Primary umbilical endometriosis

Endometriose umbilical primária

Paulo Vicente dos Santos Filho, TCBC-SE1; Marcelo Protásio dos Santos1; Samanta Castro1; Valdaldo Aragão de Melo, ECBC-SE1

ABSTRACT

Objective: to report the characteristics, evolution and outcome of patients with primary umbilical endometriosis. Methods: an observational and descriptive study of patients with primary umbilical endometriosis diagnosed between 2014 and 2017. The clinical variables evaluated were age, clinical picture, lesion characteristics, diagnostic methods, treatment and recurrence. Results: six patients diagnosed with primary umbilical endometriosis aged 28 to 45 years were operated on during the study period. They had lesions ranging from one to 2.5 cm in diameter, violet in five patients and erythematous-violaceous in one. The duration of the symptoms until diagnosis ranged from one to three years and in all the cases studied the diagnosis was made through the clinical manifestations and confirmed by histopathological analysis. No case was associated with neoplastic alterations. All patients evaluated had pain and umbilical bleeding in the menstrual period. Conclusion: umbilical endometriosis is an uncommon disease and should be included in the differential diagnosis of women as umbilical nodules. The treatment of choice is the total excision of the lesion.

Keywords: Endometriosis/surgery. Umbilicus. General Surgery.

INTRODUCTION

Endometriosis is a benign, estrogen-dependent entity, common in the gynecological clinic and associated with complaints of pelvic pain and infertility, affecting 6% to 10% of women in reproductive age. The etiopathogenesis of the disease does not comprise a single explanation in the literature. Among the classical theories, endometriosis is attributed to retrograde menstrual flow, metaplastic transformation or even iatrogenic deposition in surgical procedures1.

It is a disease that can affect several organs, such as the pelvic peritoneum, fallopian tubes, ovaries, subcutaneous tissue, Umbilicus, urinary tract, bladder, heart, kidney, lung, liver, pancreas, muscles, central nervous system, among others, which makes it a multi-systemic disease1,2. Endometriotic lesions are more frequent in the peritoneum and pelvic organs, especially in the ovaries, followed by the recto-vaginal septum. It is found less frequently in extra-pelvic regions, such as gastrointestinal (sigmoid, rectum, ileocecal and appendix) and urinary tract, extremities, subcutaneous tissue and abdominal wall2.

Cutaneous endometriosis is rare, but it is the most common extra-pelvic location, being classified as to its origin in primary and secondary forms. The secondary forms, which are the most common, are represented by the cases that arise on previous surgical scars of hysterectomies, cesarean sections, laparotomies and episiotomies. Primary or spontaneous forms are much rarer and may be located in the umbilical scar, in the perianal or inguinal region, unrelated to previous surgeries. Secondary umbilical endometriosis can affect up to 1% of women undergoing cesarean section, but their occurrence as a primary disease in patients without previous surgeries begins to gain space in the literature, although there are still few publications on the subject3.

The objective of this study is to evaluate the clinical and epidemiological characteristics of patients with primary umbilical endometriosis, as well as the treatment employed, with a bibliographic review on the
subject, since in the world medical literature only one study has a series similar to ours, the remaining being case reports and literature reviews.

**METHODS**

This is a descriptive, retrospective observational study performed at the General Surgery Service of the Medical Residency of the Charitable Foundation Surgery Hospital (FBHC) from January 2014 to December 2017. We included patients with a history of tumor in the umbilical scar associated with pain and bleeding during the menstrual period, without previous abdominal surgeries, submitted to surgical treatment and with histopathological diagnosis of umbilical endometriosis. We analyzed age, duration of symptoms, presence of infertility, intestinal complaints, weight, diagnostic method, nodule characteristics, type of treatment employed, postoperative evolution and complications.

All the patients authorized, through the Informed Consent Term, the use of information contained in their respective medical records and of the photographic records made in the pre, intra and immediate and late postoperative periods.

