

Case Report

Primary umbilical endometriosis one case report and literature review

Shizhang Yang, Ruofan Dong

Department of Obstetrics and Gynecology, Affiliated Hospital of Jiangnan University, Wuxi 214000, Jiangsu, P. R. China

Received March 24, 2021; Accepted May 19, 2021; Epub June 15, 2021; Published June 30, 2021

Abstract: Primary umbilical endometriosis is rare and its exact pathogenesis is unknown. The definitive diagnosis is histopathological, and radical surgical excision is the treatment of choice. A primary umbilical endometriosis case how has been diagnosis and treatment in our hospital will be reported as follows. To get a better diagnosis and treatment, she came to our hospital because of the increase umbilicus bleeding symptom. A detailed physical examination was performed. According to the physical examination, there was a hard purple black nodular bulge, diameter about 2 cm, in the umbilicus and a small amount of brown blood was found at the same time. We gave her a blood test in detail. The results showed that cancer indicators were normal and CA125 was 33.8 u/ml. Ultrasound examination was played. About 14*10 mm of hypoechoic mass was found in the umbilicus, the boundary was clear, the internal echo was uneven, and the ultrasound imaging showed there was a richer blood flow signal. Based on inspection result and our team's judgement, we decided to surgery directly. Abdominal wall mass resection was underwent under general anesthesia in our hospital. The lesion is removed thoroughly, there is no need for hormone therapy after operation. We followed up regularly after the operation to now, there is no symptoms of discomfort or recurrence occurred. After literature review, combining with the results of our study, we believe that personalized treatment is advocated and radical surgery is the first choice.

Keywords: Endometriosis, umbilical endometriosis, umbilical, skin diseases, extra-pelvic endometriosis, abdominal wall endometriosis

Introduction

Extra-pelvic endometriosis is a rare type of endometriosis. Abdominal wall endometriosis (AWE) appear to be relatively frequent among cases of extra-pelvic endometriosis [1]. AWE describes the involvement of ectopic endometrial tissues superficial to the peritoneum of the abdominal wall, including lesions secondary to a surgical incision and spontaneous lesion [2]. The frequency of AWE has been estimated to be 0.04%-5.5% [3]. It is common, that the AWE regularly occurred in the cesarean scar or near the surgical mark site but there are also arising incidence in primary abdominal wall endometriosis. Umbilical endometriosis is an uncommon disease and often be included in the umbilical nodules. Umbilical endometriosis is kind of endometriosis of the abdominal wall endometriosis, including primary umbilical endometriosis and secondary umbilical endo-

metriosis that develop iatrogenically at the port site after laparoscopic surgery [2]. Primary umbilical endometriosis is rare and its exact pathogenesis is unknown. The treatment of choice is the radical surgical excision of the lesion. The definitive diagnosis is histopathological, and radical surgical excision is the treatment of choice. A primary umbilical endometriosis case which has been diagnosed, treated in our hospital will be reported as follows. This study was approved by the Regional Ethics Committee of our hospital (approval number 201901) and the patient signed informed consents.

Case report

A-42-years old women, G2P1, was diagnosed as primary umbilical endometriosis. In the history of disease, she has no surgical history. She was admitted to hospital out to umbilical peri-

Primary umbilical endometriosis one case report



Figure 1. Umbilical endometrioma: brownish nodules in umbilical region. Original magnification.

odic bleeding half a year, accompanied mild pain when menstrual. Bleeding amount was less. The colour sometimes was red, sometimes was brown, but after menstrual the symptoms were disappeared, repeatedly and periodic. In the September, 2018, she accepted abdominal wall ultrasound examination at a low-grade hospital. A tubercle 19*15*15 mm was found in the umbilicus (**Figure 1**). The tubercle was hypoechoic and the boundary was clear, spiny. Blood flow signal has not been fined in the tubercle by ultrasound examination. Irregular small dark areas can be seen and posterior echo attenuation in the lesions considering the possibility of endometriosis. After all medical inspection, she has not accepted any invasive treatment or taking any oral contraceptive pill. To get a better diagnosis and treatment, she came to our hospital because of the increase umbilicus bleeding symptom. A detailed physical examination was performed. According to the physical examination, there was a hard purple black nodular bulge, diameter about 2 cm, in the umbilicus and a small amount of brown blood was found at the same time. She felt light tenderness when we touched the nodule. We gave her a blood test in detail. The results showed that cancer indicators were normal and CA125 was 33.8 u/ml. Ultrasound is often the preferred imaging examination in endometriosis disease as for it's cheap, accurate and convenient characteristics. For accurate diagnosis, the ultrasound examination was replayed. About 14*10 mm of hypoechoic mass was found in the umbilicus, the boundary was clear, the internal echo was uneven, and the

