

Umbilical Endometriosis Associated with Large Umbilical Hernia. Case Report

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Rezumat

Endometrioza ombilicală asociată cu hernie ombilicală de mari dimensiuni. Prezentare de caz

Endometrioza ombilicală este o afecțiune rară, ce survine în general unei intervenții chirurgicale generale sau laparoscopice implicând ombilicul. Endometrioza ombilicală spontană, la o persoană ce nu a suferit nici o intervenție abdominală sau uterină este extrem de rară. Grosimea maximă de penetrare a endometriozei ombilicale a fost descrisă până la nivelul fasciei. Au existat doar două cazuri descrise de endometrioza apărută în cadrul unei hernii ombilicale. Autorii prezintă cazul unei paciente cu endometrioza spontană ombilicală asociată cu o hernie ombilicală de mari dimensiuni, tratată prin excizie chirurgicală și reparare cu proteză tip mesh de material textil a herniei. Din câte cunoaștem, este primul caz descris de asociere a endometriozei ombilicale cu hernie ombilicală de mari dimensiuni ce necesită reparare prostetică a defectului de perete abdominal.

Cuvinte cheie: endometrioza ombilicală, hernie, tratament chirurgical

Abstract

Umbilical endometriosis is a rare condition, usually following laparoscopic and surgical procedures involving the umbilicus. Spontaneous umbilical endometriosis occurring without any previous abdominal or uterine surgery is extremely rare. The maximal depth of penetration of the umbilical endometriosis described is up to fascial level. There have been only two cases of endometriosis reported arising within umbilical hernia. The authors report a case of a patient with spontaneous umbilical endometriosis associated with a large umbilical hernia, treated by surgical excision and mesh repair of the abdominal wall. To the best of our knowledge, this is the first described case of the association of umbilical endometriosis with a large umbilical hernia that requires prosthetic mesh repair of the abdominal wall defect.

Key words: umbilical endometriosis, hernia, surgical treatment

Introduction

Endometriosis is defined as the presence of endometrium or endometrium-like tissue outside the endometrial cavity. This is a relatively common disease that affects up to 22% of all women, about 20%–30% of patients presenting with subfertility, and up to 45% of women with pelvic pain (1,2). Endometriosis is dominantly found in the pelvis, but when it develops outside the uterus or ovary it is called ectopic or extra-gonadal endometriosis (3). Umbilical endometriosis is a rare condition, with an estimated incidence of 0.5 to 1%

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of all patients with endometrial ectopia (4,5). The development of umbilical endometriosis usually follows laparoscopic surgical procedures involving the umbilicus (6). Spontaneous umbilical endometriosis occurring without any previous abdominal or uterine surgery is extremely rare (7,8). From 1966 to 2007, Victory et al. found only 122 reported cases of umbilical endometriosis worldwide (13). The maximal depth of penetration of the umbilical endometriosis described is up to fascial level (1,13). However, to the best of our knowledge, there have been only two cases of endometriosis reported arising within an umbilical hernia (8,14). Association of the umbilical endometriosis with a large umbilical hernia that requires prosthetic mesh repair of the abdominal wall defect is not described in the English medical literature. We report a case of a patient with spontaneous umbilical endometriosis associated with a large umbilical hernia, treated by surgical excision and mesh repair of the abdominal wall.

Case report

A 38-year-old lady was presented to a general surgeon because of a large irreducible umbilical hernia, without signs of bowel obstruction. The hernia was associated with a painful secretory lump in the umbilicus for the past five years. She had not reported history of any surgery in the past, including laparoscopy. General physical examination was essentially unremarkable. Local examination revealed a large umbilical hernia (5.5 cm in diameter) with moderately eroded red-bluish coloured skin over a firm, well-defined, tender nodular mass inside the hernial sac, measuring 3 cm in diameter. There was a minimal sero-sanguineous superficial secretion at the moment. The nodule was irreducible (Fig. 1).

The surgeon's first impression was of presence of incarcerated omentum in the umbilical hernia, with trophic skin changes, but the patient noted that the pain and constant minimal serosanguinous secretion from the umbilicus would get worse during her periods. Because of that, the surgeon paid attention to the ob/gyn anamnesis. Her menstrual cycles were regular with a normal flow. She did not give a history of cyclical pelvic discomfort or significant dysmenorrhoea, subfertility or any treatment for infertility in the past. She had a vaginal delivery fifteen years ago. She did not provide history of a hysterotomy or any other pelvic surgery, including laparoscopy, in the past. Ultrasound of the abdomen noted an echogenic nodular space occupying lesion located in the hernial sac, with no bowel loops inside. The nodule was attached to the hernial (umbilical) ring. Biochemical and haematological parameters were normal. Gynaecological examination revealed that the uterus was of normal size, anteverted, mobile and non-tender. Pelvic ultrasound noted that the uterus and adnexa appeared normal.