We performed onfalectomy with a safety margin of 1cm in all patients. We removed the umbilical scar en bloc, including the nodule, underlying aponeurosis and a small segment of rectus abdominis muscle, to prevent recurrence and rule out the possibility of malignancy. After hemostasis, we repaired the aponeurosis, and fixated the periumbilical skin. We carried out the skin synthesis with simple stitches of absorbable suture. We performed the procedures under sedation and local anesthesia.

The pathological diagnosis of endometriosis was considered when there was identification of endometrial glands, separated by endometrial stroma, within fibrous connective tissue, with areas of focal hemorrhage and chronic inflammatory process, and presence of macrophages with hemosiderin pigments.

This research was approved by the Institutional Ethics Committee with the following reference number: 1223849199.

**RESULTS**

During the study period, were admitted eight patients with umbilical scar tumor associated with bleeding and/or umbilical pain in the menstrual period, with clinical diagnosis of umbilical endometriosis (Figures 1, 2 and 3). Two of them were classified as secondary umbilical endometriosis and were not included in the study because they had previous abdominal surgeries (one case by exploratory laparotomy due to a stab wound and the appearance of umbilical and abdominal wall endometriosis a year a half later, and another due to previous cesarean section and umbilical endometriosis three years later). We classified the six other patients as having primary umbilical endometriosis and included them in the study.

![Figure 1. Umbilical endometrioma: brownish nodules in umbilical region, with progressive growth and symptomatology, more pronounced during menstrual period.](image-url)
The main complaint was pain and umbilical bleeding in the menstrual period associated with umbilical tumor (100%). Age ranged from 28 to 45 years (mean 33). The size of the lesions ranged from 1.0 to 2.5 cm (mean 1.9cm), with violaceous color in five (83%) and erythematous-violaceous in one (16%). The duration of symptoms varied from one to three years (mean 13 months). The diagnosis was clinical in all cases (100%), and the histopathological confirmation showed no malignancy in the samples evaluated.

One patient had associated umbilical hernia and underwent umbilical herniorrhaphy and the standardized resection of umbilical endometrioma. In the patients’ follow-up, one case of superficial surgical site infection (16%) occurred as a surgical complication. One patient had a new episode of bleeding on the 30th postoperative day, during the menstrual cycle, but at a lower intensity than before surgery, without new reports of bleeding in a one-year follow-up. All patients had been given birth to children by vaginal delivery. We followed the patients studied for a period of one to two years, without reports of relapses and with a good aesthetic aspect of the surgical scars. As routine, we referred all patients studied for joint follow-up with the Gynecology Service.

DISCUSSION

Extra-pelvic endometriosis can compromise multiple organs. The most frequent sites, in descending order, are intestine, skin (including the umbilical and abdominal scars), inguinal region and thigh, lungs and pleura, pancreas, meninges and vertebrae. In general, cutaneous and subcutaneous involvement is secondary to a cicatricial process following abdominal and/or pelvic surgical procedures, such as laparoscopy/laparotomy, cesarean section, hysterectomy, myomectomy, episiotomy, appendectomy, removal of Bartholin’s gland cyst, amniocentesis and intrauterine injections for abortion. Less than 30% of cases of cutaneous endometriosis appear in the absence of surgery and are referred to as primary or spontaneous cutaneous endometriosis.

