Case Report

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Endometriosis of the umbilicus in a 36-year-old woman: a case report and literature review

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Introduction and importance: Extrapelvic endometriosis is quite rare, with a reported prevalence ranging between 0.5 and 1%, and the condition is more likely to pose diagnostic challenges. This condition is more likely to pose clinical diagnostic challenges as it may mimic metastasis such as Sister Mary Joseph's nodule.

Case presentation: Herein is reported the case of a 36-year-old woman who presented with a hard nodular dark-bluish umbilicus mass that had a tendency to increase in size and was accompanied by severe pain during menstruation for ~2 years is reported. Laparotomy revealed a normal uterus without involvement of any other pelvic organ by the endometrial tissue except the umbilicus part. Histological evaluation revealed endometriosis of the umbilicus.

Clinical discussion: By far, primary endometriosis of the umbilicus is extremely rare, and most of the time, extrapelvic endometriosis involving the umbilicus would be secondary to surgical procedures involving the abdominal cavity as it was for the presented patient. Although endometriosis is rare, it should always be considered among women of reproductive age presenting with cyclic pains.

Conclusions: Meticulous investigation of patients suspected to have umbilical endometriosis helps to confirm the diagnosis and hence expedites proper management of the patients; this also prevents chances of malignant transformation of the condition despite such possibilities being extremely rare.

Keywords: endometriosis, extrapelvic endometriosis, umbilicus

Introduction

Extrapelvic endometriosis is defined as the growth of functional endometrial tissue outside the uterine cavity[1]. It affects from 3 to 10% of women of reproductive age who usually present clinically with symptoms such as dysmenorrhea, noncyclic pelvic pain, infertility, or menorrhagia, as it was in the case presented[2]. Endometriosis commonly affects the pelvis; however, extrapelvic involvement is not rare[3]. Umbilical endometriosis is quite rare, with a reported prevalence ranging between 0.5 and 1%, and the condition usually affects patients after laparoscopy or other surgical procedures involving the abdomen, as it was in the patient reported in this paper. Primary umbilical endometriosis is even by far the rarest, and the pathophysiology of this condition is not totally clear[4]. Herein, the case of a 36-year-old woman with a 2-year history of painful umbilical swelling associated with dysmenorrhea is reported along with a review of the existing literature. The patient was managed at a national consultant hospital after being referred from a district public hospital.

HIGHLIGHTS

- Diagnosis of extrapelvic endometriosis requires both clinical and laboratory findings.
- Cyclic abdominal pains among women of reproductive age with secondary infertility should raise suspicion of endometriosis.
- Raised levels of estrogen, progesterone, and estradiol hormones among women of reproductive age with cyclic abdominal pains should also raise suspicion of endometriosis.

Case presentation

A 36-year-old primigravidae African woman presented with an umbilical lump that had bluish discoloration for 2 years. She reported a history of cyclic monthly pain associated with bleeding from the umbilicus. Her past medical involves appendectomy for the past 16 years, but her family history was uneventful. On physical examination, she had a 1×2 cm purplish nodule at the umbilicus (Fig. 1). It was not tender, and there were no signs of infection. There was a 2.0 cm healed surgical scar on the upper right-side part of the abdomen as a result of an appendectomy that was done 2 years ago.

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Figure 1. Umbilical lump with bluish discoloration.

Abdominopelvic ultrasound was essentially normal. Abdominal computed tomography scan revealed hydronephrosis of the right kidney. However, the rest of the intra-abdominal organs were normal. The provisional diagnosis included Sister Mary Joseph's nodule and hemangioma. Fine needle aspiration cytology of the umbilical nodule was highly suspicious for the diagnosis of endometriosis (Fig. 2).