On the basis of clinical and ultrasound findings of the umbilical nodule with cyclic pain and secretion, the possibility of external endometriosis was kept in mind and the patient was taken up for open hernia repair and surgical excision of the nodule. After the elliptical skin excision and lateral dissection, deep in the subcutaneous tissue, we registered the presence of

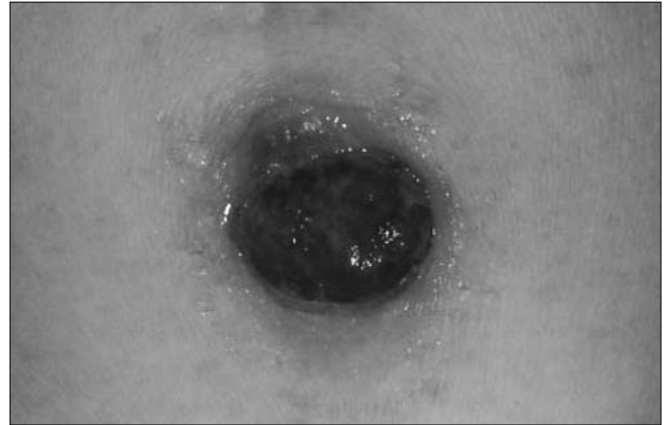


Figure 1. Macroscopic presentation of the umbilical endometriosis associated with an umbilical hernia

a firm, purplish nodule of 3 cm in diameter. The nodule itself was densely adherent to the lateral, extraperitoneal side of the peritoneal sac and musculo-aponeurotic hernial ring. The hernial sac was opened and partially excised in order to extract the nodule. After excision of the nodule, pelvic examination was performed and revealed an absolutely normal finding. The hernial defect was repaired using Prolene-mesh. The post-operative course was uneventful and the patient was discharged on the 7th postoperative day.

Surgical biopsies were fixed in 10% formaldehyde overnight, processed in paraffin wax and cut at 4 μ m. The sections were stained with H&E and immunohistochemical ABC method for oestrogen and progesterone receptor, CD10, BCL-2 oncoprotein and desmin. Histopathological examination of the umbilical nodule revealed plenty of endometrial glands surrounded by compact stroma (Fig. 2). Intensive nuclear staining of the endometrial glands and some of the cellular nuclei



Figure 2. The wall of the glandular cavity mainly consisting of a layer of tall columnar epithelia. Older and recent bleeding was observed inside the glandular cavity and in the interstitium. Spindle-shaped or stellate cells were observed in the interstitium (haematoxylin-eosin stain, original magnification x100)

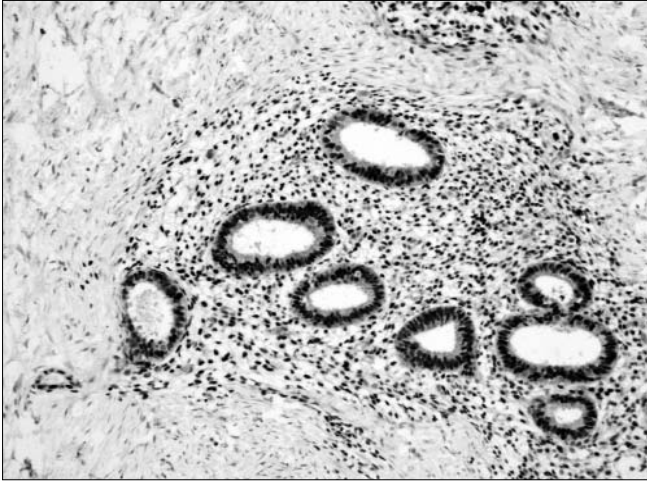


Figure 3. Intensive nuclear staining of the endometrial glands and some of the cellular nuclei from the interstitium for oestrogen receptor (immunohistochemical ABC method, original magnification $\times 100$)

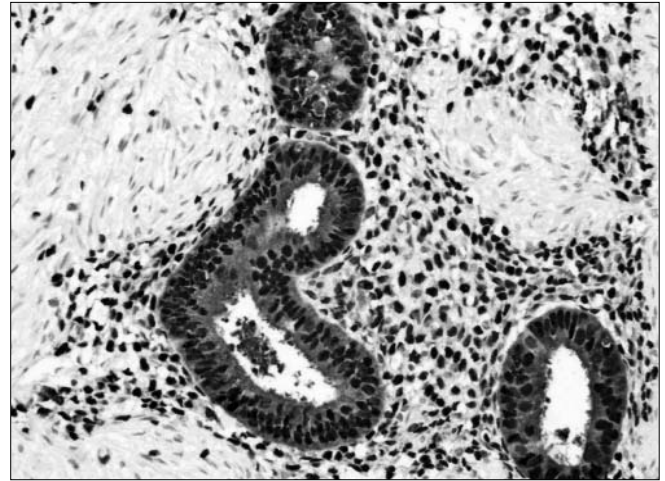


Figure 4. Strong nuclear staining of the endometrial glands and of the cellular nuclei from the interstitium for progesterone receptor (immunohistochemical ABC method, original magnification $\times 200$)

