

Clinical features, diagnosis, and management of abdominal wall endometriosis: a review

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Abdominal wall endometriosis is characterised by presence of ectopic endometrial tissue in the subcutaneous and muscle layer of the abdomen. It is usually related to previous surgical scars (commonly of Caesarean section). This article aims to review the pathogenesis, clinical features, diagnosis, and treatment of abdominal wall endometriosis.

Keywords: Abdominal wall; Cesarean section; Endometriosis; Pelvis

Introduction

Endometriosis is characterised by uterine endometrial mucosal tissue found outside the uterus¹. It usually involves pelvic organs, but 9% to 15% of cases involve extraperitoneal regions², including the bowels, the ureter, and the lungs. Abdominal wall endometriosis (AWE) is defined as the presence of ectopic endometrium between the skin and parietal peritoneum³. AWE is very rare and usually related to Caesarean section and pelvic surgeries. With the increasing trends of Caesarean section rates, the frequency of abdominal wall endometriosis is expected to rise, and it is useful for gynaecologists to be familiar with this condition.

Epidemiology

AWE is likely to be underreported owing to its rarity. The true prevalence is unknown and is estimated as 0.03% to 1%⁴. The mean patient age at presentation is 31.4 (range, 29.1-33.8) years⁵. AWE can be of primary or secondary origin. Primary AWE is not caused by surgery and accounts for around 20% of all cases; the location of ectopic tissue is often at the umbilical or groin area. Secondary AWE is associated with prior surgery and accounts for >70% of all cases, with >50% of cases relating to Caesarean section⁵.

Pathogenesis

The exact pathogenesis of AWE remains unknown. The most accepted theory to explain the formation of AWE is the direct implantation theory^{5,6}. It states that endometrial cells seed during pelvic surgery and are transported to ectopic sites. The endometrial cells then proliferate to form endometrioma. Another theory is lymphatic or haematogenous spread of endometrial cells, which may lead to deposition at scar region. This can explain the occurrence of AWE in patient without prior surgical

history⁶. In addition, there is the theory of metaplasia of abdominal wall cells into endometrial tissue under the influence of hormones⁷.

Risk factors

Prior history of abdominal or pelvic surgery is the greatest risk factor for the development of AWE. Horton et al⁵ reviewed 445 cases of AWE and reported that 57% of the cases had a prior Caesarean section and 11% had prior hysterectomy; the mean interval from index surgery to presentation was 3.6 (range, 2.5-4.8) years. Khan et al⁸ reported that body mass index was higher in the 34 patients with AWE than controls. It is hypothesised that suboptimal closure of the uterine incision or abdominal layers owing to obesity contributes to the development of AWE. Pelvic endometriosis is also a risk factor for the development of AWE. Horton et al⁵ reported that 13% of AWE has concurrent pelvic endometriosis and such incidence is similar to that of the general population (8% to 15%).

Pathology

Depending on the location at abdominal wall layers, AWE can be superficial (affecting subcutaneous tissue only and above the fascia), intermediate (infiltrating rectus muscles fascia), and deep (affecting rectus muscles)⁹. Endometriotic tissue can appear as a bluish, dark red, or black cyst or nodule with brown material, distinguishing itself from surrounding yellow subcutaneous fat. It has a hard consistency and irregular surface when found in muscles¹⁰. Microscopy shows the presence of endometrial glands, stroma, or haemosiderin pigment.

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Diagnosis

Typical presentations of AWE comprise a triad of prior history of Caesarean section, cyclical pain localised at the site of the lesion associated with menstruation, and presence of a mass lesion near a surgical scar¹¹. However, only 60% of patients demonstrate this triad of presentations¹². Abdominal mass (96%) and pain in the mass (87%) are the most common symptoms, whereas cyclic pain occurs in only 57%⁵. Patients may also complain of increase in size, bleeding, and skin discoloration of the mass in relation to menstruation².

On physical examination, there is an immobile abdominal mass that can be tender upon palpation, and the overlying skin may show discolouration¹³. The mass is usually located cephalad and lateral to the Pfannenstiel scar in Caesarean section-related cases, because the facial incision is often extended more lateral and cephalad than the skin incision³.