Despite its low incidence, the umbilical scar is the most common primary site, as reported in the six cases. Other primary cutaneous sites include the vulva, perineum, inguinal region, and extremities. It can also develop during pregnancy, more frequently in the umbilical region, and may spontaneously regress after delivery.
Cutaneous endometriosis of the umbilical scar manifests as a firm consistency nodule measuring from 0.5 to 2.5 cm, varying in color from bluish-black to intense red, brown or purpura depending on the amount of hemorrhage and the depth of penetration of ectopic endometrial tissue. Occasionally, the nodule is skin-colored\textsuperscript{1,7,9}. It is generally single, often multilobulated, although multiple discrete nodules may be present\textsuperscript{6}. Clinical symptoms include pain, hyperesthesia, bleeding, edema and growth correlated with the menstrual cycle\textsuperscript{1,7,9,10}. However, all symptoms are rarely present, and it may even be asymptomatic\textsuperscript{7}. Hemorrhage related to menstrual bleeding is absent in most cases\textsuperscript{8,6,10} according to the literature, but, in our series, all the patients presented bleeding. In our study, all the patients presented cyclic pain during the menstrual period. There were no reports of acute pain requiring emergency therapy, and pelvic pain was present in all cases. Higher degrees of endometriosis may be associated with an increased incidence of pain, but pain severity may be related to factors such as depth of infiltration and local and systemic inflammatory mediators involved\textsuperscript{10}. Victory et al., in their literature review on the subject, showed that bleeding was present in less than 50% of women with umbilical endometriosis, and usually occurred in response to the hormonal changes present during the menstrual cycle\textsuperscript{6}. They also showed a statistical association, but not clinically discernible, between the mean lesion size and the presence or not of bleeding\textsuperscript{6}. The authors also state that due to the lack of general agreement in reporting lesion size, a one-dimensional mean size was calculated from the measures reported, in the several cases reviewed, which resulted in an average size of 2.29 ± 0.2 cm. In the medical literature, the size of the umbilical nodule ranged from 0.5 to 4.0 cm in diameter, generating an average of 2.4cm. Regarding the color of the lesions, Victory et al.\textsuperscript{6} showed that the majority of the patients presented brown lesions, followed by blue, purple (violet), black and red. In our series, all lesions had similar colors, the violaceous ones predominating.

The mean age of the patients was 33 years, which is compatible with the premenopausal phase and with other series and reports described in the literature. A little different, Romera-Barba et al. presented a series of six patients with a mean age of 39.1 years, demonstrating that the disease occurs after prolonged exposure to the metaplastic and environmental factors that catalyze the development of umbilical endometriosis\textsuperscript{4}. Gynecological symptoms such as dysmenorrhea, dyspareunia, infertility and menstrual irregularities are generally present in pelvic endometriosis and absent in skin endometriosis\textsuperscript{3}. Our patients only reported dysmenorrhea. It is important to emphasize that all but one had children, since there is an intimate relation of infertility and pelvic endometriosis. Some authors use laparoscopy during resection of the umbilical lymph node to search for pelvic endometriosis, and most of the time cauterization of ectopic foci is performed. When the patient presents with infertility and exacerbated pelvic symptoms, laparoscopy is mandatory\textsuperscript{6}, which was not necessary in any of our cases.

The most common differential diagnoses of umbilical endometriosis include pyogenic granuloma, hernia, and pemphigus vegetans. Because of the variable macroscopic appearance, these lesions may initially be confused with malignant tumors, such as melanoma\textsuperscript{11}. One of our patients had an umbilical hernia associated with the nodule, only diagnosed intraoperatively. This condition is even rarer. Stojanovic et al. published, in 2014, a case of a patient with primary umbilical endometriosis associated with a large irreducible umbilical hernia\textsuperscript{11}.

The diagnosis of umbilical scar endometriosis is relatively easy. The suspicion is based initially on the medical history and physical examination. The complementary propaedeutics aims to offer subsidies for the best therapeutic option, and it is important to remember that in the case of umbilical scar endometriosis, the clinic is sovereign\textsuperscript{1,2,6,11}. All of our patients had a typical history and physical examination, and did not require complementary tests for diagnosis. Despite this, for some patients it took up to three years for the correct diagnosis after the onset of symptoms. The definitive diagnosis, however, is made only by the histological study of the nodule after its exeresis\textsuperscript{1,2,6}. In the cutaneous lesions, one can observe irregular, circular, elongated or angular glandular spaces in the reticular dermis or hypodermis,
surrounded by highly vascular and cellular stroma similar to that of the functioning endometrium. The histological aspect corresponds to the uterine endometrium in the proliferative and secretory phases.

