Abdominal Wall Endometriosis

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KEYWORDS

- Abdominal wall endometriosis • Rectus muscle endometriosis
- Incisional endometriosis

KEY POINTS

- Prior abdominal surgery, specifically history of cesarean delivery, is the largest risk factor for abdominal wall endometriosis (AWE).
- Cyclic abdominal pain, a palpable mass, and history of abdominal surgery is the pathognomonic triad for the clinical presentation of AWE.
- Abdominal ultrasonography is the first-line imaging modality for AWE; however, MRI should be considered for nodules greater than 3 cm to guide surgical management.
- Surgical approach depends on lesion size and location, with an abdominal approach preferred for superficial disease and a laparoscopic approach preferred when the bulk of disease is subfascial or involving the rectus muscle.
- Following surgical excision, patients' symptoms improve with a low recurrence rate.

INTRODUCTION

Endometriosis, defined as the presence of endometrial glands and stroma outside the uterine cavity, occurs in up to 10% of reproductive age women.[1] The pathogenesis of endometriosis is likely multifactorial, including factors such as ectopic endometrial tissue, altered immunity, imbalanced cell proliferation and apoptosis, aberrant endocrine signaling, and genetics.[2] Abdominal wall endometriosis (AWE) is a specific type of extrapelvic endometriosis in which the glands and stroma of the endometrium are found within the layers of the abdominal wall. The incidence is reported to be 0.3% to 3.5%.[3–7] It is hypothesized that endometrial cells are transported and deposited into the abdominal incision via surgical instrumentation and proliferate within the abdominal wall.[8] However, this hypothesis does not account for all cases of AWE,

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because sporadic cases in patients with no prior surgical history are reported in the literature.[9–11] Alternative hypotheses of lymphatic or hematogenous dissemination may account for cases with no surgical history.[4]

CLINICAL PRESENTATION

Similar to pelvic endometriosis, the diagnosis of AWE requires a high level of clinical suspicion. Patients are of reproductive age and present with a single mass.[12] The mean time from initial surgery to symptomatic AWE ranges from months to years.[4,5,12] Cyclic abdominal pain, a palpable mass, and a history of abdominal surgery is the pathognomonic triad, but individual cases may vary in presentation.[13] Up to 90% of patients will report cyclic abdominal pain as their primary symptom.[4,12,14] Cyclic symptoms are caused by fluctuations in the menstrual cycle with hormonal stimulation driving cell proliferation, swelling, and bleeding.[15] Patients also experience noncyclic pain and focal pain with palpation of a mass. Not all cases present with a palpable nodule, and the physical examination can be limited by body habitus. New research suggests that body mass index is an additional risk factor for AWE, making the clinical diagnosis of AWE all the more challenging.[4,16]

Focal abdominal wall tenderness close to a prior incision should increase the clinical suspicion for AWE because 80% to 90% of cases are associated with a surgical scar.[4] More than 80% of patients with AWE have a history of a cesarean delivery, and the apices of cesarean incisions are the most common location for implants. The implant can appear distant from the Pfannenstiel skin incision secondary to the lateral and superior extension of the fascia relative to the skin during cesarean delivery.[17] AWE located in surgical scars is reported after other pelvic surgeries such as abdominal myomectomies, hysterectomies, laparoscopic pelvic surgery, appendectomies, and hernia repairs.[4,18] Not all AWE cases have a history of prior surgery.[9–11,19,20] Common sites of spontaneous AWE include the groin/inguinal (58%), umbilicus (36%), and less commonly the rectus muscle (3%).[21,22] AWE that occurs in the absence of prior abdominal surgery is associated with pelvic endometriosis.[4] A thorough history followed by a complete abdominal, pelvic, and rectal examination is necessary to determine the extent of pelvic endometriosis in cases of sporadic AWE.

