

## Umbilical Endometriosis, Case Report. Is there a Causative Relation with Premenarchal Surgery?

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## 1. Abstract

Umbilical endometriosis is the most common site of cutaneous endometriosis, and it often follows surgical procedures where a iatrogenic implantation of endometriotic tissue occurs. We report a case of a woman with umbilical endometriosis with history of hernia repair during early childhood. The patient presented a 3 cm umbilical nodule that had progressively increased in size, together with dysmenorrhea and chronic pelvic pain. After physical examination and imaging techniques the patient was diagnosed of pelvic endometriosis, adenomyosis, and an umbilical nodule of 3 cm suggestive of endometriosis. A core needle biopsy of the lesion described glandular structures that express estrogen receptors surrounded by hemosiderin-lade macrophages. Hormonal treatment failed to improve symptoms and an en-block resection of the nodule was performed. The pathologist reported presence of numerous endometrial glands and residual suture threads with foreign-body granulomatous reaction within the nodule. The nature of this umbilical endometrial implantation remains elusive. Even though the infiltration of endometriotic cells at the umbilical scarring tissue

should be considered in a patient with pelvic endometriosis, the history of premenarchal surgery should not be ignored. Neonatal bleeding could be the source of endometrial cells implanting in the peritoneum and premenarchal surgery should be considered as a risk factor when cutaneous endometriosis occurs.

## 2. Introduction

Umbilical endometriosis (UE), also known as Villar's nodule, was first described in 1886 as the presence of endometrial glands in the umbilicus [1]. It is an uncommon type of endometriosis, with a reported incidence of 0.5-1% of all cases of endometriosis [5]. Despite this, it represents the most common site of cutaneous endometriosis, affecting 30-40 % of patients with abdominal-wall endometriosis [7].

Primary cutaneous UE is a rare event that occurs in absence of any previous abdominal surgery. Several theories have been proposed including coelomic metaplasia, lymphatic or even hematogenous dissemination [10], albeit its pathogenesis remain elusive. In contrast, secondary UE is caused by iatrogenic implantation of endometriotic tissue, often following surgical procedures, such as

laparoscopy [4].

Previous studies reported a higher incidence of secondary UE compared to primary. However, data from a national survey in Japan from 2020 among 116 UE reported that UE was related to previous surgery only in 30% of the cases [6]. These results were later confirmed in a systematic review that documented history of a surgical procedure in 37.6% of the 232 women with UE [5]. These surgeries comprehend caesarean section, laparotomy other than caesarean section, and laparoscopy, procedures mostly performed in women in their reproductive age.

We present the clinical case of a woman with umbilical endometriosis with previous history of an umbilical hernia repaired during early childhood.

### 3. Case Report

A 35-year-old woman presented for evaluation of an umbilical nodule, which had progressively increased in size over the past 2 years, together with dysmenorrhea and chronic pelvic pain. The patient's prior medical history revealed umbilical hernia repair at the age of 5. Besides, the patient was in follow-up for Sjogren's syndrome, Juvenile myoclonic epilepsy, and Central sensitization syndrome expressed with fibromyalgia and chronic fatigue syndrome, irritable colon, and interstitial cystitis.

At physical examination the patient had a livid, soft and cyclic painful swelling in the umbilicus of 3 cm (Figure 1), irreducible by digital pressure. The pelvic examination together with the vaginal ultrasonography revealed a retroverted uterus, with a question mark form, ovaries without cysts but firmly adhered to the sides of the uterus, suggesting endometriosis and adenomyosis.

The abdominal ultrasonography observed a hypoechoic nodule at the umbilicus of 30 x 8.3 mm (Figure 2). A core-needle biopsy evidenced glandular structures that express estrogen receptors surrounded by hemosiderin-lade macrophages supporting the diagnosis of umbilical endometriosis.

A fat suppressed T2-weighted MRI was performed to further examine the extension of the umbilical lesion. The nodule exhibited diffuse enhancement after contrast injection and linear tails of extension along the underlying aponeurosis suggesting fascial extension/involvement.

Combined oral contraception was initiated and the pelvic component responded successfully, with optimal pelvic pain management and a reduction of the pelvic endometriotic disease. However, the umbilical nodule failed to respond. An enbloc resection was performed under general anesthesia, and the histological examination described scarring connective tissue that contained numerous endometrial glands surrounded by endometrial stroma. In the core of the nodule, residual suture threads were observed together with foreign-body granulomatous reaction. In addition, the margins of the resected nodule were free of endometrial tissue. The postoperative recovery was uneventful and a month after the surgery, the patient

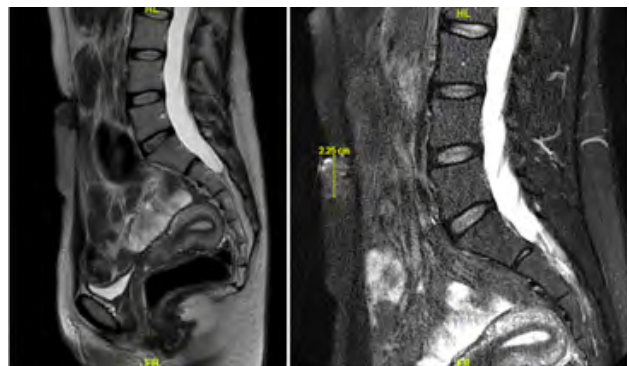
referred full remission of the painful sensation and swelling in the umbilicus.



