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Case Report

A primary umbilical endometrioma in a 38-year-old woman: a case report

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ABSTRACT

Primary umbilical endometriosis (UE) is a rare case of migration of endometrial tissue to a distant location. However, there are literature gaps surrounding this area of study and may be easily missed when forming differential diagnoses for umbilical masses. We report a case of primary UE in the setting of abdominal pain and umbilical mass. The patient is a 38-year-old female who was admitted for abdominal pain and an umbilical mass. An umbilical mass without evidence of an umbilical hernia was found on computed tomography of the abdomen and pelvis. The differential diagnosis of umbilical hernia was then introduced. Exploration and surgical excision were utilized. Pathologic frozen sections confirmed endometrioma. Both medical and surgical therapies may be used in treating UE. This case highlights the need for awareness of rare endometriosis manifestations and illustrates the diagnostic challenges when assessing for possible causes of umbilical mass. By adequately evaluating for causes such as UE, we can accurately treat it with medical and surgical therapies.

Keywords: Primary umbilical endometrioma, Umbilical mass, Endometriosis, Villar's nodule, UE

INTRODUCTION

Endometriosis affects 190 million reproductive-aged women worldwide.[1] This medical condition is characterized by the growth of endometrial tissue in locations outside the uterus, such as ovaries, fallopian tubes, the outer surface of the uterus, and other organs within the pelvis. In rare cases, endometrial tissue may also appear in more distant areas, including the umbilicus.[2] Umbilical endometriosis (UE), initially reported by Villar in 1886, is also known as Villar's node and accounts for approximately 0.5%-1% of all reported cases of endometriosis, highlighting the need for increased awareness.[3] Primary UE manifest in individuals without a history of abdominal surgery. Secondary UE arise from scar tissue formation after abdominal procedures such as cesarean delivery or, rarely, laparoscopy. The pathogenesis of endometriosis, particularly primary UE, remains poorly understood, contributing to the diagnostic challenges and delay in treatment. Improving early detection of rare endometriosis variants requires a multifaceted approach, including heightened clinical suspicion, advanced imaging techniques, and histopathological confirmation. Educating healthcare professionals about UE can lead to more timely and accurate diagnoses, enabling effective treatment strategies and positive patient outcomes. Significant gaps remain in the literature regarding the etiology, diagnosis, and management of UE, hindering the development of standardized diagnostic criteria and treatment. We present a 38-year-old woman who presented with an umbilical mass presumed to be an incarcerated hernia, which was later confirmed to be a primary UE. This case highlights the need for awareness

of rare endometriosis manifestations and illustrates the diagnostic challenges posed by this condition.

CASE REPORT

A 38-year-old woman presented to the emergency department with three days of severe abdominal pain associated with an umbilical mass. She denied fever or respiratory, gastrointestinal, or urinary symptoms. Over the past three years, umbilical swelling, thought to be a hernia, had been associated with a recurrent non-radiating, cyclical pain, for which warm compresses and ibuprofen provided only temporary relief. Her past medical history included anemia, dysmenorrhea, hyperlipidemia, hypertension, and migraine headaches. Her surgical history was limited to a cesarean section in the context of pre-eclampsia for the birth of her only child nine years prior. She suffered from menorrhagia and had undergone an extensive infertility workup, including transvaginal ultrasound, sonographic hysterogram, endometrial biopsy, sonogram, and MRI of the pelvis with findings of a retroflexed uterus, a left-sided hematosalpinx, and equivocal endometriosis in the right lateral vagina. Current medications include drospirenone-ethinyl estradiol, ferrous sulfate, gabapentin capsules, ketorolac, magnesium oxide, propranolol, sumatriptan, and terconazole. On physical exam, the patient was afebrile, hemodynamically stable, with an ill-defined, tender umbilical mass without skin change. A contrast-enhanced CT scan of the abdomen and pelvis revealed a 1.6×1.8×1.5 cm heterogeneous mass within the superficial subcutaneous fat in the deep portion of the umbilicus without any evidence of hernia. The possibility of endometrioma was entertained. She underwent exploration and surgical excision of the mass under monitored anesthesia care (MAC). Fibroadipose tissue and connective tissue with endometrial glands and stroma on the frozen and later permanent sections confirmed endometrioma. The patient was seen on follow-up with a well-healed incision and reported being pain-free.

