# Extrapelvic Endometriosis

# 20

Atilla Bokor and Nura Fitnat Topbas Selcuki

## Contents



## 20.1 Introduction

Endometriosis is not only a disease of the pelvis, but it can also be seen in other parts of the human body. Symptoms can vary depending on the affected tissue or organ. Cyclical symptoms, which occur within the first 72 h of menstruation, are called catamenial. Catamenial symptoms are present in most of the patients, at least in early stages, and may be the only sign that leads to the final diagnosis. The prevalence is not exactly known due to the irregular clinical presentation, difficulty in diagnosis, and consultation of patients to non-gynecological specialists who are not familiar with this condition. Gold standard in diagnosis is the histopathological confirmation of endometrial glands and stroma in the excised material. A trained

A. Bokor (✉)
Department of Obstetrics and Gynecology, Semmelweis University, Budapest, Hungary

N. F. Topbas Selcuki
Department of Obstetrics and Gynecology, University of Health Sciences Turkey, Istanbul Sisli Hamidiye Etfal Training and Research Hospital, Istanbul, Turkey



E. Oral (ed.), *Endometriosis and Adenomyosis*,
https://doi.org/10.1007/978-3-030-97236-3_20

pathologist in detecting endometriosis is crucial since the excised material is not from the pelvic area. Imaging techniques and endoscopy can also be used for diagnosis depending on the affected organ system. In a systematic review conducted by Andres et al. (2020) involving 179 studies, malignancy was the most common differential diagnosis (37%) among reported visceral endometriosis cases [1]. Therefore, a clinical knowledge on the possible extrapelvic locations of endometriosis is important for an accurate diagnosis and proper treatment. Treatment depends on the localization. If complete excision is possible, this is the treatment of choice; when this is not possible, long-term medical treatment is necessary [2].

Extrapelvic endometriosis is most commonly found in the abdominal wall after surgical procedures, such as cesarean section, which is followed by inguinal and umbilical endometriosis and thoracic endometriosis syndrome. Endometriosis involving the bowel is considered intrapelvic and is discussed as a part of the deep infiltrative endometriosis. Therefore, it will not be included in this chapter. However, endometriosis involving other abdominal organs will be discussed under the visceral endometriosis section. Pelvic nerve endometriosis is discussed in a separate chapter as well. Publications addressing uncommon cases of endometriosis involving the vascular, lymphatic, and central nervous system, as well as skeletal muscle and peripheral nerves, are mostly based on case reports. These entities of limited clinical relevance will be discussed at the end.

## 20.2 Abdominal Wall Endometriosis: Scar, Perineal, Umbilical, and Inguinal Endometriosis

The most frequent location of the extrapelvic endometriosis is the abdominal wall [3]. Abdominal wall endometriosis (AWE) is defined by the presence of endometrial-like tissue superficial to the peritoneum, including skin, rectus abdominis muscle, and rectus sheath [4]. AWE is frequently associated with gynecologic procedures such as caesarean section, episiotomy, laparoscopic or abdominal hysterectomy [1, 5–7]. However, this definition also includes lesions that are not a result of a previous surgical procedure. In a review of 445 cases, the pooled mean time interval between index surgery and clinical presentation of AWE was reported to be 3.6 years [7]. Many cases of this entity are often misdiagnosed as hernia, hematoma, or lipoma. Therefore, patients are usually referred to general surgery clinics. Presence of intrapelvic endometriosis has been observed in 12% of AWE patients [8].

