Atypical Sites of Deeply Infiltrative Endometriosis: Clinical Characteristics and Imaging Findings

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Abbreviations: DIE = deeply infiltrative endometriosis, FSE = fast spin echo, TES = thoracic endometriosis syndrome

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Endometriosis is defined as the presence of endometrial tissue that is located outside the uterine cavity and associated with fibrosis and inflammatory reaction. It is a polymorphic and multifocal disease with no known cure or preventive mechanisms. Patients may be asymptomatic or may experience chronic pelvic pain, dysmenorrhea, dyspareunia, or infertility. The pelvic cavity is the most common location for endometriotic implants, which usually affect the retrocervical space, ovaries, vagina, rectosigmoid colon, bladder dome, and round ligaments. Atypical endometriosis is rare and difficult to diagnose. The most common atypical locations are the gastrointestinal tract, urinary tract, lung, umbilicus, inguinal area, breast, and pelvic nerves, as well as abdominal surgical scars. Gastrointestinal lesions are the most common extragenital manifestation, and the diaphragm is the most frequent extrapelvic site. The catamenial nature of the symptoms (occurring between 24 hours before and 72 hours after the onset of menstruation) may help suggest the diagnosis, but imaging by specialists is fundamental to evaluation. Depending on the area affected, radiography, ultrasonography, thin-section computed tomography, or magnetic resonance imaging can be used to assess suspected lesions. Because isolated extragenital endometriosis is rare, concomitant evaluation of the pelvic cavity is mandatory. Surgical excision is the only therapeutic option for definitive treatment, and comprehensive disease mapping is necessary to avoid residual disease. The authors review atypical locations for endometriosis and emphasize the most appropriate imaging protocols for investigation of various clinical manifestations.

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Introduction

Endometriosis is a benign gynecologic disease affecting women of reproductive age. It is defined as the presence of endometrial tissue, glands, and stroma located outside the uterine cavity and associated with fibrosis and inflammatory reaction (1). It is a polymorphic disease that can manifest as superficial implants on the peritoneal surface; ovarian cysts called endometriomas; or deep lesions that infiltrate the peritoneal surface more than 5 mm, a condition known as deeply infiltrative endometriosis (DIE). DIE is a chronic, progressive, multifocal disease with no known cure or preventive mechanisms (2).

Clinically, DIE is characterized by chronic pelvic pain, severe dysmenorrhea, dyspareunia, dyschezia, dysuria, and infertility, although the disease may be asymptomatic or discovered incidentally (3). It is estimated to affect 176 million women worldwide, with an average of one in 10 women affected during the reproductive years.

Endometriosis can affect almost any organ or structure, although most endometriotic implants are located in the pelvic cavity. Pelvic endometriosis has a somewhat predictable and repetitive distribution...
Patients with cyclic bowel symptoms, chronic right lower abdomen and right lower quadrant pain, and severe endometriosis are at higher risk for appendiceal endometriosis.

Although it is an uncommon condition, TES is the most frequent extrapelvic manifestation of endometriosis and encompasses four clinical manifestations: catamenial pneumothorax, catamenial hemoptysis, catamenial hemothorax, and lung nodules. Pleural endometriosis manifests as catamenial pneumothorax, pneumomediastinum, hemothorax, and chest pain, and the pulmonary form manifests as catamenial hemothorax and pulmonary nodules. Catamenial pneumothorax is the most common clinical manifestation (73%), followed by catamenial hemothorax (14%), hemoptysis (7%), and lung nodules (6%).

Abdominal Wall

The abdominal wall is one of the most frequent extrapelvic locations of endometriosis. Endometriotic implants are usually embedded in the subcutaneous fatty layer and the muscles of the abdominal wall near or at the site of surgical scars. Although it is an uncommon condition, TES is the most frequent extrapelvic manifestation of endometriosis and encompasses four clinical manifestations: catamenial pneumothorax, catamenial hemoptysis, catamenial hemothorax, and lung nodules. Pleural endometriosis manifests as catamenial pneumothorax, pneumomediastinum, hemothorax, and chest pain, and the pulmonary form manifests as catamenial hemothorax and pulmonary nodules. Catamenial pneumothorax is the most common clinical manifestation (73%), followed by catamenial hemothorax (14%), hemoptysis (7%), and lung nodules (6%).