Lack of a palpable abdominal mass on examination does not exclude AWE and could suggest subfascial disease involving the rectus muscle. Rectus muscle endometriosis (RME) is a specific subtype of AWE in which ectopic endometrial tissue is located within the body of the rectus muscle.[20] RME can be isolated to the rectus muscle or can be an extension of AWE involving the subcutaneous tissue and fascia. In a single-institution case series of RME, Melnyk and colleagues[19] reported that whereas 91% had focal abdominal tenderness on examination, only 54% had palpable nodules, reflecting the subfascial location of the disease. Patients also reported pain with movements that strain the rectus muscle such as coughing, sneezing, or performing a sit-up, which can help differentiate subfascial from superficial disease.[19]

DIAGNOSIS

The differential diagnosis of AWE includes incisional hernias, lipomas, abscesses, granulomas, cysts, sarcoma, and scar tissue.[5,13,23] AWE should be included in the differential diagnosis of women with a symptomatic mass even without a history of biopsy-proven endometriosis. Final diagnosis is made after pathologic examination, but imaging is useful to narrow the differential.

Imaging

Imaging eliminates other causes of an abdominal wall mass and provides the clinician with information on the size, depth, and extent of fascial involvement to guide surgical planning.[14] First-line imaging is abdominal wall ultrasonography because this is a low-cost, readily available imaging modality. Abdominal ultrasonography can also exclude abdominal wall hernias.[23] On ultrasonography, AWE appears cystic, solid, or mixed (**Fig. 1**) and power Doppler demonstrates internal vascularity.[6,8,23] In a series of 151 cases of AWE, Zhang and Liu[18] reported a preoperative detection rate of 97% with abdominal ultrasound. Abdominal ultrasonography can distinguish subcutaneous and subfascial disease, with a reported 92% sensitivity identifying endometriosis involving the rectus body.[6] Zhang and colleagues reported a lower rate of accuracy for depth of invasion at 26.5% and described lower accuracy for size of the lesion.[18] Therefore, for AWE lesions 3 cm or larger or nodules that are difficult to palpate, the authors recommend MRI to define the location and depth of the endometriosis nodule, providing a map for surgical intervention. The authors also recommend MRI if the patient has no history of abdominal surgery, because these patients have increased rates of concurrent deep infiltrating endometriosis of the pelvis.

On MRI, AWE implants are solid hypointense or slightly hyperintense on T2- and T1 fat-suppressed images with small areas of hemorrhage appearing hyperintense compared with the surrounding abdominal wall (**Fig. 2**).[24] Recent hemorrhage displays as high signal intensity on T1- and T2-weighted images, and hemosiderin deposits show low signal intensity on T1- and T2-weighted images.[8] Various protocols for AWE are reported in the literature without consensus for the role of diffusion-weighted images and intravenous (IV) contrast.[24] Computed tomography (CT) has less clinical benefit. On CT with IV contrast, AWE typically enhances; however, the exact characteristics depend on the menstrual cycle phase, proportion of stromal and glandular tissue, and extent of surrounding inflammation and fibrosis.[8] CT findings are generally nonspecific, and we do not recommend their routine clinical use for the workup of suspected AWE.

Ultrasound-guided fine-needle aspiration (FNA) can be used to confirm suspected AWE as an alternate to surgical removal. FNA is a minimally invasive, rapid procedure that can exclude malignancy before planned surgical intervention.[6] Abdominal wall hernia must be ruled out with ultrasonography before performing FNA. Histologic confirmation can be inconclusive due to fibrosis and limited tissue available for sampling.[6,25,26] In one large study, 75% of cases were inconclusive.[25] We recommend

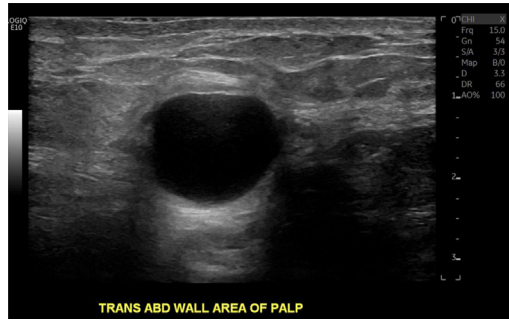

Fig. 1. Transabdominal ultrasonography with hypoechoic nodule in the cesarean delivery scar. Final pathology demonstrated abdominal wall endometriosis (AWE).

Fig. 2. AWE in the cesarean delivery scar on MRI.

FNA as an additional diagnostic tool when a patient's history and imaging is not conclusive or concerning for malignancy.

