Case Report

Umbilical endometriosis: a case report and review of the literature

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INTRODUCTION

Endometriosis was first described by Rokitansky in 1860, defined as the presence of functional endometrial tissue outside the uterine cavity. Depending on the location, it can be classified as pelvic and extra pelvic endometriosis, the last one represents only 0.9 to 1.5% of all cases. Umbilical endometriosis was first described by Villar in 1886 and since then this condition has been called Villar's nodule. Umbilical endometriosis can be primary or secondary. The primary form is rare and there are very few cases reported in the literature. The secondary form usually occurs on scars from laparotomies, caesarean sections, hernia repair and laparocopies, in which there was exposure of endometrial tissue and contact with the umbilical scar. According to the literature, until 2012 only 109 cases of umbilical endometriosis had been reported worldwide. It has an estimated incidence of 0.5-1% of all patients with extragenital endometriosis and this percentage includes both secondary scar-related and spontaneous primary forms.

Authors describe a case of a 32 years old female who presented with secondary umbilical endometriosis who underwent surgery and histopathological diagnosis of umbilical endometrioma was made.

CASE REPORT

Authors present the case of a 32 years old female with the following surgical history; 2 cesarean sections, 1 uterine curettage and surgical repair of umbilical hernia without mesh 2 years ago. She presented to the outpatient clinic of the general surgery department with the presence of 1 nodular lesion at the level of the umbilical scar that gradually increased in size, the lesion was formed 1 year after the umbilical hernia repair, it was accompanied by pain and bloody discharge starting just prior to her menstruation. Physical examination revealed a superficial hyper-pigmented dark reddish nodule located in the umbilicus of approximately 1.5 cm. The nodule was not painful at palpation, was irreducible by gentle pressure and had bloody discharge at the time of exploration. (Figure 1).
Abdominal ultrasound was performed and found that at the level of the umbilical scar, an occupational lesion of irregular morphology, poorly defined contours, heterogeneous predominantly hypoechoic with measurements of 22×16×20 millimeters, without vascularity on Doppler application was observed. During the Valsalva maneuver, no hernial protrusions were observed. Findings suggestive of an endometriotic implant at the umbilical level.

Laboratory results revealed leukocytes 6.4×10^3/μL with 50.8% granulocytes, Hb was 14.3 gm.

The following surgical findings were found: a circumferential incision is made at 1 cm from the lesion in the umbilicus, subcutaneous cellular tissue is dissected up to the aponeurosis and a tumor measuring 2×2×2 centimeters was extracted (Figure 3).

Histopathological examination of the excised tissue revealed fibro-adipose connective tissue with widespread prevalence of multiple endometrial glandular tubes and surrounding endometrial stroma. Concluding, the findings were compatible with diagnosis of umbilical endometriosis (Figure 4).

The post-surgical evolution was satisfactory, without pain or data of immediate complications. The patient was discharged from the hospital the same day. She comes to an evaluation 2 weeks later without complications.
The patient was referred to the outpatient clinic of the department of obstetrics and gynecology for evaluation and potential further consultation.

**DISCUSSION**

Endometriosis is defined as the presence of endometrial glands and stroma outside the uterus. This ectopic finding affects 7-10% of the women of reproductive age and commonly occurs in the pelvic organs. Furthermore, ectopic endometrium occurs in the abdominal wall in 0.03-1.08% of women. An umbilical endometrioma is a rare condition with an estimated incidence of 0.5-1% in all patients with endometrial ectopia.

Primary umbilical endometriosis is a rare subtype of spontaneous occurring cutaneous endometriosis with no previous scar. Secondary umbilical endometriosis can occur following laparoscopic or other surgical procedures involving the umbilicus.

The pathogenesis of endometriosis has been a much-debated issue. According to the hypothesis proposed by Sampson in 1920 endometriosis was caused by retrograde menstruation passing thorough the fallopian tube in the pelvis. However, there are other theories such as coelomic metaplasia, direct spread, iatrogenic spread, lymphatic or hematogenous spread. The theory of lymphatic and hematogenic transplantation is suggested for cases of umbilical endometriosis with pelvic endometriosis. But the disease may occur through metaplasia of urachus residues in a case of isolated umbilical endometriosis. Secondary umbilical endometriosis may occur through iatrogenic spread of endometrial cells after operations.

Umbilical endometriosis presents in the form of a nodule of variable size, whose color varies from red to blue or black. The clinic usually has a cyclical character with pain or increase in volume of the nodule during the menstrual period. Other patients describe a firm, pigmented or bluish nodule with pain and tenderness associated with cyclic bleeding or discharge from the umbilicus during menses.

Ultrasound, computed tomography or magnetic resonance imaging help assess the extent and establish the differential diagnosis. Ultrasound can be used to assess the nodule size and involvement of surrounding tissues and to evaluate other pelvic pathology, hence aiding the planning of surgical management. MRI can also be used as a method of preoperative evaluation in suspected endometriosis. MRI features of an umbilical endometriosis includes a homogeneous hypointense lesion on T1-weighted sequence with low signal on T2 weighting.

The definitive diagnosis is histopathological. The histologic diagnosis of endometrioma requires two of the three following features that is endometrial-like glands, endometrial stroma or hemosiderin pigment.

The differential diagnosis includes lesions that may appear in the umbilical area such as granulomas, keloid scarring from previous surgery, hernias, urachal cyst, dermal nevus, abdominal wall tumors and metastases of gastrointestinal neoplasia (Sister Mary Joseph nodule).

There are two treatment options for umbilical endometriosis, the medical treatment consists of the administration of analgesics and hormonal suppression with GnRH analogues, however this method presents reactivation and recurrence of the symptoms frequently.

Surgical treatment is the treatment of choice for most authors due to its low recurrence. Resection with adequate margins. The endometrioma and between 5 mm and 1 cm of adjacent healthy tissue must be included.

**CONCLUSION**

This case represents a presentation of secondary endometrioma, confirmatory diagnosis was made by histopathological study. The clinical presentation of umbilical swelling, cyclical pain and sometimes bleeding from the lesion are highly suggestive of this condition.

However, due to the variety of differential diagnoses, it is possible that specialists in other areas, apart from gynecologists, could find this pathology, more frequently general surgeons. Because it is a rare pathology in the area of general surgery, it is often confused with other clinical entities with a higher prevalence in surgery such as granulomas, incarcerated hernias. Therefore, umbilical endometriosis should always be considered in women of childbearing age with umbilical pathology. The most common form of presentation is a purplish nodule associated with pain that coincides with the menstrual period.

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