Surgical excision of ectopic endometriotic tissue is the main option for definitive treatment, and the success of the surgical procedure depends on complete removal of all lesions. Therefore, imaging mapping is at the core of clinical practice and allows a comprehensive evaluation for patient counseling and preoperative planning. The pelvic cavity, abdominal wall, small bowel, appendix, and inguinal area can be evaluated with transvaginal ultrasonography (US) after bowel preparation, high-resolution US with a linear transducer, and magnetic resonance (MR) imaging (4,7,10–13). Thoracic endometriosis can be assessed with conventional radiography, thin-section computed tomography (CT), and thoracic MR imaging (10,14).

This article describes atypical sites of endometriosis, with a review of the literature and presentation of clinical cases with imaging findings and surgical correlation. We discuss cases of endometriosis of the abdominal wall, umbilicus, inguinal area and round ligaments, small bowel, appendix, thoracic cavity, and pelvic nerves.
tissue. Alternative theories include the migration of endometriotic tissue through lymphatic and vascular vessels to distant sites (15,16). The metaplastic and migration theories may help explain distant endometriotic nodules in sites without direct contact with endometrial tissue. An association between incisional and pelvic endometriosis is uncommon.

The typical clinical manifestation of abdominal wall endometriosis is pain that usually starts after the surgical procedure and is cyclical, worsening during the first phase of the menstrual cycle. The time interval between surgery and the manifestation of scar endometriosis varies from months to years. Patients may report a mass sensation, dysmenorrhea, or bleeding, and in general they can point to the exact physical location of the endometriotic nodule (16). Despite the typical clinical manifestations, definitive diagnosis may be possible only after surgical removal of the nodule and pathologic analysis.

Various imaging modalities can be used for diagnosis of abdominal wall endometriosis, including color Doppler US, CT, and MR imaging. US performed with a linear transducer will demonstrate a hypoechoic nodule or mass with irregular contours, sometimes containing small cystic areas with or without thick content and small scattered hyperechoic echoes (Fig 2). At color Doppler US, a hypovascular pattern with small peripheral vessels is seen (17) (Fig 3). US is the most cost-effective imaging modality and should be performed first. At CT, an endometriotic nodule appears as a circumscribed solid mass that enhances with contrast agent administration. MR imaging is more accurate for diagnosis, especially when small hemorrhagic areas are present within the nodule, which increases the specificity for diagnosis. The excellent spatial and contrast resolution of the method allow better tissue definition, which is valuable in defining the extent of disease, the integrity of the surrounding muscle tissue, and other affected structures. Endometriotic

Figure 1. Schematic axial (a) and midsagittal (b) drawings of the female pelvic cavity show the typical locations of endometriotic lesions (black areas). The dotted rectangle in a represents the most common locations: the retrocervical space, pouch of Douglas, and rectosigmoid colon. The dotted line in b separates the pelvis into the anterior and posterior compartments. (Adapted and reprinted, with permission, from reference 4.)

Figure 2. Cesarean scar endometriosis in a 36-year-old woman with localized anterior pelvic pain. Transabdominal US image of the anterior hypogastric region obtained with a high-resolution linear probe shows a mixed nodule (arrows) with irregular margins and cystic areas that infiltrates the muscle layer (ML). SCL = subcutaneous layer.
lesions typically have low signal intensity on T2-weighted images, with high-signal-intensity small cystic areas and spiculated margins (Figs 4, 5a, 5b). On T1-weighted images, they demonstrate intermediate signal intensity with hemorrhagic internal areas of high signal intensity and contrast enhancement (Fig 5c, 5d). Nodules can be located exclusively inside the muscle layer or the subcutaneous fatty layer, or both. The inflammatory process can extend beyond the affected area like a ring of variable width and enhances after contrast material injection at CT and MR imaging. Episiotomy endometriosis manifests with a similar pattern, and the nodules usually contain multiple cysts (Figs 6, 7).

The main treatment of scar endometriosis is surgical resection of the affected area. Medications are not effective and provide transient relief; most patients have a recurrence of symptoms after stopping medication. Wide excision with at least a 1-cm margin is considered the treatment of choice. Fascial defects may need closure with synthetic mesh if the underlying sheet is involved. Imaging mapping is crucial for better surgical results and to avoid residual disease. The recurrence rate is low, and the risk for malignant transformation is less than 1%. The most common type of malignant transformation is clear cell carcinoma (15).

