CASE REPORT

Primary Umbilical Endometriosis: Case Report with Literature Review

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ABSTRACT

Villars nodule, also known as umbilical endometriosis, is a rare umbilical disorder, usually affecting women of child bearing age. Endometriosis is a functional endometrial tissue found outside the uterine cavity, which affects the umbilicus in 0.5–1% of all extragenital sites. Correct diagnosis is challenging and commonly missed, but once a diagnosis has been made, the recommended management is surgical excision. We are reporting a case of primary umbilical endometriosis in a 31-year-old nulliparous patient, who presented with an umbilical mass of one year duration. Histopathology confirmed the diagnosis of endometriosis and the mass was widely excised.

Key words: Endometriosis, primary, umbilical

ملخص البحث:

تعتبر بطانة الرحم المهاجره التي تصيب السرة من التغيرات النادرة والتي قد تعاني منها السيدات صغيري السن، وهي نسيج وظيفي فعال خارج الرحم، وقد يتواجد في السره بنسبة (0.5% - 1%) من بين المواقع الغير تناسلية. يصعب تشخيصها أحيانا ويكمن علاجها جراحيا. يعرض الباحثان حالة لسيدة تبلغ من العمر 31 سنة تعاني من ورم في منطقة السرة أجريت للمريضة عملية لاستئصال الكتلة المتواجدة في السره واظهر الفحص النسيجي وجود بطانة رحمية.

INTRODUCTION

Endometriosis is a common benign gynecological disease defined as the presence of functional endometrial tissue (glands and stroma) outside the uterine cavity.[1] Among women of reproductive age, it has a prevalence of 3–10%.[2] Endometriosis mostly affects the pelvic organs, including the ovaries, fallopian tubes and uterosacral ligament. Its clinical manifestations include pelvic pain, dysmenorrhea, dyspareunia, infertility and intestinal or urinary symptoms. Although different theories have been postulated to elucidate the pathogenesis of this condition, none of them has been established. Extrapelvic endometriosis is less common and has been described in almost every area of the body, including the bowel, bladder, brain and lungs.[1] Umbilicus is a rare site of involvement representing 0.5–1% of extragenital endometriosis cases. It more commonly occurs secondary to surgical intervention, but rarely presents spontaneously as a primary umbilical endometriosis.[3] We report a rare case of primary umbilical endometriosis in a nulliparous female patient.

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CASE REPORT

A 31-year-old single female patient presented to the outpatient department with an umbilical swelling of 1-year duration. She stated that the swelling was slowly increasing in size and was associated with intermittent mild pain. There was no local cyclic bleeding or pain during menses, which had been regular, but accompanied by dysmenorrhea during the preceding year. The patient's medical and surgical history was unremarkable and she was not taking any form of hormonal therapy.

The physical examination revealed a firm mass (3 cm × 2.5 cm × 1.5 cm) located in the umbilicus, covered with brown corrugated skin. It was tender on palpation with no discharge [Figure 1]. The mass was irreducible with negative cough impulse and there were no other abdominal masses, regional lymphadenopathy, or hepatosplenomegaly.

The laboratory investigations were within normal ranges. Abdominal and pelvic ultrasound revealed a 3 cm × 3 cm heterogeneous hypoechoic lesion confined to the umbilical region with no intra-abdominal or pelvic extensions.

A decision was taken to excise the mass under local anesthesia. A periumbilical incision was made and the subcutaneous lobulated mass was exposed, causing a discharge of a thick brown material. The mass was widely excised with subsequent umbilical reconstruction. The patient was discharged on the same day of surgery. The histopathologic evaluation of the excised mass revealed hemorrhagic endometrial tissue composed of endometrial glands and stroma, consistent with endometriosis [Figure 2]. The patient was re-evaluated 6 months after the surgery and no signs of recurrence were evident.

