Management options for primary umbilical endometriosis: a case report

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ABSTRACT

Background: The common location of Endometriosis is in genitalia organs, for instance, the ovary, uterus, fallopian tube, and sometimes even in the intestine, bladder, and in rare locations such as the navel, lungs, or brain. Umbilical Endometriosis is the most irregular form of Endometriosis and the most common cutaneous form of Endometriosis. Primary umbilical endometriosis diagnosis is often biased and delayed; the exact etiopathology remains unclear. Our case report discusses the diagnosis and management options for this rare disease.

Case presentation: We reported a Primary Umbilical Endometriosis case, confirmed by a history of the nodule with pain, swelling, and bleeding at the umbilicus, which occurs during menstruation. The nodule was surgically removed, and histopathological analysis shows fibromycsoid tissue with multiple forms of subepithelial endometrial glands and surrounding stroma, confirmed as Endometriosis.

Conclusion: The definitive treatment for umbilical Endometriosis is surgical excision with total removal of the umbilicus. The prognosis is good, and the recurrence rate is meager if complete excision is successfully performed.

Keywords: endometriosis, rare endometriosis, cutaneous endometriosis, umbilical endometriosis.


INTRODUCTION

Endometriosis is the endometrial-like tissue presence outside the uterus, occurring in 10-15% of women of childbearing age. Endometriosis can be found in the peritoneum, ovaries, and sometimes even in the intestine, bladder, and in rare locations such as the lungs or umbilicus. Pelvic organs such as fallopian tubes, ovaries, recto-vaginal septum, uterosacral ligaments, and peritoneum are also places where Endometriosis is commonly found. Symptoms vary depending on severity and inflammation factors, including pelvic pain, menorrhagia, painful intercourse, defecation, and micturition complaints related to menstruation.

It is reported about 1-12% of Endometriosis have extragenital Endometriosis. Umbilical Endometriosis, also known as Villar nodules, is the rarest form of Endometriosis and the most common cutaneous form of Endometriosis. Many theories have tried to explain the pathophysiology of umbilical Endometriosis. When umbilical Endometriosis occurs spontaneously without any history of surgery, it is classified as primary, and if it is the result of endometrial implantation triggered by different surgical procedures, it is called secondary endometriosis.

Several authors have suggested immune system alterations in endometriosis hosts. It has been reported to indicate an increase in B cell activity, cytokines and prostaglandins, and decreased natural cell killer’s activity in peritoneal and serum fluids. In cases of Primary Umbilical Endometriosis (PUE) with concomitant pelvic Endometriosis, local inflammation surrounding the Endometriosis causes endometrial cells to migrate to the navel, whereas in isolated PUE, its metaplastic change is suggested to be from urachal remnants.

Keloids, granulomas, benign and malignant tumors, such as primary and metastatic neoplasms, and polyps are the differential diagnosis of UE. However, there is minimal risk of malignant transformation. Because cases of PUE are rare, guidelines regarding its treatment do not exist. The main objective of our case report is to discuss and review the treatment options for PUE.
CASE REPORT

A 37-year-old woman, parity 2, came with pain and swelling at the navel, which increased during menstruation over the past year. This lesion has grown slowly over a year. Her umbilicus was tender, painful, and bleed during menstruation. The patient has been married for 15 years and has had two normal vaginal deliveries. She has been using oral contraceptives for nine years and has had regular menstrual cycles. Medical history was unremarkable, and the patient has no history of abdominal trauma, cesarean section (C-section), or other surgeries. There were no complaints of dysmenorrhea, problems with sexual intercourse, or dyschezia. Upon physical examination, we found a dark brown nodule sized 2 centimeters located in her umbilical fold (Figure 1). From transvaginal and abdominal ultrasounds, there were no signs of Endometriosis. She underwent surgical resection of umbilical nodule and performed a histopathological analysis of the nodule.

Umbilical resection was performed under spinal anesthesia. Dissection and resection around the nodule to the lateral umbilicus were done until normal tissue margin was visible. Excision was made about 2 cm in diameter, mainly around the endometriotic nodules to the abdominal fascia. Umbilical closure performed with simple interrupted sutures using Polyglycolide acid and prolene as non-absorbable sutures (Figure 2).

Histopathological findings from the resected nodule showed fibromycoid tissue, multiple subepithelial endometrial glandular structures, surrounding endometrial stroma, and no malignancy. In conclusion, the results were consistent with the diagnosis of umbilical cord endometriosis.

The patient was observed seven days after surgery with mild periumbilical pain. Clinical examination suggests no signs of local infection and inflammation in the wound. She recovered well and felt very satisfied with the surgery results (Figure 3).