**Umbilicus**

Umbilical endometriosis is rare, estimated to occur in 0.5%–1.0% of all cases of endometriosis. It can be classified as primary, also known as a Villar nodule (first described by Villar in 1886), when it appears spontaneously, or secondary, when it appears after a surgical procedure (13,18,19). About 70% of abdominal endometriosis is secondary. The umbilicus is the most common location of primary disease, followed by the inguinal region. The pathogenesis of primary umbilical endometriosis remains unclear. Different theories have been proposed, such as the migration of endometrial cells through the abdominal cavity, the lymphatic system, or embryonic remnants in the umbilical fold (eg, the urachus and umbilical vessels); genetic predisposition; and immunologic defects. Secondary endometriosis is caused by direct implantation or dissemination of endometrial tissue, such as through laparoscopy. Furthermore, it has been hypothesized that the umbilicus may act as a physiologic scar and seeding site, contributing to the etiology of secondary umbilical endometriosis (13).

The clinical manifestation typically involves an umbilical mass or firm nodule of variable size, from a few millimeters up to 6 cm, and symptoms such as swelling (90.9%), cyclic pain (81.5%), and bleeding (49.2%) concurrent with menstruation. The nodule can be brown, blue, or a faint spot. A history of concomitant pelvic disease is common (26% of cases), with dysmenorrhea, dyspareunia, or pain during defecation. There may be constant rather than periodic pain and an underlying hernia (19,20). Umbilical endometriosis is considered a marker for severe pelvic endometriosis, with increased risk for intestinal disease. The differential diagnosis for umbilical endometriosis includes benign diseases such as hemangioma, umbilical hernia, sebaceous cyst, granuloma, lipoma, abscess, and urachal anomaly and malignant diseases such as melanoma, sarcoma, adenocarcinoma, and lymphoma.
Figure 5. Cesarean scar endometriosis in a 43-year-old woman with pelvic pain and dyspareunia. (a, b) Sagittal (a) and axial (b) T2-weighted FSE pelvic MR images show a nodule (straight arrow) with heterogeneous signal intensity, irregular margins, and internal areas of high signal intensity within the rectus abdominis muscle. There is a second endometriotic nodule (curved arrow in a) in the retrocervical space that infiltrates the pouch of Douglas (rectouterine pouch) and posterior vaginal wall. (c–e) Axial nonenhanced (c), axial gadolinium-enhanced (d), and sagittal gadolinium-enhanced (e) fat-saturated T1-weighted gradient-echo MR images show the precontrast and postcontrast characteristics of the nodule (arrow), with notable enhancement seen after gadolinium injection.
Making a diagnosis of umbilical endometriosis can be difficult, and there is often a delay in diagnosis. Patients often have a lengthy disease course before clinical presentation. US, CT, and MR imaging are frequently used to assess the nature of the lesion and its vascularity. US is used to define whether the lesion is cystic or solid, but it has low specificity for diagnosis. The typical US finding is a nodular formation that occupies the umbilical scar, with ground-glass echogenicity, irregular margins, and no papillary structures with a detectable blood flow (Fig 8). MR imaging has been shown to be the best choice for diagnosis and is very useful to define the size, extension, and hemorrhagic content of cysts. It is also a valuable method to exclude concomitant abdominal disease. The typical MR imaging finding is a nodule with low signal intensity on T2-weighted images, intermediate signal intensity on T1-weighted images, internal cystic areas with high signal inten-
Inguinal Area and Round Ligaments

Inguinal endometriosis is a rare condition, accounting for 0.3%–0.6% of patients affected by endometriosis, with fewer than 60 cases reported in the literature. It is characterized by the presence of endometrial stroma and glands in the extraperitoneal portion of the round ligament, the inguinal lymph nodes, the subcutaneous adipose tissue, or even the sac wall of inguinal or femoral hernias (7,21). The typical presentation is a patient in her reproductive years (peak incidence, 30–40 years of age) who reports inguinal swelling (Fig 10). Catamenial symptoms synchronized with the menstrual period, such as variations in the size and tenderness of the mass, occur in 50% of cases and should raise clinical suspicion for inguinal endometriosis (21).