After a thorough discussion between the patient and a team of gynecologists with the patient regarding the need for surgical removal of the umbilicus and the possibility of recurrence, she consented to the removal of the umbilicus. Before the removal of the lesion, she was first managed medically using analgesics for 2 months; however, she did not show total improvement. Then the nodular umbilicus mass was completely excised, and the defect was repaired. Excision of the mass was done under general anesthesia by a gynecologist together with a master's student. She was prescribed analgesics and antibiotics and discharged home on second day postoperatively. The mass was taken for histological evaluation. Microscopically, the tissue sections from the umbilical mass revealed the proliferation of typical endometrial glands in a nonreactive endometrial stroma. The glands are lined by columnar epithelium that resembles the endometrial glandular epithelium. The subepithelium is composed of small round cells that resemble the endometrial stroma. This confirmed the diagnosis of umbilical endometriosis (Fig. 3). Therefore, it took ~2 years from the initial presentation of the signs and symptoms until when the definitive diagnosis was obtained.

The patient was followed up for 1 year postoperatively, and she denied any pain from the operated site, and there was no recurrence after the clinical evaluation was done. Moreover, for 1 year after management, she could not conceive despite her current condition being stable. This work has been reported in line with the SCARE (Surgical CAse REport) 2020 criteria[5].

Discussion

Umbilical endometriosis can be primary or secondary. In 1886, Villar reported the first case of umbilical endometriosis. Hence, this condition is also called Villar's nodule[1]. The incidence of umbilical endometriosis has been reported to range from 0.5 to 1%[3]. Umbilical endometriosis is divided into primary and secondary endometriosis[6].

Primary umbilical endometriosis implies the presence of ectopic endometrial tissue located in the umbilicus in the absence of previous surgery for either gynecological disorders or Cesarean section[7]. Primary umbilical endometriosis is known to be associated with severe pelvic endometriosis compared to secondary endometriosis[2]. On the other hand, secondary umbilical endometriosis is linked with iatrogenic dissemination and implant of endometrial cells during either laparoscopic or open surgical procedures. In terms of occurrence, secondary umbilical endometriosis is more common than primary umbilical endometriosis[4,8]. The pathogenesis of secondary umbilical endometriosis seems to be relatively easy to explain as it is to explain the origin of primary umbilical endometriosis. However, different hypotheses have been proposed, such as the embryonal rest theory of Wolfian or Müllerian remnants, the transplantation theory in which the ectopic endometrial

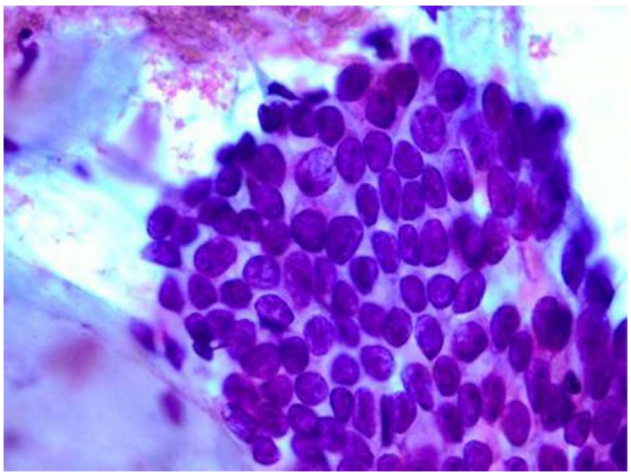


Figure 2. Papanicolaou stain showing epithelial cells without atypia which resembles endometrial origin with hemorrhage (×400).



Figure 3. Haematoxylin and eosin section showing tortuous glands surrounded by dense cellular stroma lying in a collagenous background (×100).

tissue harbors from retrograde menstruation or hematogenous/lymphatic dissemination, or a combination of them[9,10].

The typical symptoms of umbilical endometriosis include the presence of a discrete bluish-purple mass in the umbilicus, becoming swollen, painful, and bleeding concomitantly with the menstrual cycle[11]. Diagnosing umbilical endometriosis clinically is sometimes difficult because of its varying presentations in color, characteristic of pain, and size, for that matter. Therefore, malignant melanoma, paraumbilical hernia, granuloma, primary or metastatic adenocarcinoma (Sister Mary Joseph's nodule), nodular melanoma, and cutaneous endosalpingiosis should be thought of as clinical differentials. In the published series, umbilical endometriosis is misdiagnosed in 20–50% of cases[12].

