every area of the female body including the bowel, bladder, lungs, brain, umbilicus, and surgical scars [3]. Due to its varied presentations, endometriosis remains a difficult condition to diagnose and treat. Umbilical endometriosis represents 0.5% to 1% of all cases of extragenital endometriosis. It usually occurs secondary to surgical scars, but very rarely presents as primary umbilical endometriosis [4,5]. We report one such rare case of spontaneous, primary umbilical endometriosis. Due to the rarity of this entity, no guidelines for treatment exist. We report a recently observed case with the main goal being to discuss the treatment options.

## Presentation of Case

A 35-year old woman, gravida 2 para 2 normal full term vaginal delivery, was admitted to outpatient clinics with 1 year history of umbilical nodule. She stated that the nodule had slowly increased in size and had started to bleed concomitantly with the menstrual periods with pain in the previous 8 months. Her medical history was unremarkable and she denied symptoms of pelvic endometriosis such as dysmenorrhea, abdominal pain or dyspareunia. She was not taking any oral contraceptives and had regular menstrual cycles. Physical examination revealed a black, non tender



**Figure 1:** A black, non tender nodule of about 3 x 3 cm centimeter in diameter.



**Figure 3:** Endometriotic glands with mucinous type metaplasia and extravasation of the mucinous secretion into the adjacent stroma.



**Figure 2:** Local excised nodule.

nodule of about 3 x 3 cm centimeter in diameter located deep in the umbilical fold which appeared to be covered by a reddish brown discharge on menses (Figure 1). She then underwent an ultrasound scan that showed a hypochoic lesion with cystic area 2.1 x 2.8 x 1.8 cm in the umbilicus and no signs of intra-abdominal endometriosis.

The patient was offered both medical and surgical management and she opted to have depot injections of Zoladex (Goserelin acetate, 3.6mg subcutaneously, monthly). The swelling continued to persist in spite of three doses of Zoladex, and the patient then requested surgical excision. The risk of recurrence and scar endometriosis were explained to her. The patient successfully underwent wide local excision of the nodule (Figure 2). Histology confirmed the diagnosis of endometriosis and revealed the presence of endometriotic glands with mucinous type metaplasia and extravasation of the mucinous secretion into the adjacent stroma (Figure 3). No epithelial atypia was seen and the excision appeared complete. The patient was followed up 1 year after the surgery and found to be asymptomatic. Before being discharged, the patient was again reminded of the risk of recurrence.

### Discussion

The deposition of fragments of uterine endometrium in the skin is a well recognized, although uncommon, phenomenon (0.5% to 1% of extragenital endometriosis). Umbilical endometriosis was first described in 1886 and since then more than 100 cases have been described [4]. Majority of these cases occurred secondary to surgical, commonly laparoscopy, scars. An umbilical endometriotic lesion without surgical history is a rare condition [4,5]. Some case reports have also described the presence of umbilical endometriosis during pregnancy [6]. There has been great speculation about the pathogenesis of this phenomenon and several theories have been proposed. Latcher has classified these theories into three main categories: The embryonal rest theory, which explains endometriosis adjoining the pelvic viscera by Wolffian or Mullerian remnants [4,5]; the coelomic metaplasia theory, which states that the embryonic coelomic mesothelium dedifferentiates into endometrial tissue under stimulus such as inflammation or trauma [7]; and the migratory pathogenesis theory, which explains the dispersion of endometrial tissue by direct extension, vascular and lymphatic channels, and surgical

manipulation. Still others suggest cellular proliferation of endometrial cells from initial extraperitoneal disease along the urachus [8,9]. The real mechanism still remains a mystery. These patients are usually in the reproductive age group and present commonly with swelling, pain, discharge or cyclical bleeding from the umbilicus. There may be associated symptoms of coexistent pelvic endometriosis. These lesions are usually bluish-black in color and become painful, larger and bleed about the time of menses. They range in size from 0.5cm to 3cm, but can enlarge to even more enormous sizes [4]. While the diagnosis is primarily clinical, Magnetic Resonance Imaging (MRI) can be useful in evaluating patients, with suspected endometriosis. Endometriomas appear homogeneously hyperintense on T1-weighted sequences [10]. MRI also has an advantage over laparoscopy for evaluating pelvic and extraperitoneal diseases, as well as lesions concealed by adhesions. Histological findings are characterized by irregular glandular lumina embedded in the stroma with a high cellular and vascular component resembling the stroma of functional endometrium. A fairly recent study has suggested a distinctive dermatoscopic feature in cutaneous endometriosis that of comprising small red globular structures called 'red atolls' [11]. Differential diagnosis of umbilical nodules should include pyogenic granuloma, hernia, residual embryonic tissue, primary or metastatic adenocarcinoma (Sister Joseph's nodule), nodular melanoma, and cutaneous endosalpingosis. In severe cases or in the presence of pelvic endometriosis, hormonal therapy in the form of danazol or GnRH analogues can be given to the patient [12]. Surgical treatment is recommended for several reasons. Firstly, the removal of the entire lesion only enables accurate histopathological diagnosis of UE, thus excluding unusual malignant disorders of the umbilicus, such as metastases or skin neoplasms [13,14]. Furthermore, removal of UE nodule is warranted because malignant transformation of endometriotic lesions, although rare, has been described [14,15,16,17].

In our case wide local excision of lesion is done and histology confirmed the diagnosis. Although simultaneous laparoscopy has been recommended for pelvic endometriosis, this was not done because our patient was asymptomatic. Although local recurrence is uncommon, the patient has been warned of the risk of scar endometriosis and of recurrence.

## Conclusions

Endometriosis is a common gynaecological disease; however, primary umbilical endometriosis is very rare. Making a diagnosis is difficult and other causes of umbilical lesions should be considered. Surgical wide local excision is the standard treatment of this condition. The patient was followed up 1 year after the surgery and found to be asymptomatic.

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