Abstract. Background: Endometriosis is the presence of functioning endometrial tissue outside the uterine cavity. Umbilical endometriosis has been reported in more than one hundred cases and the umbilicus represents the location of 0.5–1% of ectopic endometrioses. A correct differential diagnosis can be difficult and the use of epiluminescence and MRI is suggested for the accuracy of preoperative diagnosis. Patients and Methods: We report our experience in preoperative differential diagnosis and treatment of four cases of umbilical endometriosis. An accurate anamnesis and clinical examination together with the use of 13 MHz probe ultrasonography with echocolor duplex scan led us to an accurate pre-operative diagnosis of umbilical endometriosis. The surgical approach to umbilical endometriosis represented an important step in achieving a satisfying result. Results and Conclusion: In all treated cases, a hemi-circumferential incision in the inner aspect of the umbilical opening was performed allowing adequate access to the lesion and umbilical repositioning. At the follow-up visit (from 6 to 24 months after surgery), no symptoms or signs of recurrence were evidenced. A natural-looking umbilicus was observed in all cases with minimal visible scars.

Umbilical endometriosis has been reported in more than one hundred cases and was first described by Villar in 1886 (cited in 7). The umbilicus represents the location of 0.5–1% of ectopic endometrioses (8). Its clinical manifestation often consists of a dome-shaped, brown nodule, with intermittent pain and bleeding from the umbilicus during the menstrual period. A correct differential diagnosis can be difficult and some authors propose the use of epiluminescence microscopy (ELM) (9) and magnetic resonance imaging (MRI) (10,11) for the accuracy of preoperative diagnosis.

We report our experience in preoperative differential diagnosis and treatment of umbilical endometriosis.

Patients and Methods

Between January 2003 and March 2006, four Caucasian women with an umbilical mass presented to the Department of Plastic and Reconstructive Surgery of the University “La Sapienza” of Rome. The mean patient age was 37 years (from 27 to 43 years). All patients were of a fertile age and referred to the use of hormones for contraceptive purpose or endometrial pathology (Table I). They noted the presence of a growing mass surrounding the umbilicus, all associated with similar clinical characteristics. A common feature was that the symptoms worsened during the premenstrual period (Table I).

At clinical examination, it was possible to palpate well-defined lesions, with a dimension from 1x0.5 to 2x2 cm and with a tough, wooden consistence (Figure 1, Table I). Lesions were mostly adherent to deep layers and did not show any volume increase during Vasalva’s maneuver.

Differential clinical diagnosis was among umbilical endometriosis, umbilical hernia, cyst or tumour.

An ultrasound examination of the masses with a 13 MHz probe and echocolor duplex scan were performed in all cases. Common findings were the presence of a hyperechogenic structure with small hypoechochogenic spots inside and a predominant venous pattern (Table II). No connection to the deep fascial plane was noted. At this time, a strong preoperative diagnosis of umbilical endometriosis was suspected (Figure 2).

At surgery, an hemicircumferential skin incision in the inner aspect of the umbilical opening was performed. The lesions were easily dissected from the deep plane and sent for histological analysis. (Figure 3). When it was necessary to invert the umbilicus, its repositioning required the umbilical dermis to be attached to the abdominal rectus fascia by using single sutures at cardinal points of Poliglecaprone 25 (MONOCRYL® 25 4/0) (Table II).
Histology showed the presence of endometrial tissue in all samples (Table II), allowing the confirmation of umbilical endometriosis. At the follow-up visit (from 6 to 24 months after surgery), no symptoms or signs of recurrence were evidenced. A natural looking umbilicus was observed in all cases with minimal visible scars (Figure 4).

Results and Discussion

Endometriosis is a typical benign pathology of women of a fertile age, with the highest incidence in women between 30 and 45 years. We can distinguish an internal endometriosis in which ectopic cells are located in the myometrial tissue (also called adenomyosis) and an external or ectopic one with pelvic (most common) or extra-pelvic localization (also called extragenital) (12).

A number of theories have been put forward to explain its development. According to Sampson (13), it is due to the reflux of menstrual blood and the implantation of endometrial cells into the peritoneal surface of the pelvic organs. Based on this theory, it is difficult to explain the rare occurrence of endometriosis in the breast, lung or umbilicus (14). Other observers believe that endometriosis develops from pluripotent cells of the coelom (15). This theory explains the development of endometriosis wherever coelomic epithelium or vestige occurs (i.e. in the pelvis, umbilicus and hernial sacs). Furthermore, embolization by lymphatic vessels has been suggested. According to this theory, blue dye and other materials can migrate from the pelvis to the umbilicus by retrograde lymphatic flow. No single theory can explain the spontaneous occurrence of endometriosis in all affected sites.