ultrasound imaging showed there was a richer blood flow signal. After a preliminary judgement, the focus extent limited in 2 cm unaccumulated muscularis and peritoneum. Based on inspection result and our team's judgement, we decided to direct surgery. Abdominal wall mass resection was underwent under general anesthesia in our hospital, in December 2018. Postoperative pathology suggested that endometrial glands and stroma were found in fibrous tissue, which were consistent with endometriosis. The lesion is removed thoroughly, there is no need for hormone therapy after operation. We followed up regularly after the operation to now, there is no symptoms of discomfort or recurrence occurred.

Discussion

Extra-pelvic endometriosis is a rare type of endometriosis, which occurs in a distant site from gynecological organs such as abdominal wall endometriosis. It can compromise multiple organs. Abdominal wall endometriosis (Abdominal Wall Endometriosis, AWE) is an endometriosis between the wall peritoneum and the skin. It is common, that the AWE regularly occurred in the cesarean scar or near the surgical mark site but there are also arising incidence in primary AWE. It is a benign pathema, estrogen-dependent, affecting 6% to 10% of women in reproductive age. Umbilical endometriosis is kind of endometriosis of the abdominal wall endometriosis. It will occur after prolonged exposure to the metaplastic and environmental factors which catalyze the development of umbilical endometriosis. The incidence rate is not clear. Primary umbilical endometriosis is rare type of endometriosis, but among umbilical endometriosis, primary umbilical endometriosis is more frequent than secondary umbilical endometriosis. Other primary cutaneous sites include the vulva, perineum, inguinal region, and extremities [1]. Kimball studies on the pathological of the primary umbilical endometriosis lesions find that the presence of lymphoid interstitial infiltration in the lesion tissue, and it speculated that the occurrence of the disease may be related to the endometrium through lymphatic or vascular dissemination, and then they proposed the theory of lymphatic vascular dissemination [4]. Other research suggests that immunohistochemical analysis of primary endocutaneous

Primary umbilical endometriosis one case report

Table 1. Literature review of primary umbilical endometriosis arranged in chronological

Kimball KJ, etc. Diffuse endometritis in the setting of umbilical endometriosis: a case report. <i>J Reprod Med.</i> 2008.
Mechsner S, etc. Clinical management and immunohistochemical analysis of umbilical endometriosis. <i>Arch Gynecol Obstet.</i> 2009.
Richard F, etc. Spontaneous umbilical endometriosis: a rare but clinically important entity. <i>Am Surg.</i> 2011.
Koguchi-Yoshioka H, etc. Sister Mary Joseph's nodule: Malignant transformation of umbilical endometriosis. <i>J Dermatol.</i> 2016.
Al-Quorain SA, etc. Primary Umbilical Endometriosis: Case Report with Literature Review. <i>Saudi J Med Med Sci.</i> 2017.
Genovese G, etc. Ultrasonographic findings in primary umbilical endometriosis. <i>An Bras Dermatol.</i> 2018.
Rzepecki AK, etc. A Case of Primary Umbilical Endometriosis. <i>J Cutan Med Surg.</i> 2018.
Santos Filho PVD, etc. Primary umbilical endometriosis. <i>Rev Col Bras Cir.</i> 2018.

tissue specimens was also found to be associated with the redifferentiation of multifunctional stem cells [5]. According to the above theory, it suggests a mechanism the most likely cause of primary umbilical endometriosis is hypothesized to occur by migrating through blood or lymphatic vessel and a metaplasia theory has also been proposed. Some main related literatures of this study are shown in **Table 1**.

Clinical manifestation

The abdominal wall endometriosis is divided into three types according to the location of the lesion in the abdominal wall: superficial type (only in the subcutaneous tissue layer, above the fascia), intermediate type (invading the rectus abdominis fascia) and deep type (located in the rectus abdominis, below the fascia) [6]. The typical sign of abdominal wall endometriosis is that the abdominal wall can touch hard nodules. The nodules are round or oval, with irregular surfaces, fixed, poorly defined boundaries and tenderness. If the lesion is deep, the boundary is not easy to touch, or when the nodule is less than 1 cm it is more difficult to be found. The clinical presentation are umbilical swelling, cyclical pain, and sometimes bleeding from the lesion. Common age of abdominal endometriosis onset is childbearing age. Typical abdominal wall endometriosis has a triple sign, it were history of cesarean section, periodic pain associated with menstruation, or nodules at surgical scars [7]. As the same, patients with primary umbilical endometriosis often suffer from periodic bleeding associated with menstruation, and 90% complain of swelling and mass. Although most patients complain of pain, bleeding or swelling, there are still patients without any symptoms of discomfort [4, 6, 8]. Primary abdominal endometriosis occurs mostly in the umbilical cord or groin [3]. About 80%~90% of patients found periodic mass