Scar endometriosis is the most common form of AWE, and the endometriotic loci are located near or at the site of the surgical incision. AWE is seen in 0.03–1.5% of women following caesarean delivery [5, 6]. Perineal endometriosis on the episiotomy scar is rarer. It is encountered in approximately 0.01–0.06% of women after vaginal birth with episiotomy [6, 9]. The etiology of scar endometriosis is thought to be iatrogenic through the transfer of endometrial cells into the surgical wound [10]. This mechanism is also called the metastatic theory. On the other hand,

primary endometriotic lesions of the abdominal wall are thought to occur through the metaplastic transformation of the coelomic epithelium. Primary lesions are usually located at or around umbilicus. Therefore, these lesions are called umbilical endometriosis. Umbilical endometriosis is estimated to occur in 0.5–1.0% of all cases of endometriosis and in 0.4–4% of all extragenital endometriosis cases [5, 11]. Another form of AWE, inguinal endometriosis, is defined by the presence of endometriotic loci in the extraperitoneal portion of the round ligament, in the inguinal lymph nodes, in the subcutaneous adipose tissue, and in the wall of sacs of inguinal or femoral hernias, which occur either as primary lesions or following gynecological and/or inguinal surgeries [12–16]. The actual incidence is not known. Most of the cases are observed on the right inguinal region [16].

Symptoms include local catamenial pain, diffuse abdominal pain, palpable mass with catamenial tenderness and swelling, and rarely umbilical bleeding. AWE can be identified with transabdominal ultrasonography (TAS), computed tomography (CT), and magnetic resonance imaging (MRI) in patients who are both symptomatic or asymptomatic [5, 6, 11, 17]. The appearance of AWE at TAS, CT, and MRI depends on the phase of the patient's menstrual cycle, the chronicity of the lesion, the number of stromal and glandular elements, the amount of bleeding and associated inflammation [6, 17, 18]. TAS is usually the first-line imaging modality in evaluating focal abdominal or inguinal wall thickening identified at clinical examination. With TAS, the extent and the nature of the focal lesions can be determined and abdominal wall hernias can be excluded [18]. CT and MRI are used to exclude differential diagnoses in the anterior abdominal and pelvic wall such as hernia, abscess, hematoma, and other soft-tissue tumors [6, 17, 18]. CT can be performed with or without intravenous contrast material, although the use of contrast material improves its sensitivity and specificity. The highest reported combined sensitivity of CT imaging for the diagnosis of AWE is 0.69 (95% CI: 0.48–0.86) and specificity is 0.97 (95% CI: 0.91–1.00) [17]. MRI provides better contrast resolution than CT and TAS and is superior to CT for depicting the delineation between muscle and abdominal subcutaneous tissue and infiltration of abdominal wall structures. Furthermore, MRI is preferred in younger patients because of its improved tissue characterization and lack of ionizing radiation. Recently, for the diagnosis of umbilical endometriosis, a sensitivity of 87.1% for physical examination, 76.5% for TAS, 75.6% for CT, and 81.8% for MRI was reported [5].

Ultrasound-guided fine-needle aspiration biopsy can be performed to exclude malignancy and establish a definitive preoperative diagnosis of AWE [18]. Treatment of choice is the surgical excision of endometriotic loci. The surgical therapy of AWE is often successful following a complete excision. Use of neoadjuvant and adjuvant hormonal treatment has been reported in only a few case reports. Due to lack of data, hormonal treatment is not routinely recommended. The decision should be made according to the symptoms of the patients, extend of the disease, and the presence of pelvic endometriosis.

## 20.3 Visceral Endometriosis

Endometriosis is also known to affect abdominal organs. Cases of liver, kidney, pancreas, and biliary tract endometriosis have been reported in the literature [1]. Symptoms depend on the affected organ. Liver endometriosis commonly presents with abdominal pain, abdominal mass, and acute liver failure. Flank pain, hematuria, and pyelonephritis are usually associated with kidney endometriosis. Epigastric pain and acute pancreatitis can occur in patients with pancreas endometriosis. In 62% of the patients, pelvic endometriosis was reported [1]. CT is the most commonly used imaging modality. Biopsy, if possible, can lead to definitive diagnosis. Surgical excision of the endometriotic loci is the treatment of choice. However, more radical approaches such as partial nephrectomy, partial hepatectomy, and complete nephrectomy can be performed in the presence of severe disease. Hormonal treatment can be administered following surgery.