Careful history taking and physical examination is essential for diagnosing AWE. It is estimated that 20% to 50% of scar endometriosis are correctly diagnosed preoperatively^{2,14}. Diagnosis is difficult when the mass is not palpable or presentation is atypical such as a mass without cyclical pain. The differential diagnoses include non-tumoural lesions (hernia, granuloma, haematoma, abscess, fat necrosis), benign neoplasms (lipoma, neuroma, desmoid tumour), malignant neoplasms (carcinomas, melanoma, sarcoma, metastasis) and secondary tumours (Sister Mary Joseph node).

Imaging modalities aid the diagnosis and facilitate surgical planning, especially for large AWE in which mesh placement for large fascial defects or complex abdominal closure may be required.

Ultrasonography

Ultrasonography is used to confirm the presence of lesion, to assess its size, content, location, and margin, and to differentiate cystic from solid masses. AWE usually appears as solid heterogeneous hypoechoic masses with ill-defined and irregular margins^{15,16} (Figure 1a). Echogenic spots (haemorrhage) or thick echogenic strands (fibrosis) can also be seen, depending on menstrual phase of patient¹⁷. A hyperechoic ring at the periphery of lesion represents inflammatory changes of adipose tissue¹⁸. Vascular pattern of lesion varies with the size of AWE. Lesions >15 mm are found to have intralesional vascularisation that can be demonstrated by Doppler velocimetry¹⁹ (Figure 1b). However, these findings are non-specific for AWE.

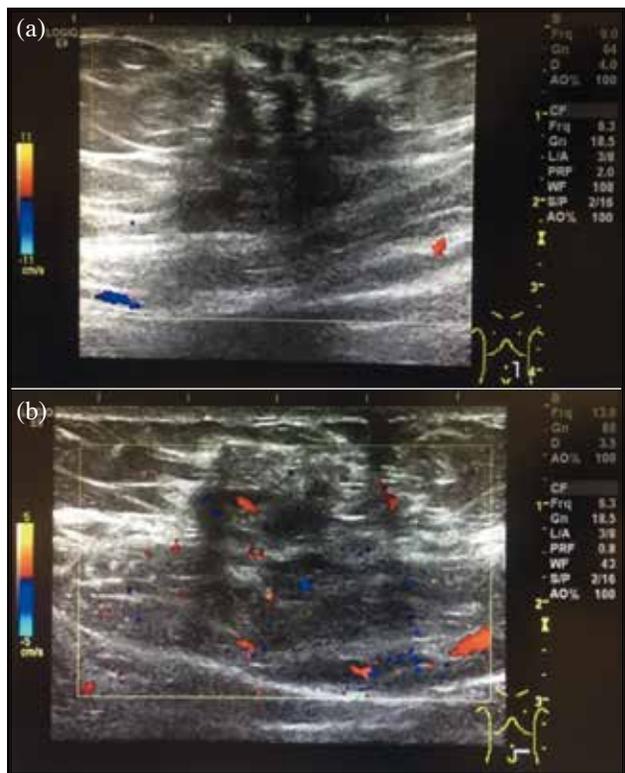


Figure 1. (a) A heterogeneous hypoechoic mass with ill-defined and irregular margins; and (b) intralesional vascularisation on Doppler ultrasonograph.

Nonetheless, ultrasonography is low cost, non-invasive, and radiation-free. Three-dimensional ultrasonography is more useful to demonstrate the depth of infiltration of the mass and the relation to surrounding tissues^{20,21}.

Magnetic resonance imaging

Magnetic resonance imaging is the modality of choice to assess soft-tissue mass. AWE appears as hyperintense heterogeneous mass on T1-weighted (with or without fat suppression) and T2-weighted images (Figure 2). For chronic scar endometriosis, lesions have speculated margins and low-signal intensity on T2-weighted images owing to its fibrotic component^{22,23}. The chronicity of the haematoma is demonstrated by the presence of haemorrhage inside the lesion²³. Advantages of magnetic resonance imaging include clearer delineation of subcutaneous tissues and muscles, more accurate assessment of the location and depth of infiltration of AWE, no ionising radiation, and the ability to detect small lesion²².

