Spontaneous Umbilical Endometriosis: Case Report of a Cyclical Painful Umbilical Swelling

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Abstract

Background: Primary or spontaneous umbilical endometriosis is the presence of ectopic endometrial tissue in the umbilicus in the absence of previous surgery. It is a rare entity but should be considered as a possible differential diagnosis in cases of umbilical disorders even if the patient has no typical symptoms of pelvic endometriosis.

Case Report: We reported a 24-year-old nulliparous lady with a 2-year history of a cyclical umbilical swelling and pain. She was managed by a complete umbilical excision and diagnosis was confirmed with histology.

Conclusion: Spontaneous umbilical endometriosis should be considered in women of reproductive age when they present with catamenial umbilical pain and nodule.

Keywords: Catamenial; Spontaneous endometriosis; Umbilicus

Introduction

The umbilicus occasionally may become a focus of clinical attention, as a variety of conditions are known to affect it. The differentials of an umbilical mass or nodule are diverse and may sometimes present a diagnostic challenge or uncertainty [1].

The umbilicus is the commonest cutaneous site of endometriosis outside of the pelvis; however, it remains a rare occurrence with an incidence of approximately 0.5-1.2% in all cases of endometriosis. [2] Umbilical endometriosis could be primary or secondary in origin. [3] Primary umbilical endometriosis develops spontaneously in the absence of any prior abdominal surgery and it is also known as Villar's nodule, first described by Dr Villar in 1886. [4] Secondary endometriosis, also known as scar endometriosis, occurs at the site of previous surgery by iatrogenic implantation in surgical scars [5].

The pathophysiology of primary (spontaneous) umbilical endometriosis is unknown. However, multiple theories have been postulated. The most accepted explanations are implantation of endometrial tissue during retrograde menstruation (implantation theory), haematological or lymphatic dissemination of endometrial cells (dissemination theory), and differentiation of pluripotent peritoneal progenitor cells into endometrial tissue (coelomic metaplasia theory) [6].

The most characteristic symptom of spontaneous umbilical endometriosis is cyclical pain together with a palpable mass in women of reproductive age. [7] The temporal association of these features in relation to the menstrual period is of utmost importance for the clinical diagnosis of the disease.

Radiological investigations have been done in many studies, but they are not conclusive for diagnosis. Histopathology remains the hallmark for the diagnosis, which reveals the endometrial glands, stroma, and hemosiderin pigments. Surgical intervention is the standard modality of treatment [8].

The aim of our case presentation is to report this uncommon condition in order to increase the index of suspicion among medical practitioners as spontaneous umbilical endometriosis is still an enigma and often, the diagnosis is delayed.

Case Report

A 24-year-old nulliparous lady who had been married for 2 years presented to the general surgery clinic with a cyclical umbilical swelling and pain of 2 years. The swelling increased in size during menstruation with associated increase in the severity of the pain and thereafter, regressed.

She attained menarche at 13 years and had regular menstrual cycles, but she has never been pregnant. She did not experience any abnormal vaginal bleeding, dysmenorrhoea, dyspareunia or pelvic pain and was not on any form of contraception. She had no gastrointestinal, urinary symptoms, weight loss, chest pain, cough or haemoptysis. Her medical and surgical history were unremarkable.

Physical examination revealed a young lady with hyperpigmented periumbilical area and a tender umbilical nodule of approximately 2x3cm, which was irreducible by gentle digital pressure (Figure 1). Umbilical endometriosis was suspected based on her clinical findings. Ultrasound scan of the abdomen only suggested a suprambilical hernia with uterine fibroids.
The patient had excision of the nodule together with the umbilicus under general anaesthesia (Figure 2). The fascia was repaired by simple interrupted sutures with nylon 2. The skin closure was done with simple interrupted non-absorbable sutures.

Histopathological examination of the excised tissue revealed a lesion composed of variable sized endometrial glands and stroma within the dermis. The glands were lined by cuboidal cells (Figures 3a, 3b). Concluding, the findings were consistent with the diagnosis of umbilical endometriosis. She did well postoperatively and was symptom free on follow up at 6 weeks.