The prevalence of inguinal endometriosis on the right side is 90%–94% as compared to the left side. The asymmetric lymphatic drainage, which favors the right inguinocrural region, might explain this phenomenon. In addition, the endometrial cells remain for a longer time on the right side owing to gravity and to protection of the left ligament by the sigmoid colon. Nevertheless, none of these hypotheses offer adequate explanation for the pathogenesis of inguinal endometriosis (22).

Endometriosis of the round ligament is associated with groin hernias (usually inguinal) in about 40% of cases, which increases the diagnostic difficulty. The presumptive diagnosis is most often confused with incarcerated hernia, lymphadenopathy, neuroma, abscess, lymphoma, lipoma, hematoma, sarcoma, or subcutaneous cysts. The final diagnosis is usually made on the basis of histopathologic results after surgical excision (7). Malignant degeneration is rare.

US, CT, or MR imaging is used to elucidate the nature of the mass. Color Doppler US will demonstrate a hypoechoic mass with absent flow, with or without cystic areas. The cystic portion is usually filled with thick fluid and has a ground-glass appearance (Fig 11). MR imaging is more accurate than US because it can demonstrate the old hemorrhagic content of the cyst (7) (Fig 12). Endometriotic cysts are unique retention cysts that undergo cyclic bleeding for many years and have morphologic characteristics different from those of other types of lesions. The thick aged blood and its high viscosity are responsible for the high signal intensity seen on T1-weighted images and the low signal intensity seen on T2-weighted images, together called the “shading sign.” The mechanism of shading is attributed to the high concentration of methemoglobin and protein within the cyst (23,24).
The treatment of choice is surgical en bloc resection of the lesion. To be locally curative and to prevent recurrence, surgical resection must include removal of the extraperitoneal portion of the round ligament. Given the high association with pelvic endometriosis, concurrent laparoscopy is indicated to treat pelvic disease (21).

**Small Bowel**

Bowel endometriosis is one of the most aggressive forms of DIE. The bowel is the most common location of extragenital endometriosis, and bowel endometriosis occurs in up to 37% of women with DIE. Although the rectosigmoid colon is the most common location (52%–72%), endometriotic implants can also be found in the small bowel, especially in the terminal ileum (4.1%–16.9%) (10).

Clinically, patients may be asymptomatic or may present with a variety of nonspecific symptoms that are frequently associated with other diseases, such as constipation or diarrhea, a palpable mass, melena, rectal bleeding, tenesmus, abdominal distention, or meteorism due to the accumulation of gas in the bowel. Gastrointestinal endometriosis can manifest as acute abdomen
with perforation, intussusception, and obstruction. Other conditions that affect the terminal ileum, such as carcinoid tumors, small-bowel lymphoma, Behçet disease, Crohn disease, and tuberculous enteritis, should be excluded in the differential diagnosis (25). Only 0.15% of patients with small-bowel endometriosis will develop a small-bowel obstruction (26).

Enteric endometriosis characteristically manifests as a mural nodule that affects the serosa, muscularis propria, and submucosa. The mucosa is usually intact. Noninvasive diagnosis of bowel endometriosis can be achieved with use of CT or MR imaging, and obtaining good distention is a key factor, particularly in the small bowel. CT enteroclysis may allow good visualization of the terminal ileum, but this technique requires retrograde opacification and ionizing radiation and is of limited value for other pelvic sites (27). MR enterography, which uses anterograde opacification, is a radiation-free imaging modality that is widely used to investigate small-bowel and ileocecal diseases (10,28,29). Endometriosis is a multifocal disease with multiple sites affected. MR imaging allows concomitant evaluation of the bowel, urinary tract, and pelvic nerves, along with other commonly involved sites such as the retrocervical space, vagina, and ovaries (10,30).