MANAGEMENT

Expectant

Patients who are asymptomatic or whose AWE is an incidental finding may be managed expectantly. We do not recommend routine imaging surveillance for asymptomatic individuals. Additional workup or treatment can be initiated in response to new symptoms.[27]

Medical

Medical management can be offered to patients as a conservative approach. Continuous combined oral contraceptive pills (OCPs), progestins, or Gonadotropin-releasing hormone agonists suppress ovulation and produce a hypoestrogenic state; this can relieve symptoms but does not decrease the size of lesions.[3] The overall success of medical management is temporary, with symptom recurrence following drug discontinuation. Medications can be used temporarily as a bridge to surgery or menopause to optimize symptom relief.

Ablation

Recent small, retrospective studies demonstrate the safety and feasibility of high-intensity focused ultrasound (HIFU) ablation for AWE.[28] Ultrasound-guided HIFU ablates AWE through coagulative tissue necrosis caused by ultrasound pressure-induced heat in the focused area.[28] The 6- and 12-month outcomes show decreased lesion size and lower pain scores with no differences in pain-free relief between patients treated with HIFU compared with those treated with surgical excision.[29,30] Advantages of HIFU include shorter hospital stays, no structural disruption to the abdominal wall layers, and minimal blood loss.[30] Thermal ablation with CT- or ultrasound-guided cryoablative needles is also described in the literature.[31,32] Long-term symptomatic relief from this procedure remains unknown with only 1 nonrandomized study comparing outcomes

with surgical excision.[31] Both techniques are promising for the nonsurgical management of AWE; however, large prospective comparative studies are necessary.[3]

Surgical

Surgical excision remains the standard of care for the treatment of symptomatic AWE. Endometriosis nodules are removed via wide local excision and an abdominal incision. Experts recommend surgical margins of at least 1 cm to ensure complete disease resection.[7,17] During surgery, the endometriosis is easily delineated from the surrounding abdominal wall based off texture and appearance. The firm, fibrotic tissue is typically pink and can have cystic areas of blue, purple, or brown (**Fig. 3**). About 60% to 70% of reported cases involve the fascia, which requires repair after excision.[4,12,14] As described later, primary fascial repair is performed for small defects (<3 cm). Experts recommend fascial repair with mesh for lesions greater than 3 cm.[3,17] Preoperative imaging is crucial to determine the size and location of the endometriosis, allowing for a planned multidisciplinary approach with general or plastic surgery for large nodules involving the fascia. If RME is less than 2 cm and without obvious physical landmark such as symphysis pubis, umbilicus, or adherent uterus for reference, ultrasound-guided needed localization can be helpful to locate the nodule and minimize dissection.

Ecker and colleagues[22] first reported a laparoscopic approach to AWE with a focus on subfascial lesions. Melnyk and colleagues[19] further described this technique in her case series of surgically managed RME. The advantages of minimally invasive surgery, including less postoperative pain, lower infection rates, shorter hospital stay, and quicker return to normal activities, are additional benefits of laparoscopy over an abdominal approach.[33] The laparoscopic approach also provides an opportunity for the treatment of pelvic endometriosis if present and the patient is symptomatic. We suggest the following approach to surgical excision. If most of the endometriotic nodule is within the subcutaneous tissue, an abdominal approach is preferred. When the bulk of the disease is subfascial and involving the rectus body, a laparoscopic approach is most appropriate.[33] The surgical procedure is billed according to the corresponding Current Procedural Terminology codes (**Table 1**).

Fig. 3. AWE nodule after surgical excision.

Table 1
Current procedural terminology codes for surgical management of abdominal wall endometriosis

CPT Code	Description
22902	Excision of abdominal wall, subcutaneous, <3 cm
22903	Excision of abdominal wall, subcutaneous, 3 cm or greater
22900	Excision of abdominal wall (subfascial), <5 cm
22901	Excision of abdominal wall subfascial tumor, 5 cm or greater