DISCUSSION

Incidence of Umbilical Endometriosis (UE) is rare and is estimated to account for only 0.4% to 1 % of all patients with extra pelvic Endometriosis. The pathophysiology is associated with pelvic surgical procedures or may occur spontaneously. The theories regarding its etiopathology are varied. Cutaneous Endometriosis occurs due to the transplantation of viable endometrial cells into the scars at the time of surgery, mainly if the surgical procedure contacts endometrial tissue (hysterectomy, C-Section, and ectopic pregnancy). Spontaneous primary umbilical Endometriosis is developed through metaplasia of urachus remnants or transport of endometrial cells from the pelvis through the lymphatic channels and blood vessels.

The exact pathogenesis of umbilical Endometriosis remains unclear. One theory held by Sampson and Scott suggests that UE is formed by endometrial tissue transferred through lymphatic and vascular channels. This theory was developed when they found that dyes and radioactive material were recovered in the umbilicus when injected into the peritoneal cavity. Some lymphatics connect the peritoneal cavity and umbilicus. These lymphatics flow along missing umbilical blood vessels. Migration of endometrial tissue is theorized to occur through these lymphatics. Another theory suggests that during laparoscopy or through the hematogenous spread, endometrial cells can reach the umbilicus by seeding and may then proliferate.

Many theories have been proposed regarding endometriosis etiopathogenesis. Novak has suggested the coelomic metaplasia theory, and Von Recklinghausen has suggested the embryonal rest theory, both of them have indicated that cells come from the Wolfian duct system or coelomic mesothelium have had a transformative change at the cellular level, which subsequently leads to Endometriosis. These theories, however, can only explain Endometriosis which occurs close to the pelvic organs. Migratory pathogenesis is thought to be the most common explanation for Endometriosis. This includes the theory...
CASE REPORT

Umbilical Endometriosis is sometimes described as dark-bluish, flesh-colored, or hemorrhagic in appearance, and can vary from several millimeters to numerous centimeters. The macroscopic appearance of UE may vary during the menstrual cycle. Differential diagnosis includes benign or malignant skin neoplasm, metastatic adenocarcinoma, and hernia or lesions which are inflammatory or infectious.

In this case report, our patient had an uncommon case of spontaneous Umbilical Endometriosis, which is endometrial tissue presence in the umbilicus without any history of C-section or any other gynecologic surgeries. The more common form, secondary Umbilical Endometriosis, is caused by implantation of endometrial cells and iatrogenic spreading during laparoscopic or pelvic surgery. This form of Endometriosis is relatively easier to explain in terms of pathogenesis. Different hypotheses have been put forward to explain primary Endometriosis, such as Wolffian and Mullerian remnants as embryonal rest theory and the transplantation theory in which retrograde menstruation or hematogenous spread causes migration of endometrial tissue. Nevertheless, the exact pathogenesis of primary Endometriosis is still unknown.

At times, the clinical diagnosis of umbilical Endometriosis is not easy. In terms of clinical findings, typical symptoms of UE include the dark brown nodule in the navel, which is swollen, painful, and bleeds during the menstrual periods. In our case report, our patient had a dark brown nodule in the umbilicus, which bleeds during the menstrual cycle. However, it is essential to note that the macroscopic appearance of UE may vary and can be described as dark-bluish, flesh-colored, brownish, or just a subcutaneous mass. Size may range from less than 1 centimeter to numerous centimeters. Umbilical Endometriosis is sometimes confused with malignant tumors such as melanoma due to variations in macroscopic appearance. Therefore, the differential diagnosis should include umbilical skin discoloration, such as cyst lipoma, abscess, benign nevus, abscess, or hernia.

Several reviews have noted that UE occurs at 37.7 years, with the youngest patient aged 23. The patient, in this case, is aged 37 years old. The mainstay for diagnosis in cases of UE is a clinical picture and a physical examination. Differential diagnosis includes benign or malignant skin neoplasm, metastatic adenocarcinoma, and hernia or lesions which are inflammatory or infectious. If the clinical picture and physical examination suggest UE, then radiological tools can detect the umbilical's connection with abdominal and peritoneal repair or an endometriotic nodule locally excision. These diagnostic tools can detect the umbilical's connection with the peritoneum or fascia. Another accurate method of UE diagnosis is Magnetic resonance imaging (MRI). In contrast, histopathological means of diagnosis such as fine-needle aspiration cytology were less accurate in most cases. The UE histopathological marker were findings of endometrial glands implanted in the stroma. These glands have a strong vascular and cellular constituent. These endometrial glands form irregular glandular Lumina. During menstruation, these endometrial glandular bleed into the umbilical skin, which leads to extravasated erythrocytes. To further confirm the diagnosis of UE, Immunohistochemistry may be used to find the expression of CD10 antigen, estrogen, and progesterone receptors. These are all markers used for stromal cells in Endometriosis. Other common findings of UE are the inflammations mediators, marked mitotic activity, and hemosiderin deposit presence in the stroma. It is crucial to do a careful assessment to rule out atypia to exclude malignancy if myxoid or hypertrophic decidual changes appear.

