Journal of Bangladesh College of Physicians and Surgeons
Vol. 41, No. 2, April 2023

Primary Umbilical Endometriosis: A Rare Entity

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Abstract

Primary umbilical endometriosis is a rare benign gynaecological disorder caused by ectopic endometrial tissue in the umbilicus. The disease typically manifests as a painful umbilical nodule associated with cyclic bleeding or discharge during menstruation. Here, a 46-year-old woman with low parity and without previous history of abdominal surgery has been reported who presented with cyclical per-umbilical bleeding and pain in the umbilical region during menstruation without any palpable umbilical nodule. The patient responded well to initial medical management, but symptoms recurred on discontinuation of the medication. Though clinically, there was no palpable nodule, and ultrasonography revealed a hypoechoic lesion in the lower umbilical region suggestive of endometrial deposition. On resuming medical management, her symptoms abated.

(J Bangladesh Coll Phys Surg 2023; 41: 173-176)
DOI: https://doi.org/10.3329/jbcps.v41i2.64508

Introduction:

Endometriosis is a common benign gynaecological condition affecting 6-10% of women in the reproductive age group[1]. The disease is defined by functioning glands and stroma outside the uterus[1,2]. It commonly occurs in the pelvic organs, especially the ovaries, the uterosacral ligaments, and the pouch of Douglas. Women with endometriosis often present with dysmenorrhoea, menorrhagia, pelvic pain, and infertility [3].

Due to its varied presentation, endometriosis often remains difficult to diagnose and treat. Extra-genital endometriosis is less common but may involve almost every part of the female body including the bowel, bladder, lungs, brain, umbilicus, and surgical scars[4]. Umbilical endometriosis represents 0.5-1.0% of all cases of extra-genital endometriosis and usually occurs secondary to surgical scar[5, 6]. Very rarely, the disease may present as primary umbilical endometriosis with an unclear pathogenic mechanism.

Case study

A 46-year-old woman with low parity (one vaginal delivery and one abortion) presented on February 2016 with spontaneous bleeding through the umbilicus during menstruation which she noticed for one month [Figure-1]. There was a pain in the periumbilical region, which persisted during the whole length of her menstrual period. She had no history of abdominal surgery, and her medical history was unremarkable. She complained of primary dysmenorrhoea, which was relieved following her first childbirth.

Her ultrasonography of the lower abdomen and periumbilical area revealed no abnormality, and serum CA-125 was within the normal limit. Thus, she was offered oral progesterone in suppressive dose (norethisterone acetate 5 mg thrice daily) for three consecutive months. However, six months later, she again developed bleeding through the umbilicus during menstruation, which was not associated with significant pain. Oral danazol 100 mg daily was started for three months, the patient became amenorrhoeic, and her symptoms abated. Unfortunately, she discontinued the medication. One year later, she again noticed umbilical bleeding during menstruation. This time the first ultrasonographic evidence was available as a tiny, septate cystic area, measuring 3 mm X 6 mm in the muscle layer of the para-umbilical region, suggestive of para-umbilical endometriosis [Figure-2A]. Treatment was started with oral dienogest 2 mg daily for six months, and she remained free of symptoms for one year. Follow-up ultrasonography two years later showed bilateral adnexal cysts. One clear cyst (2.6 cm X 1.6 cm) in the right adnexal region, another cyst having low-level echoes (2.7 cm X 2.6 cm) in the left adnexal region, and a small hypoechoic lesion measuring about 10 mm X 4.5 mm in the umbilicus at its lower part, features suggestive of small endometriotic tissue in the umbilicus [Figure-2B]. She continued medical management, with no further bleeding per umbilicus and no pelvic pain. As the patient had a very small endometriotic deposit in the umbilicus that was not approachable and symptoms alleviated by taking an oral medication, surgical intervention was unnecessary. She is on regular observation without

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Received: 03 August, 2022 **Accepted:** 07 January, 2023