Diagnosis of umbilical endometriosis involves clinical presentation and physical examination as mandatory and key issues for obtaining the right diagnosis. Fine needle aspiration cytology has been reported to be not confirmatory for diagnosing umbilical endometriosis[13]. The suspicion made from the cytopathology of the aspirate materials taken from the umbilical mass can be confirmed by means of histopathology, which also helps to exclude the presence of metastases to the umbilicus[14,15]. Immunohistochemistry using CD10 biomarkers for staining the stromal cells has also been reported to be supportive when it comes to diagnosing umbilical endometriosis[14,15]. Furthermore, imaging techniques such as MRI and computed tomography scan, on the other hand, have been found helpful in the process of diagnosing umbilical endometriosis[14,15]. Therefore, the diagnosis of umbilical endometriosis requires a meticulous collection of information for its definitive diagnosis.

Treatment of umbilical endometriosis using hormonal therapy helps to diminish the clinical symptoms temporarily[16]. However, after the cessation of hormonal therapy, it is likely that the symptoms tend to reoccur. Agonists of gonadotropin-releasing hormones and oral contraceptives are the most frequently used hormones to treat endometriosis[1,3]. In the review article by Machairiotis *et al.*[3], it was argued that almost 70% of patients with umbilical endometriosis require surgical treatment. However, consensus about standard surgical management of umbilical endometriosis is lacking, probably due to the rare nature of the disease.

The operative options include complete umbilical resection, with or without repair of the underlying fascia and peritoneum; local excision of the endometrial nodule, sparing the umbilicus[17]. Total removal of the umbilicus is the most frequently performed operation for umbilical endometriosis[15]. When umbilical endometriosis is suspected on the basis of clinical and imaging workup, surgical treatment is recommended for several reasons. Removal of the entire lesion only enables accurate histopathological diagnosis of umbilical endometriosis, thus, excluding unusual malignant disorders of the umbilicus, such as metastases or other skin neoplasms[14,15]. Furthermore, removal of the nodule is warranted because the malignant transformation of endometrial lesions, although rare, has been described[15]. Timely and early surgery is more likely to result in good cosmetic outcomes, especially when the size of the umbilical endometriosis nodule does not mandate the removal of the entire umbilical fold.

The strength of our work in presenting the case is based on the fact that we were able to perform cytological and histological evaluations of the lesion, which helped to increase the evidence for the diagnosis. In consideration of the diagnostic approach for the patient described in this paper, we faced a limitation of having included immunohistochemistry staining with CD10 antibody, which we could not manage to purchase due to lack of funds.

Conclusions

Umbilical endometriosis is an extremely rare gynecological condition that requires meticulous investigations for its diagnosis because it is more likely to be missed due to its cyclic nature as well as available other conditions which are more likely to mimic it. The decision on treatment should be tailored on the size of the lesion, duration of symptoms, and nature of the lesion so as to prevent it from being confused with metastatic lesions such as Sister Mary Joseph's nodule. While managing patients with umbilical endometriosis, the possibility of recurrence should always be anticipated. The possibility of either primary or secondary infertility as the complications of extrapelvic endometriosis must also be discussed.

Ethical approval

Not applicable.

Patient consent

Written informed consent was obtained from the patient for the publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Author contribution

J.J.Y. and E.D.M.: acquisition, organizing, and writing the first draft of the manuscript: Z.S.A.: preparation of the images. All authors reviewed the manuscript critically.

Conflicts of interest disclosure

The authors have no conflicts of interest to declare.

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James J. Yahaya agreed to be the guarantor and agreed to have full responsibility for the work.

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