Cutaneous endometriosis can be divided into a spontaneous form and an iatrogenic one, related to the presence of surgical scars for abdominal-pelvic surgery. Umbilical endometriosis is rare, having an estimated incidence of 0.5-1.0% of all cases of endometriac ectopia. More commonly, cutaneous endometriosis occurs in a surgical scar from abdominal or pelvic procedures, which include hysterectomy, caesarean sections, episiotomy and laparoscopy (16, 17).

Umbilical endometriosis may appear during active menstrual life as a small, firm, bluish-pink mass of the umbilical area, with a diameter varying from a few millimetres to 6 cm. The endometrioma may cause pain, swelling or tenderness mainly in the premenstrual period, as this endometrial tissue responds cyclically but not consistently, to the ovarian hormones. Sometimes secretion

Table I. Overview of the clinical history and findings of patients.

<table>
<thead>
<tr>
<th>Case</th>
<th>Age (years)</th>
<th>Pathological history</th>
<th>Oral contraceptive use</th>
<th>Development / Growth</th>
<th>Symptoms out of the premenstrual period</th>
<th>Symptoms during premenstrual period</th>
<th>Dimensions out of the premenstrual period (cm)</th>
<th>Colour</th>
<th>Shape</th>
<th>Consistency</th>
<th>Adhesion to deep layers</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>42</td>
<td>Twin birth at 24 years</td>
<td>Assumed for 16 years and suspended 1 year before manifestation</td>
<td>1 Year / gradual</td>
<td>None</td>
<td>Volume increase, bleeding during menses</td>
<td>2x2</td>
<td>Red-bluish</td>
<td>Hemi-spherical</td>
<td>Strong</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>36</td>
<td>Spontaneous miscarriage in the 2nd month at age 22 years; caesarean section at age 25 years; diffuse uterine fibromatosis</td>
<td>Assumed for 3 years from age 25 to age 28 years</td>
<td>1 Year / gradual</td>
<td>Tenderness, pain</td>
<td>Pain and tenderness worsening</td>
<td>1x0.5</td>
<td>Bluish</td>
<td>Nodular</td>
<td>Wooden</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>43</td>
<td>1 vaginal birth at age 31 years; 1 caesarean section at age 25 years; diffuse uterine fibromatosis</td>
<td>Estrogen-progestin therapy for endometrial cyst 4 years before</td>
<td>3 Months / rapid</td>
<td>Abdominal swelling, sense of tension and weight</td>
<td>Symptoms worsening</td>
<td>1x0.5</td>
<td>Bluish</td>
<td>Nodular</td>
<td>Tough-wooden</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>27</td>
<td>None</td>
<td>On course, assumed for 4 years</td>
<td>5 Months / rapid</td>
<td>Abdominal swelling, tenderness and pain</td>
<td>Symptoms worsening</td>
<td>1x1</td>
<td>Normal Round</td>
<td>Slightly tough</td>
<td>Strong</td>
<td></td>
</tr>
</tbody>
</table>

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or some bleeding may occur through the skin, concomitant with menses; for this reason, it is often called the “menstruating tumour”. On examination, a tender indurated umbilicus containing cystic structures, which exude old blood on pressure, is pathognomonic. Umbilical endometriosis frequently exists as a solitary lesion without accompanying pelvic disease (18).

The differential diagnosis of an umbilical lesion includes hernia, primary and metastatic neoplasms, embryological residual mass and various granulomas (e.g. pyogenic, foreign body). MRI and epiluminescence are described in the literature to aid in recognizing umbilical endometriosis before surgical extirpation and histological control (9, 10). Endometriomas appear as homogeneously hyperintense on T1-weighted sequences. They often demonstrate shading (i.e. loss of signal within the lesion) which can be seen on T2-weighted images. This shading reflects the chronic nature of an endometrioma and helps differentiate it from other blood-containing lesions. Yu et al. described how the MRI features of umbilical endometriosis were helpful to distinguish an umbilical mass secondary to endometriosis from other more sinister causes (11).

Other authors used ELM to obtain a correct differential diagnosis with cutaneous malignant neoplasms (melanoma, basal cell and squamous cell carcinomas) (9). But ELM did not reveal the eventual relationship with deeper planes, so excluding such a sinister diagnosis, needing to be accurately studied pre-operatively with other instrumental analyses.

