Primary Umbilical Endometriosis: Unusual Clinical Presentation with Microscopic Diagnosis.



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ABSTRACT

Introduction Endometriosis, commonly occurs in the pelvic organs, mostly the ovaries, with dysmenorrhea, and menorrhagia. Umbilical endometriosis is rare. Symptoms include umbilical masses with swelling and pain. So it is often not recognized based on its clinical appearance. In this report, it was presented a case of primary umbilical endometriosis, accidentally found after surgery for umbilical hernia and detected in pathologic examination of tissue.

Patient Information A 32-year old female, was presented with umbilical bulging and mild cyclic umbilical pain, without history of abdominal surgery, or symptom of pelvic endometriosis. Umbilical sonography was done in favor of fixed sac of herniation, underwent surgery, umbilical endometriosis accidentally found in pathologic examination after surgery for umbilical hernia.

Conclusion Although endometriosis is a common gynecological disease, primary umbilical endometriosis is a rare entity that make the diagnosis is difficult. Nevertheless, this disease must be considered in the differential diagnosis of any suspected umbilical mass.

Keywords Endometriosis; Umbilicus; Pain; Pathology

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[1] Umbilical endometriosis: a rare diagnosis in plastic and reconstructive surgery [2] Perforating hemorrhagic (chocolate) cysts of the ovary [3] Endometriosis-associated infertility [4] A case of umbilical endometriosis: Villar's nodule [5] Umbilical hernia masking primary umbilical endometriosis – a case report [6] Primary umbilical endometriosis. Case report and discussion on management options [7] Primary Umbilical Endometriosis: Unusual and Rare Clinical Presentation [8] Surgical scar endometriosis after cesarean section: a case report [9] Primary umbilical endometrioma: a rare case of spontaneous abdominal wall endometriosis [10] Atypical clinical presentation of primary umbilical endometriosis: a cause of a painful umbilical nodule [14] Menstruating from the umbilicus as a rare case of primary umbilical endometriosis: a case report [15] Endometriosis of the umbilicus [16] Endometriosis of the umbilicus

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Primary Umbilical Endometriosis: Unusual Clinical Presentation ...

Introduction

Endometriosis is defined by the presence of endometrial glands and stroma outside the uterine cavity $^{[1,2]}$. It affects 7-10% of women of reproductive age presenting with dysmenorrhea, menorrhagia, pelvic pain, and infertility $^{[3,4]}$ and commonly occurs in the pelvic organs $^{[5,6]}$, mostly the ovaries, the Fallopian tubes, the utero-sacral ligaments, and the recto-vaginal septum $^{[7,8]}$.

Umbilical endometriosis (UE) is rare and constitutes about 0.5% to 1% of the patients with extra genital endometriosis [9, 10], primary umbilical endometriosis without history of previous surgery is even rarer condition [5, 11]. Symptoms include umbilical masses with swelling and pain [12] during menstruation and, to a lesser extent, bleeding [10]. The differential diagnosis of the condition includes both benign and malignant tumors [12]. So primary umbilical endometriosis (PUE) is often not recognized based on its clinical appearance, considering the importance of diagnosis of cause of abdominal pain and its timely diagnosis and considering endometriosis as one of the differential diagnoses of vague abdominal pain that can lead to long-term complications if not timely diagnosed [13].

In this report, it was presented a case of primary umbilical endometriosis, accidentally found after surgery for umbilical hernia and detected in pathologic examination of tissue.

Patient Information

A 32-year old female, gravid 2, was presented to the surgeon's office with 6 month history of umbilical bulging and mild cyclic umbilical pain. She did not sense the relation between degree of bulging and pain with her menstruation. She had no history of abdominal surgery or any medical problem, and she had not symptoms of pelvic endometriosis such as dysmenorrhea, or dyspareunia. She was not taking any oral contraceptives and the patient had regular, mild painless menstrual periods. Umbilical sonography was done for the patient that was shown hypo echoic mass M: 13×10mm at the base of umbilicus in favor of umbilical cyst or fixed sac of herniation was seen.



Figure 1) Umbilicus cut sections show a dark cystic nodules surrounded by fibrosis



Figure 2) A low power field showing a dermal glandular proliferation of endometrial-type glands surrounded by endometrial stroma

With the diagnosis of umbilical sinus and hernia, underwent surgery with accompanying umbilical reconstruction and the umbilicus was send for pathology for more study. The microscopic examination revealed endometrial glands and stroma and endometriosis was diagnosed (Figures 1 and 2).

Discussion

Patient with primary umbilical endometriosis is usually in the reproductive age group typically manifested by a bluish nodule with pain and tenderness associated with cyclic bleeding or discharge during menstruation.

In Bagade *et al.* study a 35 year-old woman was presented with symptoms of spontaneous and periodic bleeding from the umbilicus for 4 months. The bleeding started two days before her menses and continued for the entire duration of her period [14]. In our case, the patient did not show any discoloration of skin, bleeding in the umbilicus or pain.

Unlike Taniguchi *et al.*'s case presentation [7], our case has not relation with menstruation, so it was impossible to establish a definitive diagnosis of umbilical endometriosis only by clinical data, surgical exam or sonography. It should be placed in many differential diagnosis of umbilical swelling such as hernia, lipoma, primary and metastatic adenocarcinoma, or nodular melanoma [14], even in women with no typical symptoms of pelvic endometriosis [7].

Also the specific feature of our case is that the patient has not previous surgery, (called primary UE) which the its different from secondary UE is that implant of endometrial cells can occur during surgical procedures [15, 16] and increase the possibility of this rare phenomenon. While the pathogenesis of secondary UE seems to be relatively easy to explain, it is harder to clarify the origin of primary UE [6].

In this regard, different hypothesis have been proposed, for primary UE such as the embryonal rest theory of Woll-fian or Mullerian remnants, the transplantation theory in which the ectopic endometrial tissue harbors from retrograde menstruation or hematogenous/lymphatic dissemination, or a combination of them ^[6]. However, the pathogenesis of primary endometriosis still remains unclear. Histopathological features are characterized by irregular glandular Lumina embedded in the stroma resembling the stroma of functional endometrium ^[14].

Conclusion

Although endometriosis is a common gynecological disease, primary umbilical endometriosis is a rare entity that make the diagnosis is difficult. Nevertheless, this disease must be considered in the differential diagnosis of any suspected umbilical mass.

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