## 20.4 Thoracic Endometriosis Syndrome

Thoracic endometriosis syndrome (TES) encompasses a variety of symptoms and radiological findings associated with the growth of endometrial foci within the respiratory system, most commonly the lung parenchyma, pleural surfaces, and the diaphragm [19–21]. These symptoms include pneumothorax, hemothorax, hemoptysis, chest pain, pulmonary nodules, endometriosis-related diaphragmatic hernia, and endometriosis-related pleural effusion [22, 23]. Symptoms usually have a catamenial pattern [1, 20, 21]. However, non-catamenial presentation has also been reported in the literature [24]. Approximately 90% of patients with TES experience catamenial thoracic pain, which is followed by catamenial pneumothorax (80%) [25, 26]. Catamenial hemothorax is observed in 14% of the reported cases and catamenial hemoptysis in 5% [22]. In majority of the cases, a right-sided hemithorax involvement has been reported [27, 28].

Although the exact pathophysiological mechanism of TES is not yet known, a multifactorial etiology is suspected. The theories already discussed as a possible explanation for the occurrence of endometriosis such as retrograde menstruation, coelomic metaplasia, lymphatic and hematogenous dissemination, and prostaglandin F2alpha involvement are also considered in the etiology of TES [26]. The right-sided predominance can be explained by the circulation of the peritoneal fluid, which flows from the pelvis through the right paracolic gutter to the right hemidiaphragm, while deviating away from the left hemidiaphragm due to obstruction of flow by the falciform and phrenicocolic ligaments [26, 29].

Symptoms arise according to the localization of the endometriotic lesions. Pleural involvement usually presents with catamenial pneumothorax, chest and/or shoulder pain and less commonly catamenial hemothorax. Pneumothorax leads to pleuritic chest pain, cough, and shortness of breath [26]. Diaphragmatic involvement with fenestrations, which are usually present in the tendinous (central) part of

the diaphragm, can cause secondary pneumothorax, serous or bloody pleural effusions, or partial thoracic herniation of abdominal organs [30, 31].

On the other hand, isolated diaphragmatic endometriosis is mostly asymptomatic, which is usually an incidental laparoscopic finding during pelvic endometriosis surgery [30]. Therefore, there is an ongoing debate on whether isolated diaphragmatic endometriosis should be considered as a part of TES or not. Endometriotic lesions of the diaphragm can cause phrenic nerve irritation. This leads to catamenial neck, shoulder, right upper quadrant, or epigastric pain [26, 32]. Parenchymal endometriotic lesions can cause mild to moderate hemoptysis and nodules can be identified with imaging.

Diagnosis of TES is challenging, as these women's symptoms may not immediately be attributed to endometriosis. Due to the respiratory related complaints, these patients usually visit thoracic clinics, which usually leads to a delay in the diagnosis. Chest X-ray (CXR), CT, and MRI are the imaging modalities of choice in diagnosis. CXR and CT are the most sensitive techniques in identifying hemothorax and pneumothorax [21, 26]. In a recent systematic review, only one study with 33 patients with diaphragmatic endometriosis evaluated the accuracy of MRI for diagnosis and reported a sensitivity of 83% with fat-suppressed T1-weighted sequences [1].

It is advisable to discuss the diagnosis and management of TES in a multidisciplinary team in a center with sufficient expertise [33]. In the case of catamenial pneumothorax, video-assisted thoracoscopic surgery (VATS) is the approach of choice for diagnosis and surgical treatment [19, 26]. Superficial lesions can be coagulated or ablated with different types of low-energy sources. The presence of larger lesions may afterwards be associated with diaphragmatic fenestrations and therefore a total excision when possible is advised [31, 34]. Small fenestrations in the diaphragm can be closed with interrupted stitches. In cases with large defects after resection, thoracoscopic suturing by a thoracic surgeon is preferable [30, 31]. Special care should be taken during the surgical interventions for the treatment of diaphragmatic endometriosis in order to preserve the phrenic nerve and vessels.