The typical MR imaging finding is a nodule with low signal intensity on T2-weighted images and postcontrast enhancement (Figs 13, 14).
Transvaginal US performed after bowel preparation is an excellent imaging modality to investigate bowel endometriosis, especially that affecting the rectosigmoid colon (31,32). Bowel preparation is used to eliminate fecal residue and gas that may impair adequate visualization of the lesions and is accomplished by (a) administering a mild laxative orally at 8:00 AM and 2:00 PM the day before the transvaginal examination, (b) instructing the patient to follow a low-residue diet on the day before and the day of the examination, and (c) administering a rectal enema consisting of 120 mL of sodium diphosphate approximately 1 hour before the examination (33). The same protocol can be used to diagnose ileocecal lesions, but a complementary US evaluation through the abdomen with a linear transducer is necessary (4,33). The nodules demonstrate the same imaging manifestations as rectosigmoid lesions, with hypoechoic nodules attached to the bowel wall and deeply infiltrating the muscularis propria (Fig 15). The MR imaging protocol should also be devoted to investigating DIE. We suggest use of an eight-channel cardiac coil, patient fasting for 4 hours, administration of vaginal gel, and bowel preparation. Bowel cleansing can be performed with an oral laxative (5 mg of bisacodyl per dose) at 8:00 AM and 2:00 PM the day before imaging and adherence to a low-residue diet the day before and the day of the MR imaging examination. Our standard MR imaging protocol includes acquisition of axial, sagittal, and coronal T2-weighted FSE images and axial fat-suppressed T1-weighted gradient-echo images in and out of phase. Dynamic sagittal and delayed phase axial images are acquired with a liver acquisition with volume acceleration (LAVA; GE Healthcare, Waukesha, Wis) sequence 50 seconds after intravenous injection of a gadolinium-based contrast material (4). Treatment consists of surgery with resection of the affected segment.

Appendix

Appendiceal endometriosis is an uncommon condition that usually is asymptomatic and is an incidental finding in patients with severe endometriosis. It can be an isolated finding on the right side of the colon, or it can be associated with ileocecal lesions. Endometriosis of the appendix causing acute appendicitis is rare and constitutes less than 1% of all appendiceal pathologic conditions that can mimic acute appendicitis (34,35). It can also be associated with intussusception into the cecum (11) (Fig 16). Lesions preferentially affect the body and tip of the appendix (Figs 17, 18).

Diagnostic imaging of appendiceal endometriosis is difficult and requires dedicated protocols. Transvaginal US with bowel preparation can be used to readily identify a nodular thickening of the appendix, but investigation of the right iliac fossa with a high-resolution linear
transducer is mandatory. MR imaging can also be useful, especially when used with enterography, but the method may have a limited performance on the right side of the colon, especially for small lesions, because of peristaltic artifacts and bowel content (28).

Patients with cyclic bowel symptoms, chronic right lower quadrant pain, and severe endometriosis are at higher risk for appendiceal endometriosis. Therefore, during laparoscopy the appendix should always be carefully inspected and resected if any abnormalities are found. A carcinoid tumor may be unexpectedly diagnosed in an incidental lesion thought to be endometriosis. Carcinoid tumors are the most common neoplasm of the appendix, with a prevalence of 0.32%. Although endometriosis is the most likely diagnosis in patients with severe DIE and an appendiceal nodule, carcinoid tumors should be included in the differential diagnosis because they cannot be ruled out by imaging findings alone (36).

**Thoracic Cavity**

*Thoracic endometriosis* is defined as the presence of functional ectopic endometrial tissue inside the thoracic cavity. The finding of endometrial implants in the airways, pleura, pericardium, and lung parenchyma is called thoracic endometriosis syndrome (TES) (37). Although it is an uncommon condition, TES is the most frequent extrapelvic type of endometriosis and encompasses four clinical manifestations: catamenial pneumothorax, catamenial hemothorax, catamenial hemoptysis, and lung nodules (7). Pleural endometriosis manifests as catamenial pneumothorax, pneumomediastinum, hemothorax, and chest pain, and the pulmonary form manifests as catamenial hemoptysis and pulmonary nodules. Catamenial pneumothorax is
the most common clinical manifestation (73%), followed by catamenial hemothorax (14%), hemoptysis (7%), and lung nodules (6%). Pericardial endometriosis is rare, with a few cases reported in the literature that manifested with ascites and pleural and pericardial effusion (38). It is estimated that 50%–85% of women diagnosed with TES have concomitant pelvic disease (39,40).