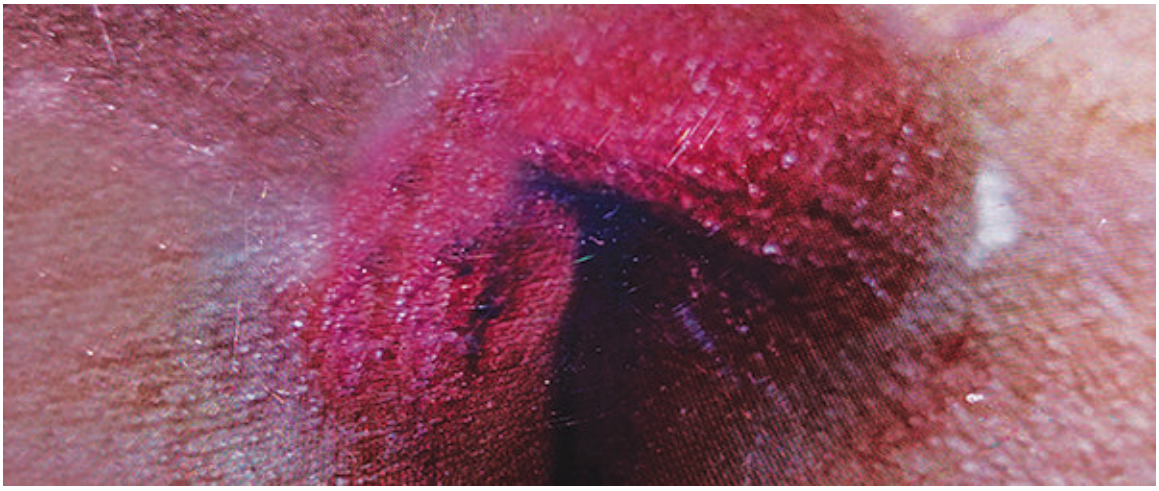

Fig.-1: *Bleeding per umbilicus*

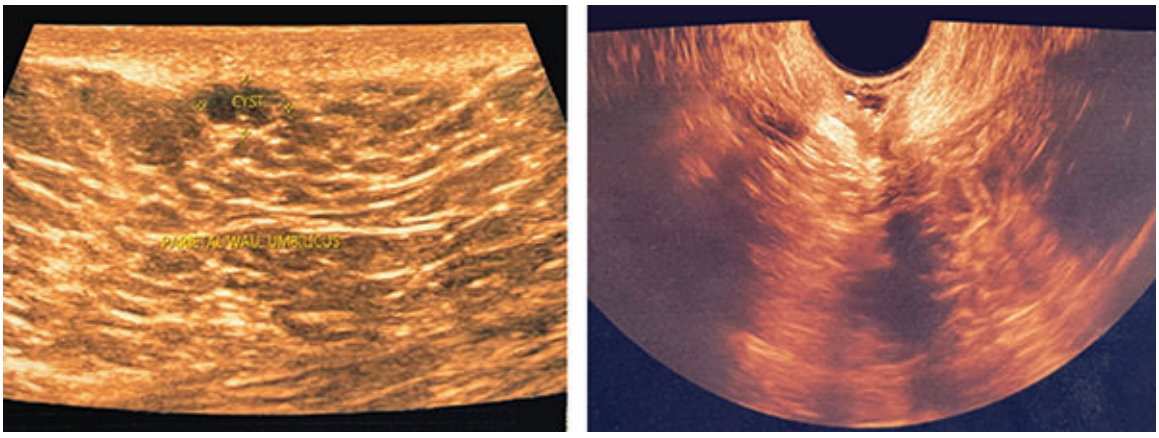

Fig.-2: *Ultrasonogram of the parietal wall of the umbilicus showing a cystic lesion in the muscle layer of the para-umbilical region [on 02/01/2018] (A), and two years later [on 11/02/2021] (B)*

having any symptoms and has no umbilical or endometrial deposit on sonographic follow-up.