pain, but corresponding skin discoloration can occur in less cases [9]. In the same, the symptoms of the patients studied in this paper was typical, with periodic bleeding with menstruation, accompanied by pain, and color changes in the skin at the lesion.

In this case, the abdominal wall lesions was about 2 cm, located in the umbilical cord, and the boundary was clear, tenderness was positive, the clinical diagnosis was not difficult. Because considering the obvious clinical symptoms, patient was considered to be given surgical treatment.

Diagnoses

The diagnosis of umbilical endometriosis is according to the typical clinical symptoms and initially on the medical history, physical examination and imaging modalities. In a recent systematic review, symptoms of swelling, pain, or bleeding at the lesions were present in 86.5%-90.9%, 80.5%-81.3%, or 44.8%-49.2% of cases, respectively [2]. Differentiating between umbilical endometriosis with other umbilical nodules is quite difficult. Published data shown that 20%-50% of umbilical endometriosis cases have been misdiagnosed [8]. It can be a challenging due to its rarity and lack of knowledge about the disease. Dermatologists and other physicians when evaluating abdominal masses in reproductive women should be remembered. In this case, the patient had an umbilical periodic bleeding, accompanied mild pain when menstrual. Ultrasonography showed typical endometriosis lesions. The clinical symptoms was obvious and diagnosis is not difficult.

Imaging examination

Imaging examination helps to determine the size, number, location, depth of the lesion and

Primary umbilical endometriosis one case report



Figure 2. Ultrasonography of the umbilicus demonstrated a 2 cm heterogeneous echogenic material within the umbilicus. Original magnification.

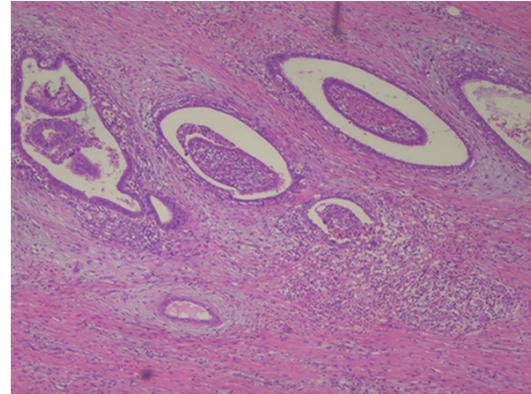


Figure 3. Histology of the umbilical lesion showing dense fibrosis and glandular structures lined by simple columnar epithelium together with endometrial stroma. Original magnification, $\times 400$.

its relationship with the surrounding organs. Ultrasound is often the preferred imaging examination. Ultrasound representation is that it showed solid, hypoechoic, cystic and multi-vascular manifestations, especially in the case of large lesions or lesion vessels anastomosed with the upper abdominal vessels, and the positive predictive value is 92.0% [10]. Magnetic Resonance Imaging (MRI) is more specific in the diagnosis of endometriosis, it can visually show the location, size, morphology, invasion range and relationship with adjacent tissues. MRI examination can be used for lesions larger, or more inaccessible masses involved between fascia and rectus abdominis [11]. Computed Tomography (CT) examination is rarely used in the diagnosis of endometriosis due to its lack of good soft tissue contrast [12]. However, in the past study that the positron emission tomography (PET-CT) was reported to be useful for the diagnosis of Malignancy [13]. Ultrasound is often the preferred imaging examination in endometriosis disease as for its cheap, accurate and convenient characteristics. In this case, about 14*10 mm of hypoechoic mass was found in the umbilicus, the boundary was clear, the internal echo was uneven, and the ultrasound imaging showed there was a richer blood flow signal (**Figure 2**). The diagnosis is basically clear so that we don't use other tests anymore.