Computed tomography

AWE appears as a solid soft-tissue mass with mild to moderate contrast enhancement^{17,23}, depending on the phase of menstrual cycle, degree of fibrosis, bleeding, and inflammatory response (Figure 3). Feeding vessels may also be seen within or near the lesion^{16,23}.

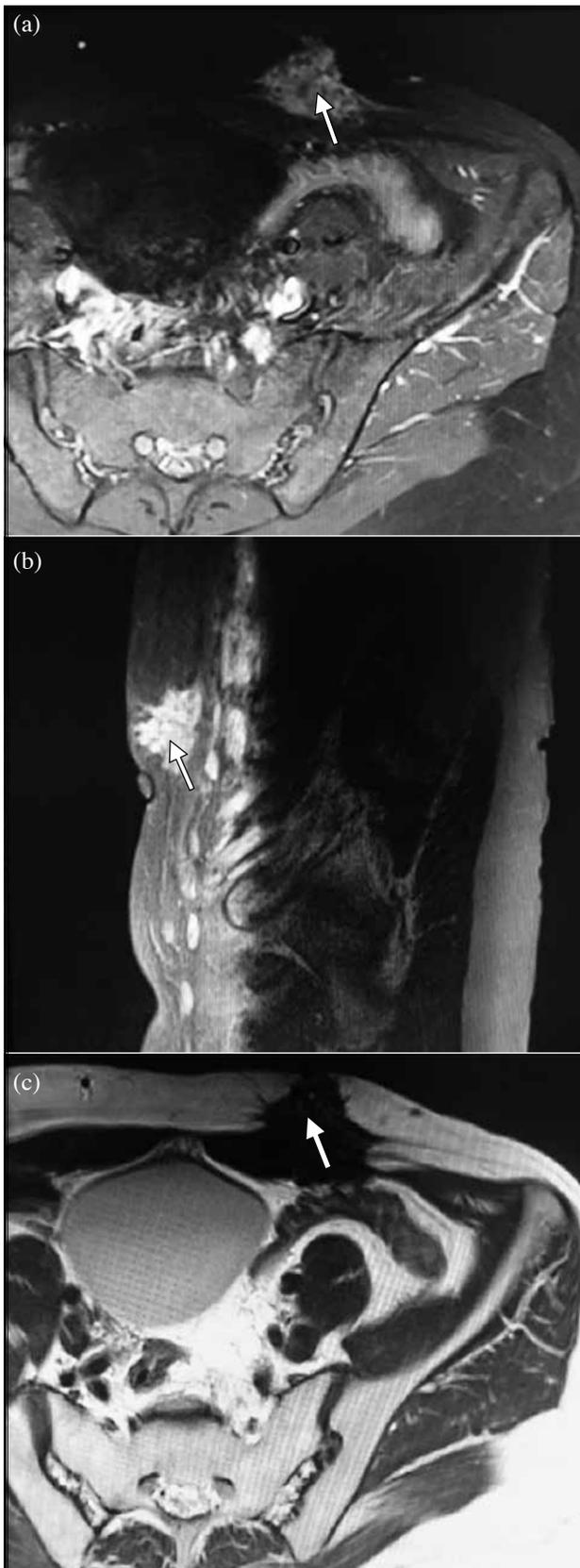


Figure 2 (a) Axial T1-weighted image showing heterogeneous hyperintense lesion (arrow); (b) sagittal T1-weighted image showing enhancement of the lesion with contrast (arrow); and (c) T2-weighted image showing speculated margins and low-signal intensity (arrow).



Figure 3. Axial contrast-enhanced computed tomography showing speculated irregular soft-tissue mass in abdominal wall.

Fine needle aspiration

Ultrasound-guided fine needle aspiration is used to confirm the diagnosis of AWE and to exclude malignancy. Incisional hernia must be ruled out before aspiration. However, its use for the diagnosis of AWE is controversial because of the risk of spreading endometriosis at the puncture site. It is advisable to include the biopsy tract in the field of resection intra-operatively²⁴. In old AWE lesions with large fibrotic content, fine needle aspiration may not yield enough tissue for sampling and lead to inconclusive results. Histologic biopsy may be required in such cases.