SURGICAL TECHNIQUES

Surgical Excision via an Abdominal Incision

The nodule is palpated and marked in the preoperative area, allowing for patient input and identification. The patient is placed in a supine position. The abdomen is prepped in the usual sterile fashion. The nodule is again palpated, and the planned incision is marked directly over the implant. A horizontal skin incision is made, approximately 1 to 2 cm larger than the size of the nodule to allow for dissection.[17] The subcutaneous layers are dissected until the nodule is identified, constantly palpating the lesion to guide dissection. Once the nodule is exposed, it is grasped with a clamp to provide elevation and countertraction. A stay suture is an alternative option for retraction. Using electrocautery, the disease is sharply dissected from the surrounding abdominal wall layers in a circumferential manner. During dissection, a curved hemostat can be helpful to delineate the fibrosis from the surrounding tissue.[19] Tactile feedback identifies the margins of the nodule, allowing for complete resection. Once the lesion is removed, the fascia is inspected. We recommend a tension-free fascial closure using delayed absorbable monofilament suture. When the fascial defect is too large for a tension-free repair, general surgery or plastic surgery should be consulted for abdominal wall reconstruction with or without mesh. The subcutaneous space is irrigated and closed if the depth of the defect is greater than 2 cm to prevent seroma and hematoma formation. Local anesthesia is infiltrated in the incision, and patients can be discharged home the same day.

Surgical Excision via Laparoscopy

The location of the nodule is palpated and marked in the preoperative area, allowing for patient input and identification. Laparoscopic entry into the peritoneal cavity is standardly performed. A 45°-angled laparoscope improves visualization of the anterior abdominal wall. Using tactile palpation, anatomic landmarks, and knowledge from preoperative imaging, the endometriosis nodule is identified. A peritoneal incision is made with electrocautery at the site of the endometriosis nodule in the anterior abdominal wall. The incision is extended with careful avoidance of the inferior epigastric vessels and the bladder. If necessary, the space of Retzius is dissected to separate the bladder from the abdominal wall endometriosis nodule.[19] The nodule is grasped from the assistant ports, and downward traction delineates the borders (**Fig. 4**). Using a mixture of sharp and blunt dissection, the nodule is progressively dissected from the surrounding abdominal wall using monopolar energy or an advanced energy device. Tactile feedback delineates the fibrosis from the normal tissue in a circumferential fashion. After complete enucleation, the fascia is carefully inspected for defects. Small defects (<3 cm) are repaired with interrupted delayed absorbable sutures using a fascial closure device (**Fig. 5**).[19] Melnyk and colleagues[19]

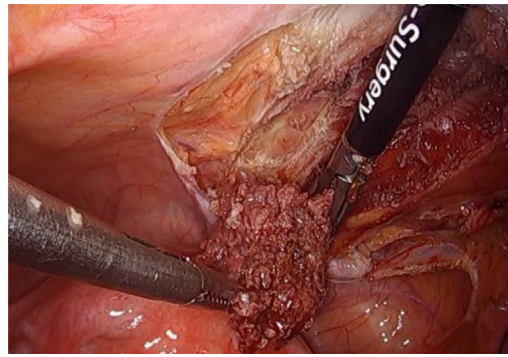

Fig. 4. During laparoscopic excision of AWE, downward traction on the lesion from the assistant port helps delineate the borders of the lesion to facilitate dissection.

first described repairing larger defects (>3 cm) with mesh and absorbable tacks. A multidisciplinary approach with general surgery, particularly a laparoscopic hernia specialist, is helpful for complex laparoscopic repairs and reconstruction.

Surgical Management of Pelvic Endometriosis

Laparoscopic evaluation of the pelvis for concurrent endometriosis should be considered in patients who present with pelvic symptoms. The reported rates of coexisting pelvic endometriosis with AWE range from 5% to 34%.[4,5,14,19,34–37] Patients with AWE and no prior surgery have a higher likelihood of having pelvic endometriosis, specifically deep infiltrating disease. Marras and colleagues[4] found that patients with concurrent pelvic endometriosis were more likely to be nulliparous, present with smaller nodules, and have nodules located in the umbilicus and suprapubic region. This study also reported that AWE associated with a cesarean delivery scar is less likely to have concurrent pelvic disease.[4] Pelvic endometriosis, when present, frequently involves the bladder (**Fig. 6**). In one series, 70% of patients who underwent laparoscopic excision of RME had concurrent pelvic endometriosis with the most common location being the bladder or ureter (60%).[19] The anatomic relationship between the anterior abdominal wall and the bladder is altered with cesarean deliveries, which could represent the reason for the high incidence of genitourinary disease. Another hypothesis is