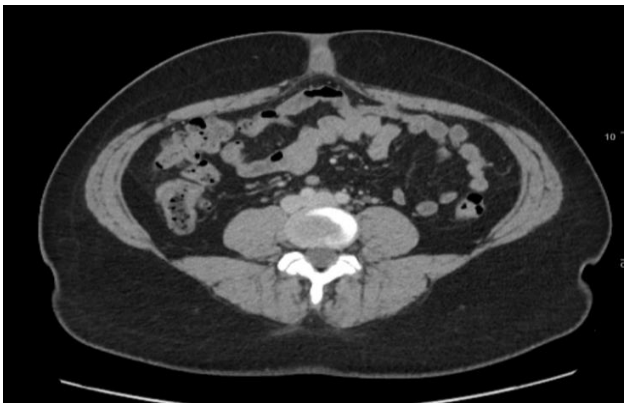

Figure 1: A transverse plane CT scan showing a 1.6×1.8×1.5 cm heterogeneous mass within the superficial subcutaneous fat in the deep portion of the umbilicus.

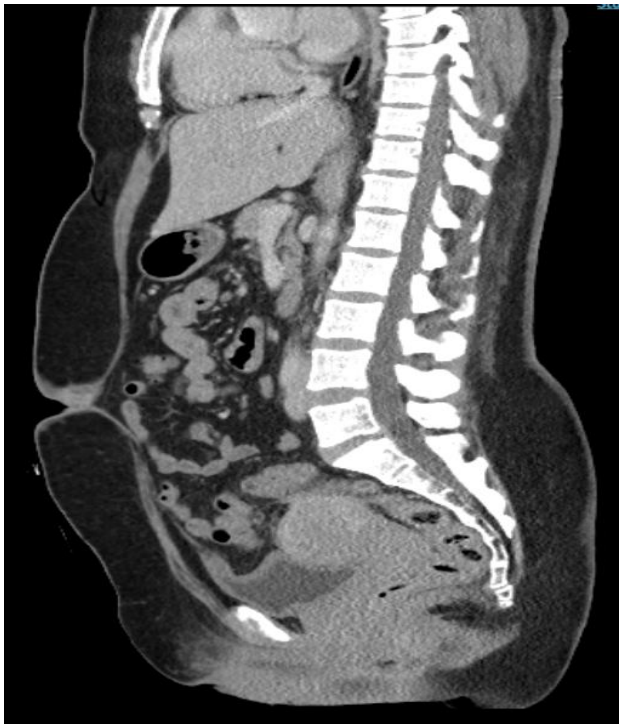

Figure 2: A sagittal plane CT scan showing the heterogeneous mass within the superficial subcutaneous fat in the deep portion of the umbilicus.

DISCUSSION

Endometriosis typically manifests within the pelvic region, affecting the ovaries, fallopian tubes, pelvic peritoneum, and the uterus's external surface. It can also develop in the abdominal wall at sites of previous surgical incisions, such as post-cesarean section or post-hysterectomy scars, and occasionally at the umbilicus. UE, also known as Villar's nodule, initially reported in 1886, is found in approximately 0.5-1% of all instances of endometriosis.[3] It typically presents as a painful, often cyclically tender, nodule in the umbilicus.[4] Two distinct types of UE are identified in the literature. Primary UE manifest in individuals without a history of abdominal surgery. They are hypothesized to result from the migration of endometrial cells via the abdominal cavity, lymphatic system, or embryonic remnants within the umbilical fold, along with genetic predisposition and immunological factors. Hildar et al recently reported the first case of primary umbilical endometrioma in a 40-year-old woman without a prior surgical history.[5] Secondary UE arise from scar tissue formation after abdominal procedures such as cesarean delivery and laparoscopy. Rahman et al described a trocar site endometrioma with cyclical bleeding after a laparoscopic gastric bypass.[6] Umbilical endometriosis has been reported recently after laparoscopic treatment of pelvic endometriosis.[7] Understanding the differences between primary and secondary development of UE is pivotal for comprehending their pathogenesis.[3]

