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Umbilical endometriosis: A case series on the different approaches in management

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Abstract:

Endometriosis is defined as the presence and growth of the glands and stroma of the lining of the uterus in an aberrant or heterotopic location. The cause of endometriosis is uncertain and involves many mechanisms including retrograde menstruation, vascular dissemination, metaplasia, immunologic, and hormonal influences to name a few. Primary umbilical endometriosis is a rare disorder defined as the presence of ectopic endometrial tissue within the umbilicus. It commonly presents with catamenial pain and bleeding from an umbilical nodule. It is a rare condition and treatment has not been standardized yet due to the limited number of cases. Surgical management is the treatment of choice to avoid local recurrence while hormone therapy may be used preoperatively for symptomatic relief. This paper reports a series of recently observed cases of primary umbilical endometriosis with main aim of discussing the different approaches in the management of this rare condition.

Keywords:

Cyclical pain, endometriosis treatment, medical management, surgical excision, umbilical endometriosis, umbilical mass

Introduction

Endometriosis is defined as the presence of endometrial-like tissue outside or beyond the lining of the uterus. The lesions can be peritoneal lesions, superficial implants, or cysts on the ovary or deep infiltrating disease.[1] It affects an estimated 5%–15% of reproductive-aged women and is detrimental to fertility. There are a variety of theories proposed about the pathogenesis of endometriosis. The embryonal rest theory states that endometriosis follows the Wolffian or Mullerian remnants. The coelomic metaplasia theory explains the presence of ectopic endometrium tissue through dedifferentiation of coelomic mesothelium. The migration theory states that endometrium tissue can spread via direct extension, vascular or lymphatic vessels. Furthermore, iatrogenic implantation after surgery is commonly seen.[2,3] Umbilical endometriosis, also known as Villar's nodule, is a rare condition and majority of the cases presented with a macroscopic nodule of the umbilicus and occurred through iatrogenic implantation.[4]

This is a case series showing the different approaches in the management of umbilical endometriosis in a tertiary hospital in the Philippines.

Case Reports

This is a case series involving three patients with umbilical endometriosis, managed in a tertiary hospital in the Philippines, who have all presented with a bleeding umbilical mass.

Patient 1: M.G. 40-year-old gravida 1 para 1 (1001)

She has no known comorbidities and no previous surgeries. Her family medical

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history was unremarkable. She had her menarche at 13 years old, with subsequent menstrual periods coming in at regular intervals, lasting for 3–5 days and soaking 3–4 pads per day with no associated dysmenorrhea. Her first and only pregnancy in 2000 was carried to term and delivered via normal spontaneous delivery with no fetomaternal complications.

Five years before admission when she noted the presence of nonfoul smelling bloody umbilical discharge, which was associated with menses [Figure 1]. There were no noted tenderness and umbilical swelling on the area. No consults were done, no medications were taken. In the interim, there was persistence of the bloody umbilical discharge which was now accompanied by a gradually enlarging violaceous umbilical nodule, which was nontender on palpation. This prompted consult at the out-patient department of a tertiary hospital where a transvaginal ultrasound revealed: three well-circumscribed heterogeneous myometrial masses, with a federation of gynecology and obstetrics classification grade 4, two of which are located at the posterior midcorpus and one at the posterofundal area. Occupying the abdominopelvic cavity is a multilocular cystic mass measuring 11.2 cm × 9.8 cm × 9.7 cm with homogenous low-level echo fluid within. A simultaneous scan of the umbilical mass showed an irregular anechoic mass measuring 1.0 cm × 1.4 cm × 1.5 cm at the subcutaneous layer of the periumbilical area giving the impression of umbilical endometriosis. She was advised surgery (exploratory laparotomy, total hysterectomy and bilateral salpingectomy with unilateral oophorocystectomy) and was given hormonal suppression while awaiting surgery. Three months before scheduled admission and surgery, she was started on gonadotropin-releasing hormone (GNRH) agonist injection, 3.75 mg IM every 4 weeks for 3 doses. After which the patient was then shifted to Desogestrel, one tablet once a day while awaiting clearance for surgery.