When immunohistochemical properties of a normal endometrium include Keratin 7+, Keratin 20 expression, besides expression of estrogen receptors, progesterone receptors, and Ki-67, a metastatic process should be suspected. Adenocarcinoma metastatic from the gastrointestinal are usually keratin-20+. Another valuable tool in determining an unclear diagnosis is CD10, which is known to be highly expressed in endometrial stromal nodules. Nevertheless, its efficacy is sometimes doubted because some fibroblasts also express this neutral endopeptidase from normal dermis. In cases of ectopic endometrium, calretinin expression is low to absent, whereas, in eutopic endometrium, it is expressed. In this case, histopathological findings showed characteristics that resemble those of glandular tissue and endometrial stroma without atypia. Our case report did not assess keratin 7+, ER- and PR receptors.

Treatments for UE are limited. Surgical excision remains to be the only ideal option for treatment. Often, surgery is managed when the ectopic tissue is least active due to hormonal conditions at the end of the menstrual cycle. The choice of surgical technique depends on the size and depth of the lesion and must be individualized for each patient. According to Victory et al., usually Umbilical Endometriosis patients require surgical excision. However, because UE is very rare, standard surgical consensus regarding its treatment is still very lacking. Currently, the procedure options include a complete umbilical resection with or without fascial and peritoneal repair or an endometriotic nodule locally excision. The most frequently performed operation for UE is total resection of the umbilicus. This choice is primarily for patients with chronic UE symptoms and enlargement of endometriotic nodules. Some literature has reported cases of UE extending to the abdominal wall tissue and even to the umbilical fascial. Surgical complication of UE is associated with large umbilical hernias which explains why such a radical approach is needed for its treatment. Even though this operation yields inferior cosmetic results, several authors have recommended a complete umbilical resection regardless of size. Hormonal therapy of umbilical Endometriosis can only temporarily reduce the clinical symptoms of UE, which will reoccur after its cessation. The most common hormone used to treat Endometriosis are oral contraceptives and Gonadotropin-releasing hormone.

Figure 3. Scar after the seventh-day post-surgery.
Agnosists. Our patient was given GnRH agonists to reduce her pelvic endometriosis/dysmenorrhea symptoms. A complete nodule excision procedure is recommended to prevent a recurrence. However, few studies follow up on the outcomes after the procedure. The risk of relapse must be fully informed to patients with simple local nodule excision. When operating on patients with UE, laparoscopic exploration should be performed to exclude further spread of intraabdominal Endometriosis since transvaginal ultrasound cannot exclude pelvic Endometriosis definitively. All patients are recommended to be evaluated by a gynecologist for coexisting internal Endometriosis, which occurs in around 15% of patients with UE. Umbilical endometriosis prognosis is good, with meager rates of recurrence if complete excision is performed. However, a few pieces of literature have mentioned the risk of scar tissue transformation due to wide excision, so follow-up is required.

When UE is diagnosed, complete or local nodule excision should be offered due to several considerations. The first reason is that the entire resection of the nodule enables accurate histopathological findings of UE and, therefore, can exclude other umbilical malignancies such as skin neoplasms or metastases. In addition, complete resection of the umbilical is required due to malignancy transformations of endometriotic lesions. Finally, early surgery yields better cosmetic results. If the size of the nodule is still tiny, it does not require extensive resection of the umbilicus.

CONCLUSION

Primary umbilical Endometriosis is an unusual and rare form of e endometriosis ectopia. Currently, its etiopathogenesis is not yet fully understood. Most studies now focus on diagnosing and managing this Endometriosis form and have identified histopathological confirmation as the primary form of diagnosis. The mainstay modality of treatment is surgical management. Early diagnosis can avoid the need to perform extensive local excisions. When coming across an umbilical mass during physical examination, practitioners of different specialties should consider PUE as a differential diagnosis.

CONFLICT OF INTEREST

The authors have nothing to disclose.

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ETHICAL CONSIDERATION

This case report did not require any ethical clearance. The patient and her families understand and agree about the publication of receptive medical data in this journal article.

AUTHOR CONTRIBUTION

All authors contributed to the study, including literature research, data collection, data analysis, and manuscript preparation.

REFERENCES