DISCUSSION

The first case of umbilical endometriosis was first described by Villar in 1886 and has been named as Villar's nodule.[2] Since the first report, more than 100 cases have been reported.[3] Different theories regarding the pathogenesis of extraperitoneal endometriosis have been proposed, however none of them have been proven. Proposed theories include coelomic metaplasia, lymphatic or vascular invasion, genetic predisposition, immunological defects and retrograde menstruation. The umbilicus is a rare location of endometriosis, presenting a prevalence of <1% of all endometriosis locations.[4] Umbilical endometriosis is more commonly associated with surgical intervention, even if the surgery did not involve the umbilicus, 57% of the cases were associated with cesarean section and 11% with hysterectomy.[5,6] However, primary umbilical endometriosis is less common.[2]

In a review of Victory *et al*., the average size of umbilical endometriosis lesions was 2.3 ± 0.2 cm in diameter, 19% were predominantly brown, 13% were blue and 10% were purple. The mean age of affected females at the time of diagnosis was 37.7. These patients presented with swelling (88%), pain (77.9%) and cyclic bleeding (47%). Surprisingly, most patients (73%) had no history of pelvic endometriosis.[7]

The diagnosis of umbilical endometriosis is primarily clinical and the typical symptoms include the presence of a bluish mass in the umbilicus that bleeds or becomes painful or swollen during menses. Magnetic resonance

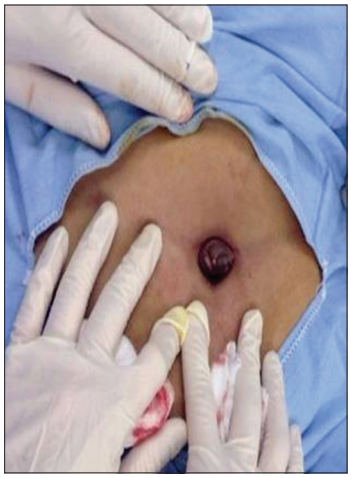

Figure 1: Gross picture of the umbilical mass

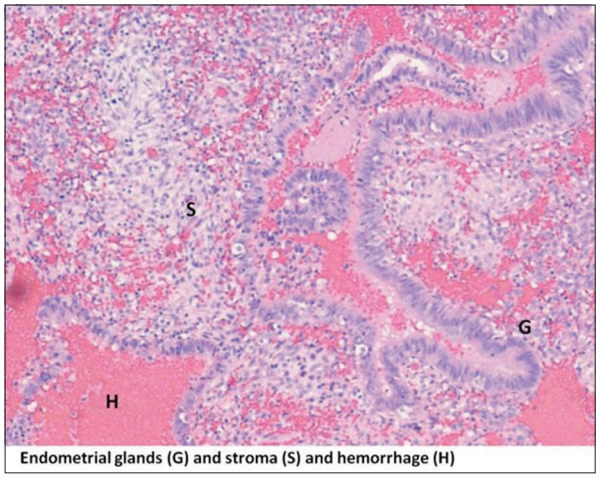

Figure 2: Microscopic view of the umbilical mass

imaging and excisional biopsy offer an accurate modality of diagnosis. However, the yield obtained by fine needle aspiration biopsy can show questionable results.[8] Differentiating between umbilical endometriosis and other umbilical nodules is quite difficult as evidenced by published data, which have shown that 20–50% of umbilical endometriosis cases have been misdiagnosed.[2] Differential diagnoses of umbilical nodules include keloid, pyogenic or foreign body granuloma, umbilical or paraumbilical hernia, benign or malignant skin tumors and metastatic adenocarcinoma, more commonly known as Sister Mary Joseph nodules.

The management of umbilical endometriosis is not yet standardized, most probably due to the rarity of the disease. In a review of Victory *et al*., almost 70% of patients required surgical management. Surgical procedure options include complete umbilical resection or local resection of the endometriotic mass sparing the umbilicus.

The most frequently performed procedure is complete resection of the umbilicus, especially if the mass is relatively large.[8] However, if the patient prefers to keep the integrity of the umbilicus, local resection of the mass sparing the umbilicus is performed with excision of a rim of normal surrounding tissue to decrease the likelihood of recurrence. Medical management using progesterone, danazol, norethisterone and GnRh analogs are not reliable, although some studies have reported a decrease in the mass size and relative relief of the associated symptoms.[8] Therefore, surgical management is the preferred management of umbilical endometriosis for the following reasons:

1. Unreliable medical treatment
2. The removal of the entire mass enables accurate histopathological evaluation
3. Malignant transformation such as clear cell adenocarcinoma although rare, but has been reported in literature.[8,9]

CONCLUSION

Umbilical endometriosis is a rare umbilical disorder. Nevertheless, it has to be considered as a diagnosis in women of reproductive age who have an umbilical disorder. A thorough gynecologic history and examination is recommended to know the extent of the disease.

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Conflicts of interest

There are no conflicts of interest.

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