from the interstitium for oestrogen (Fig. 3.) and progesterone (Fig. 4.) receptor, confirmed the diagnosis of umbilical endometriosis. We registered also intensive intracytoplasmic immunoreactivity for BCL-2 oncoprotein of the glandular epithelium and moderate immunostaining for desmin in some of the spindle-shaped cells arranged around the glandular cavity.

Discussion

The most common locations of the extra-gonadal endometriosis are the lower parts of the gastrointestinal tract and the peritoneal surfaces. External endometriosis, involving the subcutaneous tissues, have also been reported in the presence of a surgical scar following surgeries such as hysterectomy, hysterotomy, caesarean section, episiotomy and laparoscopy (4,5).

The first description of an umbilical endometrioma is credited to Villar in 1886 (7), and hence the condition is sometimes referred to as Villar's nodule. Umbilical endometriosis is a rare condition, with an estimated incidence of 0.5 to 1% of all patients with endometrial ectopia (4,5). The development of umbilical endometriosis usually follows laparoscopic surgical procedures involving the umbilicus (6). Spontaneous umbilical endometriosis that occurred without any previous abdominal or uterine surgery is extremely rare (7,8). The pathogenesis of spontaneous endometriosis is not clear. The most widely accepted is the "hypothesis of migration – or retrograde menstruation", with the reflux of menstrual blood and implantation of the endometrial cells of the target organs (9); The second theory is "induction theory", the theory of "coelomic metaplasia" with the transformation from mesothelium to endometrium-like tissue under the influence of regurgitated endometrium. This theory is based on the observation that pluripotent cells of the coelom are able to differentiate into both endometrial and peritoneal cells (10). In the cases of cutaneous endometriosis, another possible mechanism could be a retrograde lymphatic flow with the

implantation of intra-abdominal endometrial cells into the subcutaneous tissue. (11). In the development of spontaneous umbilical endometriosis, as in the case presented, it is possible that the umbilicus acts as a physiological scar with a predilection for endometrial tissue (12).

Also, this may represent intra-abdominal endometriosis within the hernia sac which has prolapsed through the hernia orifice. (7). However, our patient did not have any intraperitoneal or pelvic endometriosis.

From 1966 to 2007, Victory et al found only 122 reported cases of umbilical endometriosis worldwide (13). The maximal depth of penetration of the umbilical endometriosis described is up to fascial level (1,13). However, to the best of our knowledge, there have been only two cases of endometriosis reported arising within an umbilical hernia (8,14).

The differential diagnosis of umbilical endometriosis is sometimes difficult. These lesions can be confused with a pigmented malignant tumour such as a melanoma, or in the cases that present with a subcutaneous mass such as a benign nevus, a lipoma, an abscess, a cyst, or a hernia, as well as a metastatic deposit from a systemic malignancy, should be considered. The use of Doppler US or MR could be useful, but not pathognomonic (5). Given the classic history and physical examination, no further studies are required before proceeding to the operative room for local excision. The diagnosis is often made incidentally by histological examination after surgical exploration and excision of the lesion (5,15,16).

Surgery is based upon radical excision of the lesion intact with minimal spillage in order to decrease recurrence (7). After surgery, a careful gynaecological assessment is recommended, considering that further intraperitoneal localization may be seen in the majority of cases /91%/ (17).

In our case, a large abdominal wall defect required a mesh repair, which results in lower recurrence rates and less discomfort in the long term, while mesh repair is not associated with an increased incidence of complications.

Association of the umbilical endometriosis with a large umbilical hernia requiring prosthetic mesh repair of the abdominal wall defect is not described in the English medical literature. So far, there have been very few reports on the use of mesh following the excision of cutaneous (1,7) or inguinal endometriosis, and no reports of recurrence of endometriosis in these patients. A longer follow-up on these patients is required to demonstrate this, due to the possibility of mesh contamination by endometriosis. (18). 28 months ago, we did not register the recurrence of endometriosis in our patient.

Conclusion

Endometriosis involving the subcutaneous tissues of the umbilicus must be suspected in a woman presenting with localized cyclical pain and swelling, in association with menstruation. Endometriosis within an umbilical hernia is a very rare condition. The diagnosis is often made incidentally by histological and immunohistochemical examination after surgical exploration and excision of the lesion. Large abdominal defect could be repaired using a prosthetic mesh with no recurrence of endometriosis.

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