Figure 1: A case of umbilical endometriosis seen on day 2 of menses

Upon admission at the same tertiary hospital last October 2019, on abdominal examination, there was a 10 cm × 10 cm cystic mass noted at right lower quadrant, which was slightly movable and nontender. There was also a 3 cm × 4 cm soft, fixed, nontender purplish nodule at the umbilicus. The pertinent physical examination findings showed that at the right adnexa, a 10 cm × 10 cm × 5 cm cystic, slightly movable, nontender mass was appreciated. On rectovaginal examination, uterosacral nodularities were noted.

The patient underwent total hysterectomy with bilateral salpingectomy, right oophorocystectomy, adhesiolysis, enterolysis, and wide excision of umbilical mass. Gross examination showed a 2.0 cm × 2.0 cm × 0.5 cm fleshy, bluish colored mass with cystic areas and hemorrhages on the excised umbilicus [Figure 2]. Final histopathological examination of the mass showed stratified squamous epithelium of the skin of the umbilicus with endometrial glands and stroma on the dermis [Figure 3], conclusive of an umbilical endometriosis. Postoperatively, this patient did not note any recurrence of the umbilical mass nor did have any episode of umbilical bleeding nor discharge [Figure 4].

Patient 2: M.B. 30-year-old gravida 4 para 3 (3003)

She has no known co-morbidities and no previous surgeries. She had her menarche at 11 years old, with subsequent menstrual periods coming in at regular intervals, lasting for 3–4 days and soaking 3–4 pads per day with no associated dysmenorrhea. Her last menstrual period was last November 8, 2020, giving her an amenorrhea of 33 weeks and 2 days as of this writing. This is her fourth pregnancy. All her other pregnancies were carried to term and delivered via normal spontaneous delivery with no fetomaternal complications. She currently has her prenatal consultations done at a lying-in clinic in Las Piñas where she plans to give birth.

Three years prior to consult when she noted a pea-sized, gradually enlarging, nontender, violaceous mass at the umbilicus. The mass would bleed in conjunction with her menstrual periods. In the interim, there was

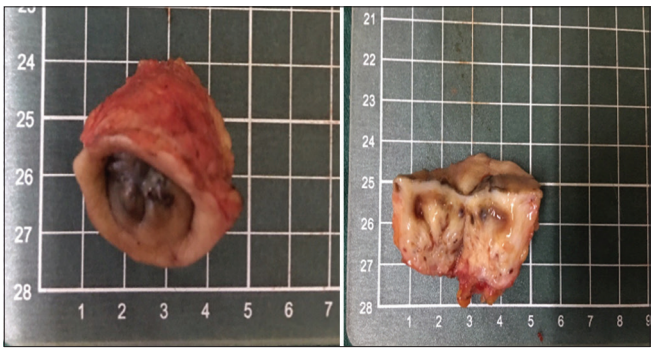

Figure 2: Upon umbilical excision, a 2.0 cm × 2.0 cm × 0.5 cm fleshy, bluish-colored mass with cystic areas and hemorrhages was seen

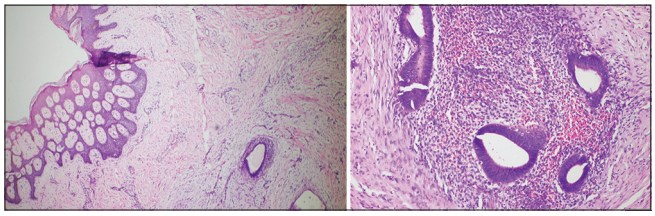

Figure 3: Final histopathological examination of the mass showed stratified squamous epithelium of the skin of the umbilicus with endometrial glands and stroma on the dermis

persistence of the gradually enlarging violaceous umbilical mass, which was now associated with tenderness on palpation noted during menses. One year prior to consult she was seen at a family medicine clinic in an out-patient department of a tertiary hospital. An abdominal computed tomography scan was requested which revealed the following findings: "presence of a bulky enhancing heterogenous lower uterine segment and cervical focus with adjacent nodularities. In this case, where endometriosis is considered, endometriotic infiltrations is a possibility. However, a neoplastic process cannot be totally ruled out. An umbilical focus, which may also be part of endometriosis is also noted."

