INTRODUCTION

Endometriosis is defined by the presence of endometrial tissue (glands and stroma) outside the uterine cavity. It is a common gynecological disease that affects 5% to 10% of fertile women (1). The pelvis is the most common site of the disease, giving rise to the common presenting symptoms of pelvic pain, dysmenorrhea, dyspareunia, cyclic bowel or bladder symptoms, and infertility (1, 2).

Extrapelvic presentations have been described in almost every area of the female body including the lungs, brain, umbilicus, and surgical scars (3). Extrapelvic endometriosis is less common but even more difficult to diagnose and treat due to the extreme variability of its presentation.

However, primary and iatrogenic umbilical endometriosis is uncommon and accounts for only 0.5% to 1% of all patients with endometrial ectopia (4). We report one case of primary umbilical and pelvic endometriosis.

Case presentation

A 32-year-old Caucasian, nulliparous woman presented to the clinic with a one-year history of spontaneous and cyclic umbilical bleeding. Bleeding started at the beginning of her menses and continued for the entire duration of her period. It was accompanied by pain and swelling in the umbilical area. The patient had a regular, heavy and painful menstrual cycle. She had a previous history of abdominal pain, dyspareunia and infertility and had received 3 cycles of IUI three years before without any results. She did not report using any hormonal contraception. There was nothing of note in her medical history, including no previous abdominal surgeries. On examination, a 10 mm by 10 mm fixed umbilical nodule was noted. The nodule was coated in a reddish brown secretion and had the appearance of a pigmented tumor (Fig. 1).

No modification of hematological markers for CA 125 were detected. Magnetic resonance study with T2 and T1 fat saturation showed 2 round masses of 0.3 cm under the...
Primary umbilical endometriosis

umbilical scar (Fig. 2). Uterine section demonstrated the presence of one endometriotic nodule of the left utero sacral ligament. The patient received medical management with Lutenyl for two months. On taking Lutenyl the swelling stopped and restarted at the suspension of the therapy. The patient was offered surgical management and she underwent laparoscopy with excision of the nodule and umbilical reconstruction.

Techniques and instrumentation

Four months after diagnosis, on day 25 of her menstrual cycle, the patient was hospitalized and underwent surgery. Sub-costal access was gained by Verres-Palmer needle to obtain pneumoperitoneum and a 5 mm camera was introduced to explore the abdominal cavity. No lesions were observed in the peritoneum, with the exception of umbilical endometriosis localized to the abdominal wall. Three suprapubic 5 mm trocars were installed for the surgery. The pouch of Douglas was occupied by dense adhesions and the left ovary was adherent to the abdominal wall. Adhesiolysis was performed. The procedure revealed the presence of two endometriotic nodules on the left and the right utero sacral ligaments. Intraperitoneal, complete excision of these nodules was performed by coagulation and section of the tissues by a bipolar “Robi” device, scissors and Manhes grasping forceps (Karl Storz, Tuttlingen, Germany), based on the standard technique generally used in our department.

We made a V-like incision of the skin and the para-umbilical tissue, followed by a 30 mm large, deep, diamond-shaped excision of the endometriotic nodules. These lesions penetrated up to the rectus sheath and musculature of the abdominal wall. A 60 mm Prolene mesh was inserted to prevent future herniation. Histological examination of the specimen confirmed the
diagnosis of endometriosis and revealed the presence of endometrial glands and stroma. No epithelial atypia was detected (Fig. 3). No complications occurred during surgery and the operative time was 180 minutes. There were no post-operative complications and only analgesic treatment was administered to the patient.

DISCUSSION

Umbilical primary endometrioma is a rare extra-uterine localization of endometriosis with a documented neoplastic risk (5, 6). It is often difficult to distinguish primary umbilical endometriosis from other benign and malignant masses of the abdominal wall. Common differential diagnoses include benign nevus, lipoma, abscess, cyst, residual embryonic tissue, pyogenic granuloma, hernia, and cellulitis. Umbilical endometriosis may resemble a malignant melanoma, primary adenocarcinoma or the “sister Mary Joseph nodule”—a metastatic adenocarcinoma. While changes to radiological imaging during the menstrual cycle mask the presence of umbilical endometriosis, magnetic resonance imaging (MRI) has been shown to differentiate umbilical endometriosis from other pigmented skin lesions (7, 8).

However, MRI cannot confirm the presence of all endometrial implants in the peritoneum (9, 10). Therefore, in all patients with umbilical endometriosis, even if asymptomatic, we recommend a diagnostic laparoscopy to ensure a complete surgical approach. Progestins or GnRH analogues may be used in the medical management of umbilical endometriosis (11, 12). A diagnosis of endometriotic lesions is supported by a subsequent improvement in symptoms. Medical management prior to surgical intervention has been advocated (13). However, it is our experience that this can lead to incomplete excision of endometriotic lesions and an increased risk of recurrence if undertaking a subsequent surgical procedure. The timing of the surgery is important: we perform the operation directly prior to menstruation to better localize lesions (14).

Excision is the mainstay of treatment for this condition. The depth of endometriotic lesions can only be established surgically. Thus, patients should be made aware prior to surgery that it may be necessary to repair the rectus sheath with a mesh. There have been a small number of cases where a mesh repair has been performed, with no subsequent recurrence documented to date.

Unlike spontaneous endometriosis, scar endometriosis can be avoided by irrigation of surgical wounds prior to closure to reduce the risk of implantation of endometrial cells (15). Histology is the mainstay of diagnosis of cutaneous endometriosis. Endometrial glands and stroma are usually detected within a background of fibroadipose tissue. Immunohistochemical analysis reveals and progesterone receptors and is sometimes necessary to confirm the diagnosis (16).

CONCLUSIONS

Primary umbilical endometriosis is a very rare disease. Differential diagnosis is difficult because of other umbilical lesions of similar appearance. Complete excision is the mainstay of treatment for this condition. Diagnostic laparoscopy should be suggested in these patients.

Conflict of interest statement: None.

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Primary umbilical endometriosis