The cases of umbilical endometriosis observed here had typical clinical manifestations. The use of a low cost and non-invasive instrumental analysis such as ultrasonography with echocolor duplex scan was exhaustive to strongly suggest the diagnosis of umbilical endometriosis. This achievement was due to the fact that, nowadays, ultrasonography can be carried

<table>
<thead>
<tr>
<th>Case</th>
<th>Ultrasonography plus echo-colour duplex</th>
<th>Macroscopical findings at surgery</th>
<th>Umbilical reconstruction</th>
<th>Histology evolution</th>
<th>Post op.</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Venous dominant pattern; hyperechogenic vessels with hypoechoic spots inside the mass. Size: 17x13x19 mm. Exclusive umbilical localization, absence of connections with deep tissues and constant growth pattern.</td>
<td>Inferior hemicircumferential incision. Trunk-conic shape lesion with wide implantation base on the inferior portion of the umbilical pedicle requiring its incision.</td>
<td>Repositioning of umbilical stalk to linea alba with interrupted poliglecaprone 25 4/0 sutures. Umbilical dermis attached to abdominal rectus fascia with poliglecaprone 25 4/0 single sutures at cardinal points. Direct skin closure with 4/0 nylon and compressive dressing.</td>
<td>Presence of endometrial glands in a context of fibrotic and chronically inflamed deep dermis.</td>
<td>No symptoms or signs of recurrence 6 months later.</td>
</tr>
<tr>
<td>2</td>
<td>Hyperechogenic pattern of the mass with some hypoechoic areas inside; few vascular spots in the peripheral areas and a venous dominance structure. Size: 9x5x8 mm. No connections with deep tissues.</td>
<td>Lateral hemicircumferential incision. Nodular lesion without connections to deep structures.</td>
<td>Direct skin closure with 4/0 nylon and compressive dressing.</td>
<td>Tissue resembling endometrium with typical glands and stroma in a fibrotic middle dermis.</td>
<td>No symptoms or signs of recurrence 11 months later.</td>
</tr>
<tr>
<td>3</td>
<td>Hyperechogenic nodule with hypoechoic spots inside and microcalcifications. Typical vascular features characterized by a venous dominance with slow venous flow after digital pressure. Size: 10x6x9 mm. No relationship with deep layers.</td>
<td>Lateral hemicircumferential incision. Nodular lesion without connections to deep structures.</td>
<td>Umbilical dermis attached to abdominal rectus fascia with poliglecaprone 25 4/0 single sutures at cardinal points. Direct skin closure with 4/0 nylon and compressive dressing.</td>
<td>Tissue resembling endometrium with typical glands and stroma</td>
<td>No symptoms or signs of recurrence 11 months later.</td>
</tr>
<tr>
<td>4</td>
<td>Hyperechogenic structure with small hypoechoic spots inside. Presence of venous flow increased by digital pressure around the lesion. No connections to the deep fasci muscular plane.</td>
<td>Lateral hemicircumferential incision. Nodular mass protruding above linea alba.</td>
<td>Umbilical dermis attached to abdominal rectus fascia with poliglecaprone 25 4/0 single sutures at cardinal points. Direct skin closure with 4/0 nylon and compressive dressing.</td>
<td>Endometrial tissue in a fibrotic stroma.</td>
<td>No symptoms or signs of recurrence 2 years later.</td>
</tr>
</tbody>
</table>
out at high definition, increasing its accuracy in the diagnosis of various pathologies. In the observed cases of umbilical endometriosis, recurrent findings were the presence of a hyper-echogenic structure with small hypoechogenic spots inside, a predominant venous pattern, and the absence of connections to the deep fascial plane. The spontaneous dominant venous flow is a characteristic of benign tumours (19). The absence of continuity with the deep fascial plane allows its differential diagnosis from invasive malignancies and hernias (20, 21).

Based on our experience, we believe that the combination of the typical clinical manifestation, in particular the symptoms worsening during the premenstrual period, together with the aforementioned echographic findings...
strongly points to a diagnosis of umbilical endometriosis. Therefore, the use of more expensive and accurate examinations, such as MRI, are not required and should be confined to atypical and uncertain cases of umbilical masses.

Another important point in the management of umbilical endometriosis is the surgical approach. Umbilical repositioning is mandatory and the surgeon should aim for the removal of such benign lesions with minimal visible scars and a natural-looking result. In all the cases treated here, a hemi-circumferential incision in the inner aspect of the umbilical opening was performed. This allowed adequate access for the lesion to be excised and an adequate umbilical repositioning. During this procedure, it is important to obtain an inverted umbilicus and to use, when required, internal absorbable sutures between the umbilical dermis and the abdominal rectus fascia to guarantee umbilical inversion.

Conclusion

The combination of clinical features of typical umbilical endometriosis and distinctive ultrasonography findings allowed an accurate preoperative diagnosis of umbilical endometriosis. We consider these preoperative findings indicative and consistent enough to make accurate preoperative diagnoses of umbilical endometriosis.

In the treatment of such benign lesions, umbilical repositioning with a minimal visible scar is mandatory to obtain a natural-looking umbilicus and a satisfying result.

References


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