Pathological

Endometriosis of skin is often characterized by local hematoma. Endometriosis in almost all areas has the same microscopic findings. There

are obvious endometrial glands and interstitial components in the pathological tissues. In our study case, histology of the umbilical lesion showing dense fibrosis and glandular structures lined by simple columnar epithelium together with endometrial stroma (**Figure 3**). In some large ectopic tissues, there is no typical endometrial glands and stroma on the wall of the capsule, but can be seen phagocytic macrophages containing hemosiderin.

Antidiastole

Umbilical cord endometriosis needs to be distinguished from abscesses, hematoma, lipoma, sebaceous cyst, fibroma, sarcoma, lymphoma, neuroma, and metastatic lesions [14, 15].

Treatment

Surgical treatment has been reported effectiveness [2]. The treatment of abdominal wall endometriosis is mainly surgical treatment, auxiliary drug treatment [16]. Hormone therapy is not curative it can be used preoperatively for relief of symptoms and reduce the size of large lesions prior to surgery. However, it is associated with side effects such as amenorrhea and the dienogest. A GnRH agonist, and oral contraceptives were effective for improving symptoms in 91.7%, 81.8%, and 57.1% of patients, respectively [2]. If the patient presents with infertility and exacerbated pelvic symptoms, a laparoscopy should be mandatory. Previous hormonal treatment may be an option for larger swelling, as it may reduce the size before surgery [17]. Simplicity drug therapy is oral contraceptives

Primary umbilical endometriosis one case report

and Gn-RH agonist. It can relieve symptoms but does not reduce the lesions. At present, the superficial and small lesions can be treated by cryoablation, high intensity focused ultrasound (Haifu Dao) and alcohol injection hardening [18, 19]. Surgical management is the preferred management of umbilical endometriosis for the following reasons [20]. Because the ectopic endometrium has the characteristic of invasion, with the course of disease prolonged, the more large of the lesions, the more tissue defect and the greater the incision tension in operation, resulting in more difficult suture, tissue healing ability weakened. Malignant transformation is estimated to occur in 0.6 to 0.8% of cases of ovarian endometriosis, but umbilical endometriosis malignant transformation has only rarely been reported in the literature [13]. Therefore, it should be treated as early as possible by surgery. For the patients treated by surgery, if the lesion is more than 3 cm, the patch should be placed during the operation, which has a good effect on the postoperative incision recovery [21]. Almost 70% of umbilical endometriosis patients required surgical management. Surgical procedure options include complete umbilical resection or local resection of the endometriotic mass sparing the umbilicus [20]. At present, there is no laparoscopic application in the treatment of umbilical endometriosis. The size of the umbilical nodule ranged from 0.5 to 4.0 cm in diameter, generating an average of 2.4 cm from the medical literature. Because the clinical symptoms of this patient are obvious, the lesions is about 2 cm at the umbilicus, the patient was performed surgical treatment. A wide resection of umbilical endometriosis can make the umbilicus disappear from the abdominal wall and can cause cosmetic problems so that a preoperative consultation with plastic surgeons and umbilical reconstruction should be considered.

It was not necessary to routinely perform laparoscopy to investigate abdominal foci as proposed because of higher hospital costs, except the patient presents with infertility and exacerbated pelvic symptoms. Radical surgical excision is the preferred choice. As to now, no postoperative recurrence was reported in cases of radical surgery resected to the peritoneum. In this study, the umbilical endometriosis lesions is about 2 cm so that we didn't use a patch and stitched the incision directly when surgery. The recurrence rate of umbilical endometriosis

after local resection is approximately 10% [2]. In this article, the patient has a good recovery after surgery, follow-up to the present no symptoms of recurrence.

After literature review, combined with the results of this study, the clinical significance of this study is that individualized treatment is advocated and radical surgery is the first choice. Furthermore, it would be advisable to develop a registry through multidisciplinary collaboration for better understanding and treatment of extra-pelvic endometriosis.

Acknowledgements

We gratefully thank the valuable cooperation of the women, who were included in this paper. The women have given consent to publish her case. This study was supported in part by Department of Obstetrics and Gynecology and Ethics Committee Of Affiliated Hospital of Jiangnan University. No conflict of interest exists in the submission of this manuscript, and manuscript is approved by all authors for publication. We gratefully thank the valuable cooperation of the women, who were included in this paper. The women have given consent to publish her case.

Disclosure of conflict of interest

None.

