Primary Abdominal Wall Endometriosis Masquerading as an Umbilical Hernia

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Background	An accurate diagnosis of abdominal wall endometrioma can be difficult to ascertain and can be easily mistaken for other more common surgical issues.
Summary	Despite its prevalence, endometriosis presents a diagnostic hurdle due to its varied presentation. Though it is a common condition, obtaining an accurate diagnosis can be anything other than straightforward. It is classified by the presence of endometrial tissue found outside the uterus. Endometrial deposits can be found throughout the abdomen and pelvis, as well as in more far-flung locations such as the lungs or abdominal wall. Its clinical presentation can be highly varied and vague. Common presenting symptoms include pelvic pain, dyspareunia, and infertility. Symptoms can be cyclical or entirely unrelated to menstruation. Conversely, some women are asymptomatic and can have the disease discovered incidentally. Finally, there are uncommon presentations that can further delay and complicate accurate diagnosis. Accurate diagnosis of endometriosis is important because the condition can cause significant distress to affected patients and has been linked to certain gynecologic malignancies. We present the case of a 31-year-old female who was referred to a general surgeon for a complaint of a painful umbilical mass. She was diagnosed with an umbilical hernia and elective repair was discussed. After being briefly lost to follow up, she returned months later, now with bleeding from the umbilicus in addition to pain. She was taken for diagnostic laparoscopy and a fat-containing incarcerated umbilical hernia was found. Separate from this, she was also found to have mass in the fascia and muscular tissue around the umbilicus. This was resected, and the surgical pathology ultimately returned as endometriosis. She had no other evidence of intra-peritoneal disease.
Conclusion	While this presentation is uncommon, similar instances of umbilical endometriosis have been described. As such, we feel that this case has value in reinforcing that common surgical issues like an umbilical her- nia can be complicated by and mask the presence of concurrent diagnoses. The general surgeon needs to consider all differential diagnoses when confronted with what seems like a common surgical problem.
Key Words	umbilical; hernia; endometriosis; mass; bleeding; abdominal wall

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Case Description

A 31-year-old woman presented in early 2021 for planned umbilical hernia repair due to intermittent umbilical pain. Upon physical exam, her abdomen was soft, not distended, and nontender, with signs suggestive of an incarcerated umbilical hernia containing fat. Notably, there was no evidence of active bleeding during the examination, but rather, irritation around the skin above the hernia was observed. The patient had a well-healed, low transverse incision from a previous C-section. An ultrasound conducted previously in the emergency department showed no presence of a mass in the area.

The patient was temporarily lost to follow-up as she pursued charity care but returned later that summer, reporting near-daily umbilical bleeding. Her medical history consisted of two cesarean sections and a childhood open appendectomy. Subsequently, she underwent a diagnostic laparoscopy and umbilical hernia repair in the operating room.

During laparoscopy, the surgeon identified an umbilical hernia sac containing omental fat. Additionally, a 2.2 × 2.0 cm mass was observed within the muscle above the fascia, separate from the hernia. This mass seemed to reduce through the hernia defect once the adhesions were taken down. It is likely that this was the mass palpated before the operation, initially mistaken for incarcerated hernia contents. The mass was resected with 0.5 cm margins of healthy tissue. Pathology confirmed nodular endometriosis with adjacent dermal scarring. No further endometriosis or C-section scar involvement was observed. The hernia defect was closed using a laparoscopic figure-of-eight suture passer and a 15 cm round mesh secured with tackers. The attending surgeon completed the case with assistance from a surgical resident. At the four-week follow-up, the patient was recovering well, with the resolution of her preoperative pain and bleeding.

Discussion

Endometriosis is a condition characterized by the presence of endometrial glands and stroma outside of the normal uterine cavity.¹ Women will often present with symptoms of pelvic pain, dysmenorrhea, or infertility. Despite its prevalence, endometriosis remains underdiagnosed and is a major cause of hysterectomy and hospitalization in the US, with a total annual societal burden estimated in 2009 at a staggering \$69.4 billion.² The mean latency from onset of symptoms to diagnosis is 6.7 years, in part due to the need for surgical pathology for definitive diagnosis.³ Endometriosis, characterized by the presence of estrogen-dependent tissue in ectopic locations, commonly affects the pelvic region, including the ovaries, uterine ligaments, pelvic peritoneum, and rectovaginal septum. However, in rarer cases, these tissues may be found in more distant locations, including the bowel, lungs, kidneys, and even the brain and anterior abdominal wall.⁴

Endometriosis pathogenesis remains a topic of debate. Sampson's theory, which is widely accepted, proposes retrograde menstruation as the culprit. Menstrual effluent and endometrial tissue flowing back through the fallopian tubes during menses implant in the peritoneal cavity. This is supported by a higher incidence of endometriosis in women with uterine outflow obstructions.^{5,6} However, Sampson's theory does not completely explain cases of endometriosis outside of the peritoneum⁴ or its presence in premenstrual girls.⁷

An alternative theory, coelomic metaplasia, proposes endometriosis develops because of metaplasia from cells lining the visceral and abdominal peritoneum. Given the shared embryologic cell lineage between the thoracic, abdominal, and pelvic peritoneum, this theory explains the presence of distant ectopic tissue sites.⁸

Sampson also suggested the theory of lymphovascular metastasis, indicating that endometrial cells could spread through lymphatic and hematogenous routes to various ectopic sites.⁹ However, while plausible, it is likely not the primary mechanism of ectopic tissue spread disease, given the relatively low incidence of extrapelvic endometriosis.