She was then referred to the OB-GYN Department but was unable to follow up due to the pandemic. Transvaginal ultrasound revealed the following findings: "Normal sized retroverted uterus, secretory phase endometrium, normal right ovary, left ovarian cyst consider physiologic cyst, umbilical mass, probably endometriotic cyst." During face-to-face consult at the OB-GYN outpatient department, the umbilical mass was noted to be 2.0 cm × 2.0 cm × 2.0 cm, well-circumscribed, soft, nontender, brownish-violaceous in color. An initial incisional biopsy of the umbilical mass revealed a histopathological diagnosis consistent with endometriosis. The long-term plan for the patient was wide excision of the umbilical mass. She delivered by spontaneous vaginal delivery to a live baby boy with an unremarkable postoperative course. There was no progression of the said umbilical mass in size and in pain severity during the course of her pregnancy; hence, no further consult was done.

Patient 3: M.O. 40-year-old gravida 2 para 2 (2002)

She have no known comorbidities nor previous surgeries. She had her menarche at 13 years old, with subsequent menstrual periods coming in at regular intervals, lasting for 3–4 days and soaking 3–4 pads per day with no associated dysmenorrhea. All her pregnancies were carried to term and delivered via normal spontaneous delivery with no fetomaternal complications.

Five years before consult when she had a history of manipulation of her umbilicus which led to an abrasion.

Figure 4: One-year postumbilical mass excision showed no recurrence of any umbilical mass, bleeding nor discharge

This was accompanied by umbilical pain with a pain score of 1–2 out of 10. She noted a gradually enlarging, soft, flesh-colored umbilical mass initially pea-sized with no bleeding nor foul smelling discharge. No consults were done and no medications were taken. In the interim, there was persistence of the umbilical pain, which was now associated with menstruation and a gradually enlarging violaceous umbilical mass. This prompted consult at the outpatient department.

Initially seen by general surgery, an abdominal computed tomography scan was requested which showed an enhancing soft-tissue nodule along the midline at the umbilical region which appears to extend deep into the linea alba likely represents an abdominal wall endometrioma. During this interval, the patient reported an umbilical pain score of 10 out of 10 during menses not relieved by intake of pain medications. Patient was then referred to the gynecology service for further evaluation and management of the umbilical mass, likely an umbilical endometriosis.

A transvaginal ultrasound was done with the following findings: *"Normal sized anteverted uterus, Proliferative phase endometrium, Normal ovaries"*. A concomitant abdominal scan revealed a periumbilical mass described to be an ovoid unilocular cystic mass measuring 2.6 cm × 2.6 cm × 2.1 cm with medium level echo fluid within with the consideration of umbilical endometriosis. The initial plan for this patient was treatment with GNRH agonist, but the patient opted Medroxyprogesterone acetate 150 mg intramuscularly every month for 3 months then every 3 months thereafter for 2 years due to financial constraints. Due to the pandemic community restrictions, the patient initially scheduled for face-to-face consult was lost to follow up. Currently, the patient has no vaginal bleeding, discharge, urinary or bowel movement changes. There was no noted change in the

size of her umbilical mass but with a decrease in pain score from 10 to 8 out of 10 [Figure 5].

Discussion

Endometriosis is the presence and growth of the glands and stroma of the lining of the uterus in an aberrant or heterotopic location. The cause of endometriosis is uncertain and involves many mechanisms including retrograde menstruation, vascular dissemination, metaplasia, genetic predisposition, immunologic changes, inflammation. and hormonal influences such as estrogen dependence and progesterone resistance. The typical patient with endometriosis is in her mid-30s, nulliparous and involuntarily infertile, and has symptoms of secondary dysmenorrhea and pelvic pain. Aberrant endometrial tissue grows under the cyclic influence of ovarian hormones and is particularly estrogen dependent; therefore, the disease is most commonly found during the reproductive years.