The primary risk factor for developing an umbilical endometrioma in scar tissue is a history of obstetric surgical procedures, particularly cesarean sections. A 2007 report on 117 cases identified early hysterotomy, before the 22nd week of pregnancy, alcohol consumption, and increased menstrual flow as significant risk factors. Additionally, laparoscopic examinations and biopsy punctures have been linked to umbilical scar endometriomas. Less common surgical procedures, such as congenital inguinal hernia repairs, can also result in endometriomas at the incision site, sometimes mimicking hernia recurrence.[8]

From a clinical standpoint, umbilical endometrioma manifests as a reddish, purplish, or blackened nodule at the umbilicus, typically measuring between 0.5 and 3.5 cm in diameter. This condition often induces discomfort, swelling, and bleeding in the umbilical region. The presence of pain, often worsening with menstruation, and the physical finding of a mass are classic clinical findings of endometrioma.[3] Catamenial pneumothorax can also occur as a rare manifestation of extra-pelvic endometriosis, affecting approximately 7% of all cases of pneumothorax. Yu et al reported a 35-year-old Thai woman with umbilical pain and menstrual bleeding over three months, with subsequent identification of right pneumothorax on chest X-ray, leading to the diagnosis of catamenial pneumothorax.[9] Recurrent umbilical bleeding from an endometrioma was reported and erroneously presumed to be due to the patient's daily use of a cotton swab soaked with peroxide for umbilical cleansing.[10] While less common, these symptoms can provide valuable clues and should be considered in assessing patients with umbilical nodules or masses.

Less common locations of endometrioma include appendiceal endometriosis, first identified in 1860, presenting asymptomatically or with lower gastrointestinal bleeding.[11] A swelling in the right inguinal region with cyclical pain was described in a 40-year-old woman with endometrial glands and stroma found on pathology.[12]

Differential diagnoses of umbilical endometrioma include omphaloliths, which are hard, black stones within the umbilicus with pain, inflammation, and discharge. Umbilical abscesses and epidermoid cysts can all be associated with a palpable mass, redness, and pain. Lastly, dermatological conditions such as psoriasis or eczema of the umbilicus can cause irritation and discomfort. They rarely may be confused with endometriomas.[13] Our patient stated she had previously been given a diagnosis of umbilical hernia, as intermittent episodes of severe pain, against a backdrop of chronic discomfort, were presumed to be related to increased intra-abdominal pressure during menses. A delay in diagnosis is not unusual. A 36-year-old primigravid African woman with an umbilical lump that had bluish discoloration for whom a definitive diagnosis was obtained after two years.[14] Our patient had a painful umbilical mass for approximately five months before her first presentation to a surgical outpatient clinic in 2022, where she was diagnosed with an umbilical hernia, and the definitive diagnosis of umbilical endometrioma was made in 2024.

Imaging plays a crucial role in assessing endometriosis as it can visualize ectopic endometrial tissue, especially in cases of abdominal wall endometriosis. CT scans of abdominal wall endometrioma, though lacking specificity, typically reveal a solid-looking soft tissue mass near the surgical scar area, which might exhibit enhancement following the injection of intravenous contrast. This mass usually appears hyperattenuating compared to adjacent muscle tissue, although the degree of attenuation can vary. Depending on the blood supply to the endometrioma, it may contain blood products leading to differing attenuation levels, resulting in a heterogeneous appearance. CT imaging is beneficial for ruling out other diagnoses in abdominal wall masses, such as hernias, soft tissue tumors, and abscesses.[15] Our patient underwent a contrast-enhanced CT that revealed a heterogeneous mass within the superficial subcutaneous fat in the deep portion of the umbilicus. According to Chen et al susceptibility-weighted imaging (SWI) to look for signal voids in the cyst wall or within the lesion can facilitate the diagnosis of endometrioma. SWI has improved the ability of MRI to assess various diseases, especially hemorrhagic infiltrating lesions. Characteristics involve the accumulation of macrophages loaded with hemosiderin in the cyst wall and necrosis at the periphery due to recurrent bleeding. SWI represents a pioneering method highly attuned to blood degradation elements, manifesting as minute areas of signal voids.[16]