Discussion

Endometriosis is the presence of functional endometrial tissue and glands outside the endometrial cavity. [9] While umbilical involvement is uncommon, it is more prevalent than other extra-pelvic localizations. Primary or spontaneous umbilical endometriosis is a rare entity but should be considered as a possible differential diagnosis in cases of umbilical disorders even if the patient has no typical symptoms of pelvic endometriosis. [10] Various cases have been reported in African-indigenous women presenting with umbilical nodules, sometime associated with a history of previous uterine surgery. [11] The highlights of the case described herein, is that the patient had spontaneous umbilical endometriosis, i.e. the presence of endometrial tissue in the umbilicus in the absence of previous surgery for either gynaecological disorder or caesarean section.

Spontaneous umbilical endometriosis commonly manifests as a red, brownish, or flesh-coloured umbilical nodule. Pain, bleeding, or swelling of the lesion are the most prevalent symptoms, especially during menstruation. An asymptomatic variant, on the other hand, should not be neglected. [12] In our patient, catamenial pain and swelling were the most noticeable symptoms. In a systematic review by Victory et al., umbilical swelling was present in almost 90% of the cases with less than 50% having bleeding and about 80% having pain. Pain is caused by tissue inflammation, distension, and cyclical changes.

The differential diagnosis of an umbilical nodule includes umbilical hernia, pyogenic granuloma, keloid, melanoma, primary or metastatic carcinoma which is eponymously known as Sister Mary Joseph’s nodule. The presence of such a nodule has been generally regarded as a sign with a grave prognosis and had implied therapeutic abstention. The most common primary sources in descending frequency are colonic, gastric, and ovarian cancer. Nevertheless, it has recently been demonstrated that most umbilical lesions are benign. [13,14] However, it is important to consider all causes, including endometriosis, in patients who present with an umbilical mass.

Although preliminary diagnosis is made based on history and physical examination, imaging may aid in preoperative evaluation. The sensitivity and specificity of ultrasonography, computerized tomography and magnetic resonance are low in the diagnosis of umbilical endometriosis. None of these imaging modalities have a pathognomonic finding of umbilical endometriosis. [15] They are useful for ruling out involvement of other organs and establishing the depth of lesions. For example, ultrasound can be used to assess the size of the nodule, the involvement of surrounding tissues, and to evaluate other pelvic pathology, hence aid in the planning of surgical management. In the index patient, even though ultrasound missed the diagnosis of umbilical endometriosis, it aided in establishing the presence of the uterine fibroids. Although rare, primary umbilical endometriosis coexisting with uterine fibroids has been reported by Mba et al. in Enugu, Nigeria. It should be suspected in women of reproductive age who complain of cyclical umbilical disorders in addition to abdominal swelling or other symptoms of uterine fibroid.

The tentative diagnosis which is based on clinical findings is confirmed by histology just as was the case with this patient. Apart from

Figure 1: Umbilical nodule with surrounding area of hyperpigmentation.

Figure 2: Intraoperative image of excised umbilicus with nodule.
endometrial glands and stroma, histology may also show hemosiderin deposits, inflammatory changes, and marked mitotic activity. Immunostaining with CD10, which is positive in the endometrial interstitium, is useful when glands are not seen on histology [16].

Treatments for umbilical endometriosis are limited but Victory et al. believe that surgical management is the treatment of choice. The choice of the surgical technique depends on the size and depth of the lesion and must be individualized. Because umbilical endometriosis is very rare, standard surgical consensus regarding its treatment is still very lacking. Currently, the options include a complete umbilical resection with or without fascial and peritoneal repair or a local excision of an endometriotic nodule. The most frequently performed operation for umbilical endometriosis is total resection of the umbilicus. [17] This option carries a low risk of recurrence if complete excision is successfully performed. [18] The patient in the present report had an en-bloc resection of the umbilicus and fascial repair. Some other authors have equally recommended a complete umbilical resection regardless of the size even though this operation results in inferior cosmetic outcome. [19,20] Hormonal therapy in umbilical endometriosis can only temporarily reduce its clinical symptoms, which will recur after its cessation if surgery is not done. The hormones commonly used are oral contraceptives and Gonadotropin-releasing hormone agonists.

**Conclusion**

Umbilical endometriosis is a rare entity which should be considered in women of reproductive age presenting with catamenial umbilical pain and nodule. It is important to recognize that it can occur spontaneously in patients with no previous surgical history as was the case with our index patient. The treatment of choice is surgical excision, and diagnosis is confirmed by histopathological examination. Hopefully, our case report will raise awareness of this uncommon condition and provide understanding of the clinical presentation and treatment.

**References**