Cutaneous endometriosis is relatively uncommon and occurs when endometrial glands and stroma reside in the skin and it can be divided into primary and secondary endometriosis. Primary or sometimes referred to as spontaneous umbilical endometriosis was first defined by Villar in 1886, hence the term "Villar's nodule." The pathogenesis for primary cutaneous endometriosis remains unclear and develops spontaneously without any history of local surgery or may be associated with metaplasia of urachal remnants. All of the three patients presented in this case series are primary umbilical endometriosis due to the fact that they all presented with an umbilical mass that is accompanied by catamenial umbilical pain and all of them had no history of abdominal or pelvic surgery. In contrast, the secondary cutaneous endometriosis, which is also called scar endometriosis, is believed to occur due to seeding after abdominal or pelvic surgery. Its color can range from blue or violaceous to brown or skin-colored. Discoloration occurs as a result of bleeding into the lesion with hemosiderin deposition, which can be seen at histopathological examination. The skin is an uncommon location for endometriosis, and cutaneous endometriosis cases comprise <1% of all cases in large series[5] and the umbilicus is a physiologic scar that is a preferred site because of the lymphatic and hematogeneous spread and direct extension of endometrial cells through the round ligaments of the omphalomesenteric remnants.

Figure 5: A case of umbilical endometriosis treated medically, and received 3 doses of depot medroxyprogesterone acetate 150 mg IM every month for 3 months since July 2021–September 2021. There was no noted change in the size of the umbilical mass but with a decrease in pain score from 10 to 6 out of 10

The second case is that of an umbilical endometriosis in pregnancy, a rare occurrence and with only limited cases recorded as of this time. It is associated with the hormonal response of endometriotic tissue to the increased estrogen production during the first trimester of pregnancy causing the catamenial pain and bleeding. A case report on umbilical endometriosis in pregnancy in 2004 presented a case of a 28-year-old at 16 weeks age of gestation presenting with a tender reddish-brown polypoid nodule within the umbilical depression. An excisional biopsy was done and the nodule was histologically confirmed to be endometriosis. No therapy was given and the lesion spontaneously resolved 2 months after the biopsy was taken (at 24 weeks' age of gestation).[6] The researchers discussed that as the pregnancy progress and there is a shift from estrogen to progesterone predominance, this will cause stromal decidualization and sclerosis of endometrial lesions bringing about a decrease in symptoms.

In differential diagnosis of umbilical nodules, benign diseases such as secondary endometriosis, hemangioma, umbilical hernia, sebaceous cyst, granuloma, lipoma, abscess, keloid, and urachus anomaly, and malignant diseases such as Sister Mary Joseph's nodule, melanoma, sarcoma, adenocarcinoma, and lymphoma should always be considered. Sensitivity and specificity of ultrasonography, computerized tomography, and magnetic resonance are low in the diagnosis of umbilical endometriosis. None of these imaging techniques have a pathognomonic finding of umbilical endometriosis. Ultrasonography can provide some information on the size of the nodule and its cohesion to adjacent tissues. Histopathological analysis confirms the diagnosis of umbilical endometriosis with the visualization of endometrial glands and stroma in the underlying skin.

Once a diagnosis of either primary or secondary cutaneous endometriosis is established, the definitive treatment is surgical excision with wide margins. A study done on 33 women with cutaneous endometriosis showed that 31 of the women had a surgical history that suggested secondary cutaneous endometriosis. The majority of these women (90%) received surgical excision. Overall, cutaneous endometriosis recurred in three women (9%), suggesting surgical excision is a reasonable management option.[5] In the first case, a wide local excision of the umbilical mass was done for a definitive management

of the umbilical endometriosis with no postoperative administration of progestins. On her follow-up consult done 1 year postoperatively, the patient did not have any recurrence of umbilical mass nor did not have any catamenial umbilical pain, thereby strengthening the evidence that surgical excision is the treatment of choice in decreasing the recurrence of umbilical endometriosis.