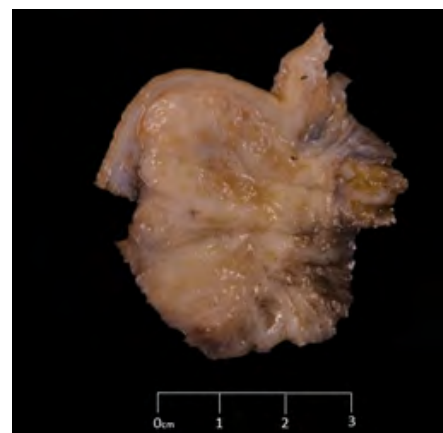
**Figure 1:** Umbilical endometriosis nodule



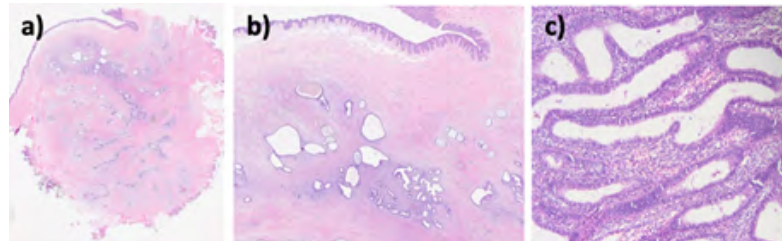
**Figure 2:** Abdominal ultrasonography revealing an umbilical nodule



**Figure 3:** Magnetic resonance imaging an umbilical nodule in a sagittal fat suppressed T2-weighted sequences and contrast enhanced image



**Figure 4:** Macroscopic view of the excised nodule.



**Figure 5:** Microscopic view of the nodule; a) nodular lesion composed of scar-like connective tissue, with numerous endometrial-type glands; b) glandular structures and stromal cells in the deep dermis are identified in more detail, in relation to the epidermis; c) glandular and stromal cellularity at a higher magnification (x10).

#### 4. Discussion

Cutaneous endometriosis is related to previous surgeries in less than 30% of the cases, where a iatrogenic implant of endometriotic cells occur during the surgical procedure. However, these cases are mostly performed after menarche, once endometrial shedding and menstrual bleeding begin to occur.

This case represents an uncommon form of umbilical endometriosis, since the patient underwent umbilical hernia repair during early childhood, long before the menarche.

Discerning whether this case corresponds to a primary or a secondary UE is key to better understand the physiopathology of the disease. It has been suggested that when pelvic endometriosis coexists, the lymphatic and hematogenous spread of endometrial tissue through the round ligaments or omphalomesenteric remnants could explain the growth of an UE nodule [12].

Despite this later, the development of an UE nodule arising independently from the pelvic disease cannot be excluded. In this regard, the epithelial and stromal cells may have developed from the persistence of coelomic epithelial cells in the umbilicus by metaplasia [8]. It is known that coelomic epithelial cells are present physiologically in the umbilicus during embryogenesis, although regress at 12 weeks gestation once the physiological omphalocele regress. However, the persistence of an umbilical hernia after birth has been speculated to be a possible causative factor for the persistence of multipotent coelom epithelial cells within the umbilicus [8, 9]. Interestingly, the surgery performed in our patient was reported to be an umbilical hernia repair at the age of 5, and even though the patient presented pelvic concomitant disease, the hypothesis of the coelomic metaplasia cannot be excluded.

Sampson, in 1920s, hypothesized that endometriosis results from the retrograde menstruation that reaches the pelvis through the fallopian tubes [11]. Brosens, however, argued that Sampson's theory could only explain endometriosis occurring after menarche, once endometrial shedding and menstrual bleeding begin to occur [2].

Previous authors had described that the neonatal uterus is capable of shedding, and that even though overt vaginal bleeding is only observed in about 5% of neonates, occult bleeding occurs in the majority of neonates [3].

In this regard, Brosens et al. proposed that the functional plugging

of the endocervical canal in neonates could promote the retrograde flux of endometrial stem cells into the peritoneum and disseminate in the pelvis at the time of neonatal uterine bleeding [3]. This hypothesis could justify cases of endometriosis that rise soon after menarche and even in premenarchal girls.

If we assume the possibility of endometrial stem cells present in the peritoneal cavity before menarche, abdominal surgeries during childhood could therefore lead to the iatrogenic spread of these cells in scars, and once hormonal stimulation occurs these cells could grow into endometriotic nodules. Hence, we need to consider the possible causative relation between an umbilical hernia repair in early childhood and the growth of an umbilical endometriotic nodule, particularly on a patient who already suffered from pelvic endometriosis and adenomyosis.

#### 5. Conclusion

The physiopathological mechanism involved in the umbilical nodule presented herein remains elusive. The lymphatic or hematogenous spread needs to be considered particularly when pelvic endometriosis coexists. However, the history of an umbilical hernia needs to be acknowledged. The persistence of an umbilical hernia after birth entails a higher risk of coelomic metaplasia at this site. Despite this, an abdominal surgery can act as a risk factor if we assume the possibility of the neonatal spread of endometrial stem cells into the peritoneal cavity, especially in a woman with concomitant pelvic endometriosis.

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