Regarding treatment, we used the algorithm adopted in our Service for the management of patients with umbilical endometriosis (Figure 4). Simple surgical excision is the choice, and should be broad to ensure complete cure. Previous hormonal treatment may be an option for larger tumors, and may reduce their size before surgery. This was not necessary in our cases. We indicated surgical treatment for all patients, since the lesions were candidates for total resection. Omphalectomy was necessary in all cases. We do not deem it necessary to perform laparoscopy to investigate abdominal foci as proposed by some authors. In our view, in addition to higher hospital costs, such a routine can also bring greater risks inherent to the method. We reserve intraoperative laparoscopy for cases with great suspicion of pelvic endometriosis based on the clinical history or in those with an already performed imaging diagnosis. As our patients did not present preoperative complaints of intestinal cramps, diarrhea, infertility or atypical pelvic pain, we did not indicate laparoscopy. We performed resection of the lesion with sedation and local anesthesia. Local anesthesia has benefits in the prevention of postoperative pain, lower incidence of nausea, vomiting and urinary retention. Another important aspect to be evaluated is hospital costs. When we consider only the material and drugs used in anesthetic procedures, the use of local anesthesia with intravenous sedation has obvious advantages. In our routine patients were discharged on the same day. The surgical technique used is always the same, respecting the safety margins. In some cases, especially in those where nodulation distorts the umbilical architecture, omphalectomy is necessary, a fact observed in all our patients, who required umbilical reconstruction after the surgical procedure for better aesthetic appearance.

![Figure 4](image-url)

**Figure 4.** Algorithm adopted in our Service for the management of patients with umbilical endometriosis.

The prognosis of umbilical endometriosis is good. Recurrences are unusual. However, malignant transformation has been reported in percentages ranging from 0.3 to 1.0% and should be suspected in recurrent...
or fast-growing lesions. In none of our cases was there malignancy. Our patients did not present recurrence until now. Among the cases described in the literature, only one patient presented recurrence after four years, which is why a follow-up should be continued for a longer period of time.

Umbilical endometriosis is, therefore, a rare condition, but should be considered in the differential diagnosis in cases of umbilical scar lesions in women of childbearing age, even if they are asymptomatic. The definitive diagnosis is histopathological, and surgical excision is the treatment of choice.

RESUMO

Objetivo: relatar as características, evolução e desfecho de pacientes portadoras de endometriose umbilical primária. Métodos: estudo observacional e descritivo de pacientes portadoras de endometriose umbilical primária diagnosticada entre 2014 e 2017. As variáveis clínicas avaliadas foram: idade, quadro clínico, características das lesões, métodos diagnósticos, tratamento e recidiva. Resultados: seis pacientes com diagnóstico de endometriose umbilical primária, com idades entre 28 e 45 anos foram operadas no período do estudo. Elas apresentavam lesões que variavam de 1,0cm a 2,5cm de diâmetro, de cor violácea em cinco pacientes e eritemato-violácea em uma. O tempo de duração dos sintomas até o diagnóstico variou de um a três anos e em todos os casos estudados o diagnóstico foi feito por meio das manifestações clínicas e confirmado por meio da análise histopatológica. Nenhum caso foi associado com alterações neoplásicas. Todas as pacientes avaliadas apresentavam como manifestação clínica dor e sangramento umbilical no período menstrual. Conclusão: a endometriose umbilical é uma doença pouco frequente e deve ser incluída no diagnostico diferencial de mulheres como nóculo umbilical. O tratamento de eleição é a exérese total da lesão.


REFERENCES


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Mailing address:
Paulo Vicente Filho
E-mail: paulovicentefilho@gmail.com /
marcelo_protasio@yahoo.com.br