Endometriosis may be medically managed using hormonal therapies to suppress the growth of ectopic endometrial tissue or surgically managed by excising it. Specific to umbilical endometriosis, surgery is often required, especially for large endometriomas (greater than 4 cm), due to the risk of rupture or torsion. It is also indicated in infertile women who have exhausted medical management, such as in vitro fertilization and hormonal therapy. This requires meticulous planning and execution, including discussion about extensive surgical resection to ensure the complete removal of the endometriotic lesion with clear margins, minimize damage to surrounding tissues, provide satisfactory umbilicus reconstruction, and decrease the recurrence rate. Aesthetic considerations are paramount to reconstructing the umbilical area post-resection with a satisfactory cosmetic outcome without compromising the procedure's effectiveness. The excised tissue examination is crucial for confirming the endometriosis diagnosis and assessing the excision's completeness. Small asymptomatic endometrioma in patients older than 35 years old should not be treated surgically.[17]

Preventing recurrences of endometriosis, particularly after surgical removal, may involve ongoing medical

therapy to suppress the menstrual cycle. Hormonal therapy such as estrogen-progestin preparations, gestagens, progesterone-releasing intrauterine systems, and gonadotropin-releasing hormone agonists may be utilized. The two-component oral contraceptives are crucial in reducing pain, minimizing heavy menstrual bleeding, and eliminating retrograde menstruation, thereby preventing the spread of endometrial debris outside the uterus.[18] Although recurrence plays a major role in our consideration of treatment, the desire for pregnancy is also an important component. For patients who plan for future pregnancy after surgery, post-operative hormonal therapy may increase spontaneous pregnancy rates. However, hormonal therapy is correlated with an increase in recurrence versus surgical removal.[19] Patients who are infertile or older than 35 years old are suggested to undergo in vitro fertilization. If future childbearing is not considered, the most efficient recurrence prevention is unilateral oophorectomy with contralateral ovarian sparing.[18]

Two reported cases of endometrioma recurrence in a 30-year retrospective study of 55 patients with umbilical endometriosis.[3] A Japanese series of 90 patients over ten years reported two cases of malignant transformation, 1.34% recurrence at six months and 6.35% at 12 months.[20] In the literature, there are few noted treatments for recurrent endometriosis. In 2007, a case report noted chemical cautery may help cure small lesions in the setting of umbilical endometriosis status post laparoscopic therapy of pelvic endometriosis.[7] Dienogest (DNG) was also suggested. A multicenter data study revealed significantly improved pain relief, CA-125 levels, and size reduction of endometriomas with a mean treatment duration of 24 weeks.[21] In addition, while rare, cancerous degeneration of endometriosis has been reported, as illustrated by Hirata's findings of 0.02% malignancy.[20] Obata et al reported a case of clear cell adenocarcinoma arising from umbilical endometriosis in a 60-year-old woman who had undergone a hysterectomy for uterine myoma at the age of 38. They observed endometriosis adjacent to the clear cell adenocarcinoma, with histopathological evidence of transformation from endometriosis to carcinoma at the umbilicus. The carcinoma expressed HER-2 protein and exhibited strong mesothelial characteristics. Careful monitoring is recommended.[22]

CONCLUSION

UE presents a unique clinical challenge due to its rarity and potential for misdiagnosis, often requiring a multidisciplinary approach. Imaging modalities play a crucial role in identifying these lesions. Surgical excision remains the mainstay of treatment, particularly in patients seeking fertility or experiencing significant discomfort, with meticulous attention to complete removal and preserving cosmetic outcomes. Recurrence and malignant transformation of umbilical endometrioma are rare, but ongoing monitoring and consideration of adjuvant therapies are recommended.

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