The etiology of TES is unknown and not completely elucidated. Several theories have been proposed, including (a) coelomic metaplasia of the distant tissues into ectopic endometrial tissue; (b) migration resulting from the migration of endometrial tissue from the uterus to the pelvis through the peritoneal fluid from retrograde menstruation to the subdiaphragmatic area; (c) metastatic or lymphovascular microembolization, which suggests a metastatic spread of endometrial cells to the lungs through venous or lymphatic vessels; (d) physiologic hypothesis, where high levels of circulating prostaglandin F2 as a result of a collapsed endometrium cause vasoconstriction and bronchospasm, resulting in rupture of the alveolar tissue and pneumothorax; and (e) transgenital-transdiaphragmatic passage, where the passage of air through congenital or acquired (secondary to endometriosis) diaphragmatic defects may result in ectopic implants (37,41).

Catamenial pneumothorax is characterized by repeated episodes of pneumothorax that are synchronized with the menstrual cycle. A pneumothorax occurring from 24 hours before...
to 72 hours after the onset of menstruation is described as catamenial. It is observed in 20%-30% of women with spontaneous pneumothorax. In most cases, pneumothoraces are typically right sided and small to moderate in size. A possible explanation for the right-sided predominance is the preferential clockwise flow of the peritoneal fluid circulation: there is more extensive diaphragmatic lymphatic drainage on the right side, and the anatomic barrier formed by the round and hepatic falciform ligaments predisposes right subdiaphragmatic deposition of endometrial implants (42). A review of cases of catamenial pneumothorax reported lesions in the right chest in 91.7% of patients, in the left chest in 4.8% of patients, and in both sides in 3.5% of patients, findings that indicate a right-sided predominance (43).

Symptoms of TES are typically catamenial, and chest pain is the most common clinical manifestation (90%), followed by dyspnea (31%), hemoptysis (7%), and scapular pain and cough (rare). About 30%-60% of patients are infertile. The CA-125 concentration is significantly higher in women with TES compared to those without TES, although this finding is nonspecific (8).

Diagnosis of TES is difficult and remains a clinical challenge unless TES is strongly suspected by the typical clinical history. It is interesting that the mean age of patients with thoracic endometriosis is higher than the mean age of patients with pelvic endometriosis, with a mean time lapse of 10 years from the initial diagnosis of pelvic endometriosis to the diagnosis of thoracic endometriosis (44). Repeated pneumothorax or hemorrhage during menstruation followed by a symptomless intervening period is diagnostic for thoracic endometriosis. Histologic confirmation of ectopic endometrial tissue is not always feasible, nor is histologic examination always performed; histologic confirmation is obtained in only one-third of cases (37,39). At physical examination, diminished or absent breath sounds on the affected side are heard. Diagnostic imaging includes chest radiography, thin-section CT, and MR imaging. Chest radiography and thin-section CT may reveal pneumothorax, pleural effusions, nodules, opacities and nodular infiltrates, thin-walled cavities, segmental atelectasis, and bullous formation (39) (Figs 19–21). MR imaging is an excellent imaging modality with high
Pelvic imaging examinations, including transvaginal US after bowel preparation and pelvic MR imaging, should be performed to investigate pelvic disease because of the high prevalence of concomitant thoracic and severe abdominal endometriosis, which varies from 50% to 100% (9,44).

TES can be treated conservatively, with surgical excision, or with combined therapy. Hormonal suppression may be considered but...
Video-assisted thoracoscopic surgery (VATS) with removal of all ectopic endometrial tissue, closure of diaphragmatic defects, and pleurectomy is considered the reference standard for both definitive diagnosis and treatment (Movie 2) (8,38,44,45). A multidisciplinary approach of combined VATS and laparoscopy is recommended to treat pelvic, diaphragmatic, and thoracic endometriosis in a single procedure (44). The primary surgical goal is to minimize recurrence by precise localization of the lesion and radical removal. Ectopic endometrial implants in the lungs are removed by wedge resection or limited lung segmentectomy.