Discussion:

Primary umbilical endometriosis (PUE) is a rare entity but should be considered in the differential diagnosis of umbilical disorders even without typical symptoms of pelvic endometriosis. However, the umbilicus is a preferred spot for cutaneous endometriosis. Villar first described the disease in 1886; since then, more than a hundred cases have been described. But an endometriotic umbilical lesion without a surgical history (i. e. primary umbilical endometriosis) is a rare entity [2, 7]. The pathogenesis of PUE is not fully elucidated. Possible explanations could be the migration of endometrial cells to the umbilicus through the abdominal cavity, the lymphatic system, or the embryonic remnants in the umbilical fold, such as the urachus and the umbilical vessels [8-10]. The disease is prevalent among Caucasian and Asian women, whereas the black race can be a protective factor[11].

PUE is typically manifested as a firm, pigmented or bluish nodule with pain and tenderness associated with cyclic bleeding or discharge during menstruation [8]. Our patient had occasional cyclical bleeding through the umbilicus, variable in amount, sometimes frank bleeding or a few drops of blood collected in the umbilicus. She did not have significant dysmenorrhoea, dyspareunia or pelvic pain, and there was no palpable pelvic mass or umbilical

nodule. The literature reports a 13-15% incidence of the simultaneous presence of pelvic endometriosis[12]. As such, although some authors suggest a concomitant pelvic evaluation, this approach is not mandatory and should be considered in cases with a high index of suspicion for pelvic endometriosis [3,5,8].

Different diagnostic modalities include ultrasonography, computed tomography of the abdomen, and magnetic resonance imaging. However, laparoscopic surgery with histopathology is the gold standard. Zhai et al. stated that fine needle aspiration biopsy (FNB) is useful for diagnosing cutaneous endometriosis[13]. Although there are no reports on the diagnostic accuracy of FNB in umbilical endometriosis, Zhao et al. stated that in 75% of cases, FNB might not come to a conclusive diagnosis in abdominal as well as umbilical endometriosis[14]. As there was no palpable nodule or mass in our patient, a tissue diagnosis could not possible. Compatible clinical features and sonographic evidence helped us to reach a clinical diagnosis.

Management of umbilical endometriosis could not be standardized owing to the scarcity of cases. Victory and co-workers, in a review, showed that almost 70% of patients required surgical treatment[15]. Medical management using progesterone, danazol, norethisterone, and GnRH analogue has not shown reliable results. Nonetheless, some authors reported significant clinical improvement in relieving symptoms and reducing the size of the endometrial nodule using medical and hormonal treatment[1,16]. Thus, medical management cannot be enthusiastically recommended due to its relatively low success rate, temporary alleviation of symptoms, serious side effects and recurrence following cessation of drugs. It is also known that the abdominal wall and scar endometriosis are less responsive to hormonal therapy. But in our report, it was observed that the patient responded well to medical management without any further bleeding. She was under regular follow-up for the last six years, and repeat ultrasonography showed no residual umbilical disease. As there was no approachable tissue, we could not go for FNAC. Also, further evaluation by CT scan or MRI were not done, which is a limitation of our case report.

Conclusion:

Finally, this case report draws our attention to an uncommon disease, the diagnostic conundrum of PUE, in the absence of a typical lesion. It highlights the role of hormonal management of umbilical endometriosis. Our patient had a compatible clinical feature and radiological finding of PUE. As there was no palpable mass, surgery and histological confirmation could not done. Though she had recurrence with two drugs (norethisterone acetate and danazol) at earlier period, lastly she responded well to dienogest. Since, the patient is now at perimenopausal age, and she may have full recovery within next one or two year, she may continue medical management as required.

Conflict of interest

There is no potential conflict of interest to declare.

Disclosure

Appropriate written informed consent was obtained for the publication of this case report.

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