## 20.5 Other Sites

In a recent systematic review by Andres et al., a total of 19 case studies of nonabdominal and nonthoracic sites of endometriosis were reported [1]. These rare sites included six cases involving the central nervous system (one on brain, one on lumbar vertebra, and four on the conus medullaris). Twelve patients with endometriosis on extrapelvic muscles and peripheral nerves, and one case of nasal endometriosis. The age of this population ranged from 21 to 58 years.

In all cases involving the central nervous system, the extrapelvic muscles and the peripheral nerves, patients presented with paresthesia and catamenial pain radiating to the associated anatomical structures and dermatomes. Surgical excision was the definitive treatment in 91% of muscular and peripheral nerve endometriosis cases,

with complete and partial improvement of symptoms in 90.9% and 9%, respectively. Adjuvant hormonal therapy after muscular and peripheral nerve endometriosis resection was reported in 33.3% of these cases, with GnRH-analogues and OC.

## References


1. Andres MP, Arcoverde FVL, Souza CCC, Fernandes LFC, Abrão MS, Kho RM. Extrapelvic endometriosis: a systematic review. J Minim Invasive Gynecol. 2020;27(2):373–89.
2. Dunselman GA, Vermeulen N, Becker C, Calhaz-Jorge C, et al. ESHRE guideline: management of women with endometriosis. Hum Reprod. 2014;29(3):400–12.
3. Ideyi SC, Schein M, Niazi M, Gerst PH. Spontaneous endometriosis of the abdominal wall. Dig Surg. 2003;20(3):246–8.
4. Goksever Celik H, Karacan T, Kaya C, Uhri M, Savkli AO, Yalcin Bahat P, et al. Abdominal wall endometriosis: a monocentric continuous series and review of the literature. J Endometriosis Pelvic Pain Disord. 2019;11(2):95–101.
5. Hirata T, Koga K, Kitade M, Fukuda S, Neriishi K, Taniguchi F, et al. A national survey of umbilical endometriosis in Japan. J Minim Invasive Gynecol. 2020;27(1):80–7.
6. Chamié LP, Ribeiro DMFR, Tiferes DA, Macedo Neto AC, de, Serafini PC. Atypical sites of deeply infiltrative endometriosis: clinical characteristics and imaging findings. Radiographics. 2018;38(1):309–28.
7. Horton JD, Dezee KJ, Ahnfeldt EP, Wagner M. Abdominal wall endometriosis: a surgeon's perspective and review of 445 cases. Am J Surg. 2008;196(2):207–12.
8. Davis AC, Goldberg JM. Extrapelvic endometriosis. Semin Reprod Med. 2017;35(1):98–101.
9. Jubanyik KJ, Comite F. Extrapelvic endometriosis. Obstet Gynecol Clin N Am. 1997;24(2):411–40.
10. Woodward PJ, Sohaey R, Mezzetti TP. Endometriosis: radiologic-pathologic correlation. Radiographics. 2001;21(1):193–216; questionnaire 288-294.
11. Bergqvist A. Extragenital endometriosis. A review. Eur J Surg. 1992;158(1):7–12.
12. Pérez-Seoane C, Vargas J, de Agustín P. Endometriosis in an inguinal crural hernia. Diagnosis by fine needle aspiration biopsy. Acta Cytol. 1991;35(3):350–2.
13. Brzezinski A, Durst AL. Endometriosis presenting as an inguinal hernia. Am J Obstet Gynecol. 1983;146(8):982–3.
14. Quagliarello J, Coppa G, Bigelow B. Isolated endometriosis in an inguinal hernia. Am J Obstet Gynecol. 1985;152(6 Pt 1):688–9.
15. Majeski J. Scar endometriosis manifested as a recurrent inguinal hernia. South Med J. 2001;94(2):247–9.
16. Wong WSF, Lim CED, Luo X. Inguinal endometriosis: an uncommon differential diagnosis as an inguinal tumour. ISRN Obstet Gynecol. 2011;2011:272159.
17. Yarmish G, Sala E, Goldman D, Lakhman Y, Soslow R, Hricak H, et al. Abdominal wall endometriosis: differentiation from other masses using CT features. Abdom Radiol (NY). 2017;42(5):1517–23.
18. Gidwaney R, Badler RL, Yam BL, Hines JJ, Alexeeva V, Donovan V, et al. Endometriosis of abdominal and pelvic wall scars: multimodality imaging findings, pathologic correlation, and radiologic mimics. Radiographics. 2012;32(7):2031–43.
19. Alifano M, Trisolini R, Cancellieri A, Regnard JF. Thoracic endometriosis: current knowledge. Ann Thorac Surg. 2006;81(2):761–9.
20. Johnson MM. Catamenial pneumothorax and other thoracic manifestations of endometriosis. Clin Chest Med. 2004;25(2):311–9.
21. Rousset P, Rousset-Jablonski C, Alifano M, Mansuet-Lupo A, Buy J-N, Revel M-P. Thoracic endometriosis syndrome: CT and MRI features. Clin Radiol. 2014;69(3):323–30.