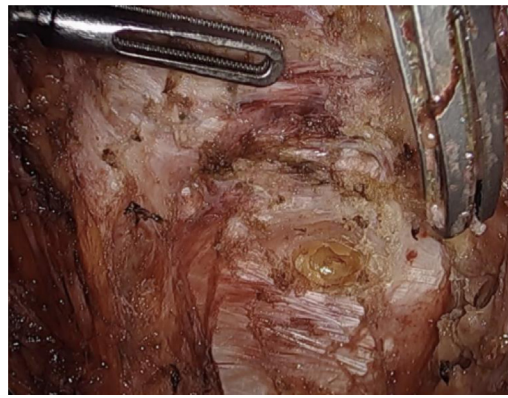

Fig. 5. Small fascial defect is noted after laparoscopic excision of AWE.

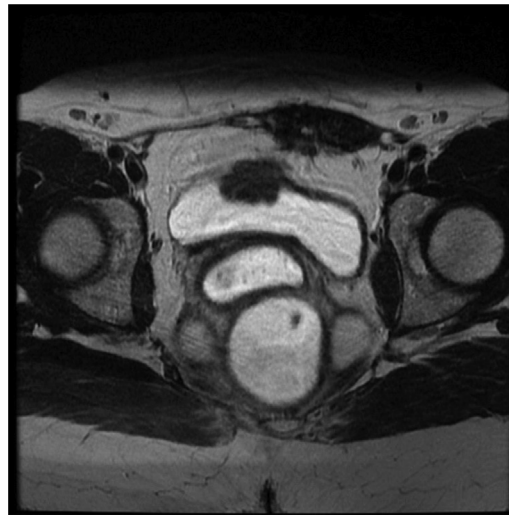

Fig. 6. MRI image with AWE and concurrent bladder endometriosis nodules.

that endometrial cells are seeded into the bladder when dissecting the bladder flap during a cesarean delivery. We recommend surgical exploration of the pelvis in patients with AWE who present with symptoms of endometriosis such as dyspareunia, dyschezia, and/or pelvic pain, with a heightened clinical suspicion in patients with spontaneous AWE, because their incidence of concurrent pelvic endometriosis is higher than the general population.[21]

Treatment Outcome

Surgical management of AWE results in symptom relief with most studies reporting greater than 90% complete relief on short-term follow-up.[12,18,19,21,38] Recurrence risk ranges from 4.3% to 11%, and recurrences can be minimized by ensuring adequate margins during surgery.[4,12,21,38] Recurrences typically occur years later, and only one study demonstrated a lower recurrence rate in patients treated with hormonal medications compared with those not on suppression.[4,12,18] Malignant transformation of AWE is extremely rate with a reported incidence of 0.3% to 1%.[39] The evidence is limited to case reports, with 46 cases described in the literature. Two-thirds of malignancies are clear cell carcinoma.[3,39] Compared with benign lesions, cancerous masses are larger at diagnosis with a median diameter of 9 cm and present at a later interval, with median time of 18 years from initial surgery to diagnosis.[39,40] The clinical presentation is otherwise similar to that of benign AWE.[39–42] In cases of suspected malignancy, we recommend early referral to gynecologic oncology because these cancers are aggressive and associated with poor clinical outcomes.

Prevention

There is no evidence that specific surgical techniques prevent incisional seeding during abdominal surgery and the subsequent development of AWE.[3] Suggested strategies include irrigation of the abdominal incision, changing gloves before fascial closure, and using fresh surgical instruments to close the fascia.[12] Although these strategies may reflect best surgical practice, there is no evidence that they will prevent the development of AWE.[3,17] Because greater than 80% of cases of AWE are

associated with a cesarean delivery, preventing the primary cesarean delivery may decrease the prevalence of subsequent AWE.