Risk of malignancy

Malignant change in abdominal wall endometriosis is rare and estimated to be 0.3% to 1%¹⁷. Risk factors include advanced age, postmenopausal, and lesion diameter >9 cm²⁵. Malignancy should be suspected in cases with multiple recurrences, lack of response to treatment, and sudden rapid growth²⁶. Clear cell carcinoma is the most common histological subtype²⁷. Wide excision with clear margins is a preventive option.

Management

Wide local excision with negative margins is the treatment of choice for AWE, as it provides both definitive diagnosis and treatment, with a success rate of 95%. Complete excision of the lesion and adjacent fascia (with a clear margin of at least 1 cm on all sides of the lesion) is important in reducing the chance of recurrence¹⁰. Inadequate resection results in around 9% of recurrence². Large lesions and involvement of rectus muscles are associated with a higher recurrence rate²⁸. So far, no study

has assessed the relationship between the size of surgical margin and the recurrence rate.

Small lesions located at subcutaneous layer can be removed easily, whereas large infiltrating lesions extending to aponeurosis, muscles, or even peritoneum are technically difficult to be excised. A large fascial defect may require placement of a mesh or constructing an aponeurotic muscle flap to cover the defect, and the procedure is usually performed by general surgeons²⁹. For large and deep lesions, preoperative assessment with magnetic resonance imaging or computed tomography and planning with the general surgeon is essential. Surgery should be performed at the end of menstrual cycle, as the lesion is minimal^{12,30}. For lesions with ill-defined borders, frozen section can be obtained intra-operatively to ensure adequate negative margin and minimise resecting unaffected tissues.

High-intensity focused ultrasound ablation is non-invasive and has favourable outcome, although pathological diagnosis of the lesion is not feasible. Ultrasound wave is used to induce coagulative necrosis in targeted endometrial tissues. In 51 women followed up for 4 years, high-intensity focused ultrasound ablation resulted in reduction of pain at the mass to visual analogue score 1 and significant reduction of lesion volume to 25% at 1 month, although one case had first-degree skin burn and the overall relapse rate was 3.9%³¹.

Percutaneous cryotherapy administers tissue ablative freezing temperature to induce tissue necrosis by inserting a cryoprobe into the lesion. Maillot et al³² compared outcome of surgical excision versus cryotherapy and reported similar pain relief and lesion size reduction. Cryotherapy preserves abdominal wall integrity and function and has better cosmetic outcome, compared with surgery. However, it is not suitable for large and deep AWE. More prospective studies with larger sample size are required to establish the effectiveness and safety of high-intensity focused ultrasound ablation and percutaneous

cryotherapy.

Medical treatment is not effective for AWE. Oral contraceptives, progesterone, and Danazol result in improvement on symptoms only but not resolution of the lesion^{3,5,33}. Risk of recurrence is high with discontinuation of medication³⁴. Use of gonadotrophin agonist promptly improves symptoms but does not change lesion size³⁵. Medical treatment is mainly for symptomatic control and shrinkage of the lesion size before operation. Combination of surgical re-excision and postoperative adjuvant medical therapy is suggested in patients with recurrent AWE³⁶.

Follow-up

Patients should be followed up to monitor recurrence. Recurrence rate after surgical treatment is 4% to 11% and usually occurs in a year after surgery³⁷.

Prevention

Preventive measures to reduce occurrence of AWE include meticulous haemostasis during uterine surgery, irrigating intra-abdominal cavities vigorously with high jet solution before abdominal closure, prompt removal of surgical sponges from operative field, gentle handling of uterine tissue, and using separate needles for suturing uterine and abdominal wall³⁸⁻⁴⁰. However, no trials have been conducted to evaluate the effectiveness of these measures in prevention of AWE.

Conclusion

AWE is rare. Careful history taking and physical examination are crucial to make the diagnosis. Imaging modalities enable assessment of the lesion extent and preoperative planning. Surgical excision with negative margin offers curative treatment. High-intensity focused ultrasound ablation and cryotherapy are non-invasive new alternatives.

Declaration

The authors have no conflict of interest to disclose.

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