22. Bobbio A, Canny E, Mansuet Lupo A, Lococo F, Legras A, Magdeleinat P, et al. Thoracic endometriosis syndrome other than pneumothorax: clinical and pathological findings. Ann Thorac Surg. 2017;104(6):1865–71.
23. Kaya C, Iliman DE, Eyuboglu GM, Bahceci E. Catamenial pneumothorax: multidisciplinary minimally invasive management of a recurrent case. Kardiochir Torakochirurgia Pol. 2020;17(2):107–9.
24. Selcuki NFT, Yilmaz S, Kaya C, Usta T, Kale A, Oral E. Thoracic endometriosis; a review comparing 480 patients based on catamenial and non-catamenial symptoms. J Minim Invasive Gynecol. 2021;S1553-4650(21):00384–8.
25. Joseph J, Sahn SA. Thoracic endometriosis syndrome: new observations from an analysis of 110 cases. Am J Med. 1996;100(2):164–70.
26. Nezhat C, Lindheim SR, Backhus L, Vu M, Vang N, Nezhat A, et al. Thoracic endometriosis syndrome: a review of diagnosis and management. JSLS. 2019;23(3):e2019. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6684338/
27. Gil Y, Tulandi T. Diagnosis and treatment of catamenial pneumothorax: a systematic review. J Minim Invasive Gynecol. 2020;27(1):48–53.
28. Korom S, Canyurt H, Missbach A, Schneiter D, Kurrer MO, Haller U, et al. Catamenial pneumothorax revisited: clinical approach and systematic review of the literature. J Thorac Cardiovasc Surg. 2004;128(4):502–8.
29. Kirschner PA. Porous diaphragm syndromes. Chest Surg Clin N Am. 1998;8(2):449–72.
30. Ceccaroni M, Roviglione G, Farulla A, Bertoglio P, Clarizia R, Viti A, et al. Minimally invasive treatment of diaphragmatic endometriosis: a 15-year single referral center's experience on 215 patients. Surg Endosc. 2021;35(12):6807–17.
31. Working group of ESGE, ESHRE, and WES, Keckstein J, Becker CM, Canis M, Feki A, Grimbizis GF, et al. Recommendations for the surgical treatment of endometriosis. Part 2: deep endometriosis. Hum Reprod Open. 2020;2020(1):hoaa002.
32. Fukuda S, Hirata T, Neriishi K, Nakazawa A, Takamura M, Izumi G, et al. Thoracic endometriosis syndrome: comparison between catamenial pneumothorax or endometriosis-related pneumothorax and catamenial hemoptysis. Eur J Obstet Gynecol Reprod Biol. 2018;225:118–23.
33. Ciriaco P, Muriana P, Lembo R, Carretta A, Negri G. Treatment of thoracic endometriosis syndrome: a meta-analysis and review. Ann Thorac Surg. 2020;113(1):324–36.
34. Roman H, Darwish B, Provost D, Baste J-M. Laparoscopic management of diaphragmatic endometriosis by three different approaches. Fertil Steril. 2016;106(2):e1.