Another option for patients who do not wish to undergo surgery is treatment with hormonal agents alone, such as GnRH agonists or danazol. These agents work by preventing cyclical proliferation of the endometrial tissue. In one case series, Drs. Robert Purvis and Steven Tyring (1994) describe two cases in which medical management was attempted, one with danazol and the other with leuprolide. In both cases, hormonal therapy was able to relieve associated pain, and in one case the nodule diminished in size by approximately 50%. However, the patient treated with monthly injections of subcutaneous leuprolide experienced return of her pain between her second and third treatment cycles. Both patients opted for surgical excision of their lesions within 1 year due to the unfavorable side effects of the hormonal therapies.

Another medical management option for patients who opted not to undergo surgical management for endometriosis would be the administration of progestins as a first-line hormonal therapy in the management of pain in endometriosis. Dienogest, a fourth-generation progestin works by binding to the progesterone receptor and, when taken continuously, inhibits systemic gonadotropin secretion and has local anti-proliferative and anti-inflammatory effects on endometriotic lesions.[7] In a study done by Pathiraja and Ranaraja in 2020, the authors presented a case of vulvar and umbilical swelling suggestive of endometriosis. The patient underwent complete surgical excision of the umbilical mass. After the surgery, the patient had few cycles of Depot Medroxyprogesterone Acetate (DMPA) injection and after 6 months, she was asymptomatic. In the third case presented where the patient was given Medroxyprogesterone acetate 150 mg intramuscularly, there was noted partial relief of the umbilical pain. Oral contraceptives (OC), progestin, or GnRH agonists may be effective in improving the symptoms and can be an option for patients who do not want surgery. However, there are no sufficient data available to prove the effectiveness of the use of medical management such as Dienogest and DMPA in the treatment and in preventing the recurrence of extra-genital endometriosis after resection.

Although surgical excision with wide margins is the gold standard for therapy, if the decision is made to opt for hormonal therapy, surveillance with histopathology should be considered due to case reports noting instances of malignant transformation. Surgical choices include either total umbilical resection with or without repair of underneath fascia and peritoneum, or local excision of endometrial nodule with preserving umbilicus. Total resection of umbilicus is mostly preferred. Local excision of endometrial lesion should be performed by achieving adequate edge of the surrounding normal tissue to avoid local recurrence.

A Clinical Practice Guidelines for the treatment of extragenital endometriosis in Japan was published in 2020 at The Journal of Obstetrics and Gynecology by Tetsuya Hirata *et al.* and it stated that relapse of umbilical endometriosis following excision occurred more than 2 years after making surgical management effective in the short term, but the long-term recurrence rate and complications remain unknown. With this, the authors recommended that it is preferable to excise the lesion using wide local resection. Umbilical deformities associated with wide local resection may occur and would sometimes require umbilical reconstruction. The authors also noted that although medical therapy is useful in the relief of endometriosis-associated symptoms, the administration of medical therapy is likely to be prolonged and the disease may recur after discontinuation of medication. Preoperative administration of GnRH agonists is not recommended because of the increased risk of incomplete resection due to reduced tumor size and difficulty in its localization.[8] Preoperative treatment with dienogest or combined OCs may be effective, but it is difficult to treat the endometriosis with hormone treatment alone, while surgical approach is preferable to conservative treatment because of complete surgical excision and histological confirmation.

Conclusion

A woman presenting with an umbilical lesion with notable violaceous discoloration and accompanied by tenderness associated with menstruation, umbilical endometriosis should be considered and early diagnosis and prompt treatment is provided. Wide local umbilical resection should be preferred to decrease the incidence of local recurrence, attain a favorable cosmetic outcome and relief of symptoms. There has been no sufficient evidence on the management of umbilical endometriosis in pregnancy. Medical management in the form of progestins aid in alleviating the symptoms associated with endometriosis, however, do not entirely eliminate the presence of endometrial glands and stroma in an ectopic location within the umbilicus and would eventually resort to surgical management as treatment of choice.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in

the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Authorship contributions

Janelle Marie G. Doloiras - Involved in the conceptualization, methodology, validation, formal analysis, resources, writing-original draft, writing-review and editing, visualization.

Regina Paz A. Tan-Espiritu - Involved in the conceptualization, formal analysis, resources, writing-review and editing, supervision.

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Conflicts of interest

There are no conflicts of interest.

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