Address correspondence to: Shizhang Yang, Department of Obstetrics and Gynecology, Affiliated Hospital of Jiangnan University, Wuxi 214000, Jiangsu, P. R. China. E-mail: yangsz1986@126.com

References

- [1] Andres MP, Arcoverde FVL, Souza CCC, Fernandes LFC, Abrão MS and Kho RM. Extrapelvic endometriosis: a systematic review. *J Minim Invasive Gynecol* 2020; 27: 373-389.
- [2] Hirata T, Koga K and Osuga Y. Extra-pelvic endometriosis: a review. *Reprod Med Biol* 2020; 19: 323-333.
- [3] Horton JD, Dezee KJ, Ahnfeldt EP and Wagner M. Abdominal wall endometriosis: a surgeon's perspective and review of 445 cases. *Am J Surg* 2008; 196: 207-12.
- [4] Kimball KJ, Gerten KA, Conner MG and Richter HE. Diffuse endometritis in the setting of umbilical endometriosis: a case report. *J Reprod Med* 2008; 53: 49-51.

Primary umbilical endometriosis one case report

- [5] Mechsner S, Bartley J, Infanger M, Loddenkemper C, Herbel J and Ebert AD. Clinical management and immunohistochemical analysis of umbilical endometriosis. *Arch Gynecol Obstet* 2009; 280: 235-42.
- [6] Grigore M, Socolov D, Pavaleanu I, Scripcariu I, Grigore AM and Micu R. Abdominal wall endometriosis: an update in clinical, imagistic features, and management options. *Med Ultrason* 2017; 19: 430-437.
- [7] Khan Z, Zanfagnin V, El-Nashar SA, Famuyide AO, Daftary GS and Hopkins MR. Risk factors, clinical presentation, and outcomes for abdominal wall endometriosis. *J Minim Invasive Gynecol* 2017; 24: 478-484.
- [8] Fancellu A, Pinna A, Manca A, Capobianco G and Porcu A. Primary umbilical endometriosis. Case report and discussion on management options. *Int J Surg Case Rep* 2013; 4: 1145-1148.
- [9] Zhang J and Liu X. Clinicopathological features of endometriosis in abdominal wall—clinical analysis of 151 cases. *Clin Exp Obstet Gynecol* 2016; 43: 379-383.
- [10] Genovese G, Veraldi S, Passoni E and Nazzaro G. Ultrasonographic findings in primary umbilical endometriosis. *An Bras Dermatol* 2018; 93: 297-298.
- [11] Takeuchi M, Matsuzaki K and Harada M. Susceptibility-weighted MRI of extra-ovarian endometriosis: preliminary results. *Abdom Imaging* 2015; 40: 2512-2516.
- [12] Busard MP, Mijatovic V, van Kuijk C, Hompes PG and Van Waesberghe JH. Appearance of abdominal wall endometriosis on MR imaging. *Eur Radiol* 2010; 20: 1267-1276.
- [13] Koguchi-Yoshioka H, Oshima H, Manago E, Kuwae K, Shioji M, Adachi K, Nakamichi I, Hoshi M, Ikeda K and Matsumoto C. Sister Mary Joseph's nodule: malignant transformation of umbilical endometriosis. *J Dermatol* 2016; 43: 1449-1450.
- [14] Ideyi SC, Schein M, Niazi M and Gerst PH. Spontaneous endometriosis of the abdominal wall. *Dig Surg* 2003; 20: 246-248.
- [15] Rzepecki AK, Birnbaum MR and Cohen SR. A case of primary umbilical endometriosis. *J Cutan Med Surg* 2018; 22: 325.
- [16] Santos Filho PVD, Santos MPD, Castro S and Melo VA. Primary umbilical endometriosis. *Rev Col Bras Cir* 2018; 45: e1746.
- [17] Richard F, Collins J and Britt LD. Spontaneous umbilical endometriosis: a rare but clinically important entity. *Am Surg* 2011; 77: e246-247.
- [18] Zhu X, Chen L, Deng X, Xiao S, Ye M and Xue M. A comparison between high-intensity focused ultrasound and surgical treatment for the management of abdominal wall endometriosis. *BJOG* 2017; 124: 53-58.
- [19] Bozkurt M, Çil AS and Bozkurt DK. Intramuscular abdominal wall endometriosis treated by ultrasound-guided ethanol injection. *Clin Med Res* 2014; 12: 160-165.
- [20] Al-Quorain SA and Al-Yahya TA. Primary umbilical endometriosis: case report with literature review. *Saudi J Med Sci* 2017; 5: 74-76.
- [21] Comănescu M, Potecă A, Lăzăroiu A and Sajin M. Endometriosis of the abdominal wall. *Rom J Morphol Embryol* 2007; 48: 195-197.