DISCUSSION

AWE is a rare form of endometriosis with which many providers are unfamiliar, resulting in a delay in diagnosis and patient suffering. Patients experience symptoms for 1 to 3 years before surgical treatment.[14,18,36] The clinical presentation of cyclic focal abdominal pain adjacent to a cesarean delivery scar is pathognomonic, yet in patients with this presentation Andres and colleagues[21] cited a preoperative clinical diagnosis of only 39%. A heightened clinical suspicion is crucial to making the diagnosis, and education should be directed to general surgeons and primary care providers, because up to 70% of patients with AWE are referred to general surgery.[21]

Experts differentiate between the 2 types of AWE based on pathogenesis and clinical presentation. The most common presentation of AWE is following major abdominal surgery, typically a cesarean delivery. Endometrial cells are iatrogenically transplanted into the surgical incision, and over time the endometriosis grows and proliferates in response to hormonal stimulation. The incidence of AWE following a cesarean delivery is 0.5% to 1% and is not related to the number of prior cesarean deliveries, timing of delivery, or reason for cesarean delivery.[3,4,22] AWE after cesarean delivery presents a singular nodule in the apices of the Pfannenstiel incision without concurrent pelvic endometriosis. Although cesarean delivery is the most common preceding surgical procedure, AWE can grow in scars from other pelvic surgeries including trocar sites from laparoscopic surgery.[14]

Spontaneous AWE that occurs in the absence of a prior surgical incision is the rarer type of AWE and most commonly presents in the groin or at the umbilicus. Endometrial cells are transplanted via lymphatic drainage from the pelvis to the groin where they implant and grow. The pathogenesis of spontaneous umbilical endometriosis is less clear and hypothesized to be secondary to vascular or lymphatic spread to the narrowest portion of the abdominal wall.[22] Ecker and colleagues[22] reported that patients presenting with no prior surgical history had higher rates of dysmenorrhea and were more likely to be nulliparous compared with AWE following cesarean delivery. Concurrent pelvic endometriosis, specifically deep infiltrating endometriosis, is more common in this population and likely contributes to the pathogenesis.

Surgical excision is standard of care for both variations of AWE, and preoperative imaging informs surgical planning. Abdominal ultrasonography is the first-line image modality to confirm the diagnosis and exclude other causes of an abdominal wall mass. MRI is recommended for patients with larger lesions and in patients in whom suspected AWE is difficult to palpate on physical examination. Surgical approach should be determined by the size, location and layers of the abdominal wall involved, and with surgeon comfort. The authors recommend a laparoscopic approach when the bulk of disease is subfascial and involving the rectus muscle. The traditional abdominal approach is preferred if the lesion is located in the subcutaneous tissue. For both surgical techniques, preoperative collaboration with a multidisciplinary team including general or plastic surgery is recommended for lesions with greater than 3 cm of fascial involvement. Two-thirds of AWE involves the fascia, and mesh is necessary in up to 10% of cases.[17] Larger size and deeper nodules invading the rectus are associated with higher rates of recurrent disease, and obtaining a 1 cm margin is the recommended practice to reduce recurrence.[17,21] Careful preoperative planning allows for complete surgical resection. Following surgery, patients have good short-term pain relief, but long-term outcomes are not published in the literature.

With the increasing rate of cesarean deliveries worldwide, the incidence of AWE will likely increase and continue to represent a diagnostic challenge for clinicians. Therefore, physician education regarding clinical presentation and management of AWE is increasingly important. The available literature is low quality, because most publications are retrospective single-institution case series and expert opinion with short-term follow-up. Andres and colleagues[21] suggest establishing a worldwide registry to improve evidence-based care of this disease. This registry would allow for prospective comparative research studies to answer important clinical questions.

CLINICS CARE POINTS

- Prior abdominal surgery, specifically a cesarean delivery, is the largest risk factor for AWE, with 80% of cases associated with a prior surgical scar.
- The pathognomonic triad of AWE is cyclic abdominal pain, a palpable mass, and a history of abdominal surgery.
- Abdominal ultrasonography is the first-line imaging modality for AWE, with preoperative detection rates as high as 97%.
- MRI should be considered for lesions greater than 3 cm to inform surgical planning.
- Medical management with OCPs, progestins, or GnRH agonists can be considered for temporary symptom relief but will not reduce lesion size.
- Surgical excision with 1-cm margins is standard of care for treatment of AWE.
- AWE involves the fascia in two-thirds of cases, and a multidisciplinary surgical team is necessary when the fascial defect is greater than 3 cm to perform abdominal wall reconstruction with or without mesh.
- Surgical management results in symptomatic relief, with reported rates of complete relief at short term follow-up as high as greater than 90%.

DISCLOSURE

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