The prevalence of women with endometriosis can be difficult to assess. Affected women can have a wide array of presenting symptoms or be asymptomatic altogether. It is estimated that approximately 10% of all reproductive-aged women are afflicted in one form or another.¹⁰ Symptoms such as changes in the size of the lesion or bleeding can be cyclical with menstruation ¹¹ or occur independently of the menstrual cycle. It can also be diagnosed incidentally during abdominal surgery. In reproductive-aged women undergoing laparotomy or laparoscopy, intra-abdominal endometriosis is reported in 15% to 44% of cases.¹²

Endometriosis, while predominantly pelvic, can also manifest in extrapelvic sites. One such rare occurrence is abdominal wall endometriosis (AWE), which accounts for 0.3-1% of cases. Studies strongly link AWE to prior cesar-

ean sections,¹³ with a reported relative risk of 27 for its development post-cesarean.¹⁴ This association is thought to arise from possible exposure of endometrial cells and high blood volume to the subcutaneous tissue during the procedure, presenting later as implants in the surgical scars that form. However, the overall incidence of AWE after cesarean section remains low (0.26% over 25 years).¹⁵ Cutaneous endometriosis is another rare presentation (0.5% to 1%).¹² It has been suggested that the umbilicus, a physiologic scar, can serve as a nidus for endometrial tissue implantation, similar to scars from abdominal surgeries.¹¹ The formation of an endometrioma at the site of entry into the abdomen, a phenomenon known as scar endometriosis, occurred after surgery in 1.08% of patients undergoing mid-trimester abortion and 0.03% to 0.4% postcesarean section.12

This case presents a unique instance of abdominal wall endometriosis following two cesarean sections (4 and 6 years prior). Notably, the incisional scars from these procedures were not involved with her umbilical pathology. The mass, located within the abdominal wall fascia and subcutaneous tissues, caused progressive umbilical pain and, eventually, recurrent bleeding with increasing frequency. Interestingly, the presentation involved both the abdominal wall and skin, sparing prior surgical scars and the pelvis entirely.

Given their location and novelty, abdominal wall endometrial nodules can be mistaken for other pathology, including granulomas, melanomas, cysts, lipomas, hernias, or umbilical metastases from gastroenterological cancer (known as Sister Mary Joseph Nodule).¹² Literature suggests varying rates of concurrent pelvic endometriosis associated with abdominal wall endometriosis, estimated to be approximately 15–30%.¹⁶ Furthermore, women diagnosed with pelvic endometriosis are more likely to have a concurrent umbilical nodule. However, in this specific case, laparoscopy revealed no evidence of peritoneal disease.

Endometriosis diagnosis presents challenges due to the variability in symptom presentation and lesion appearance. While imaging modalities like ultrasound, CT, or MRI are used for initial evaluation, their accuracy is limited by the diverse characteristics of endometriosis lesions, which can range from cystic to solid or mixed. Additionally, blood tests like CA-125 may not provide definitive diagnostic value as elevated levels are nonspecific and lack diagnostic reliability.¹⁷

Consequently, a definitive diagnosis of endometriosis relies on pathological analysis of tissue obtained through surgical biopsy or excision.¹⁸ When it comes to treatment, surgical intervention with wide excision (1 cm margins) is generally preferred. This is because hormonal treatments, while commonly used for management, have high recurrence rates and androgenic side effects.¹²

A prior systemic review found recurrence rates ranging from 0–29% even after surgical excision of abdominal wall endometriomas.¹³ Long-term follow-up studies are crucial to definitively understand recurrence risks and guide proper patient counseling.

The diagnosis of endometriosis has important clinical and socioeconomic consequences. Despite being generally considered a benign condition, studies have linked endometriosis to ovarian clear cell carcinomas and ovarian endometrioid carcinomas.¹⁹ Ectopic endometrial tissue can also undergo malignant transformation itself.²⁰Additionally, women with endometriosis have a higher risk of systemic chronic inflammation, heightened oxidative stress, increased risk of coronary artery disease, infertility, increased healthcare costs, and lost productivity due to absenteeism.^{1,2,7,8,10,13,21}

Conclusion

After the discovery of the umbilical endometriosis, our patient has been recovering well. Follow-up revealed excellent recovery: complete resolution of preoperative bleeding and well-healed incisions. Notably, she lacked classic endometriosis symptoms, and the surgical inspection confirmed no pelvic lesions. Therefore, with complete excision of the umbilical mass and no evidence of further disease, her treatment is complete. However, she will be encouraged to follow up with her gynecologist and monitor for common symptoms and recurrent lesions.

While endometriosis most commonly presents with dysmenorrhea, infertility, and pelvic pain, these symptoms can be vague and unrecognized, thus prolonging diagnosis. An accurate diagnosis can be particularly challenging when a common condition presents unusually, as was the case here. Here, the challenge was compounded by an unusual presentation: a painful abdominal mass coexisting with an umbilical hernia. Surgical resection of the mass led to the definitive diagnosis of endometriosis upon pathological examination. Nonetheless, accurate endometriosis diagnosis is crucial due to its links with various comorbidities and its rare potential for malignant transformation. This empowers clinicians to effectively educate patients about their recurrence risk and the very low possibility of cancer development. This case serves as a reminder for general surgeons to maintain a broad differential diagnosis, even when presented with seemingly uncomplicated surgical cases.

Lessons Learned

This case highlights the importance of maintaining vigilance during surgery for general surgeons. Even when encountering seemingly routine cases, unexpected findings like endometriosis can arise. A thorough understanding of such gynecologic conditions is crucial, as they can present outside the expected purview. While generally considered benign, endometriosis holds a rare potential for malignant transformation and recurrence. Postoperative patient counseling and close follow-up are essential for optimal management.

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