Pelvic Nerves

DIE is a chronic debilitating condition that can manifest with a variety of symptoms as a result of the aggressive and infiltrative nature of the disease. Neural entrapment is a possible occurrence and has been described in different pelvic nerves, such as the sciatic, obturator, femoral, and pudendal nerves and the inferior hypogastric and lumbosacral plexus (46–50). The entirely retroperitoneal path of these nerves raises a question about the pathogenesis of the endometriosis, since the retrograde menstruation hypothesis cannot explain subperitoneal implants. Lymphatic and vascular metastases with dissemination of endometrial cells to the retroperitoneum is a possible hypothesis. Another proposed explanation is the neural theory, in which the sympathetic subgroups neuropeptide Y–sympathetic nerve can play a role in the growth of retroperitoneal endometriosis because of the induced angiogenesis (46). Direct infiltration from...
a large retrocervical lesion to the parametrium is
the most common manifestation, with posterior
extension into the presacral space (affecting the
splanchnic and sacral nerves) or inferior exten-
sion affecting the inferior hypogastric plexus path.
Large paracervical lesions can also spread into the
lateral pelvic wall, infiltrating the obturator nerve
path as well as the ureters.

Although it is a rare cause of sciatica, en-
dometriosis should be considered if the sciatic
pain has a cyclical occurrence concomitant
with the menstrual cycle (51). Typical symp-
toms are catamenial L5–S1 sciatica, gluteal
pain, and locomotor deficits (of the foot). With
evolution, symptoms may progress to loss of
sensation and motility in the involved area.
Catamenial sciatica may be associated with
signs of other nerve trunk involvement (ie, puden-
dal or inferior hypogastric plexus). Over time, the
sciatic pain may become permanent (48).

Symptomatic obturator nerve endometriosis
is a rare occurrence. The obturator nerve and
its branches supply the muscle and skin of the
medial thigh. Aggressive infiltration of the retro-
peritoneal space by paracervical endometriotic
lesions can cause obturator infiltration. Patients
may present with thigh adduction weakness or
difficulty ambulating (49).

The pelvic floor is another possible route for
extension of endometriotic lesions, mainly from
a retrocervical lesion with parametrium and
paracolpium infiltration. The levator ani muscle
may be compromised, along with the rectovagi-
nal fascia.

Comprehensive neurologic and radiologic
evaluation is mandatory to rule out lumbar disk
disease, spondylotic nerve root compression,
primary neural tumor, and metastasis. Early
diagnosis of endometriosis infiltrating the nerve
is important to prevent irreversible damage to
the nerve and corresponding muscle. Results of
electrophysiologic studies can help in positive
diagnosis of peripheral nerve damage and can
help localize the disease to the nerve trunk or

Figure 23. Diaphragmatic endometriosis in a 44-year-old woman with a history of repetitive pneu-
mothoraces. (a) Axial fat-saturated T1-weighted gradient-echo MR image shows a high-signal-intensity
nodule (arrow) attached to the diaphragm. (b, c) Coronal (b) and sagittal (c) gadolinium-enhanced fat-
saturated T1-weighted MR images show enhancement of the diaphragmatic nodule (arrow). (d) Lapa-
rosscopic photograph shows multiple endometriotic spots and nodules (dashed oval) with hemorrhagic
content on the right hemidiaphragm.
nerve root involved, but they do not indicate the mechanism of the damage (48). MR imaging is the best imaging modality to investigate neural involvement. MR imaging findings can vary from solid spiculated nodules or complex cystic masses to cystic lesions with thick walls (Figs 25–27). Cystic components often exhibit high signal intensity on T1-weighted images and low signal intensity on T2-weighted images. Adjacent muscles and bones usually demonstrate a diffuse increase in signal intensity on T2-weighted images and diffuse contrast enhancement due to the inflammatory process and edema (51).

Many treatment options can be tried to offer patients relief, such as physiotherapy, analgesics, tricyclic antidepressants, antiepileptics, and surgery with neurolysis and decompression (52). Laparoscopic eradication of endometriosis with nerve-sparing technique and somatic nerve decompression is considered the reference standard (52,53).
Figures 26, 27. (26) Endometriosis affecting the pelvic nerves in a 24-year-old woman with right leg pain and dyspareunia. (a, b) Axial (a) and coronal (b) T2-weighted FSE MR images show a left retrocervical nodule (arrows) infiltrating the inferior hypogastric plexus with posterior extension close to the lumbosacral plexus. A right paracervical nodule (arrowheads) infiltrates the obturator nerve. (c) Sagittal T2-weighted FSE MR image shows an extensive low-signal-intensity retrocervical lesion (straight arrows) with spiculated margins infiltrating the left inferior hypogastric plexus and the pelvic floor close to the puborectalis muscle (*). There is associated left-sided hydronephrosis (curved arrow) due to ureteral infiltration and stenosis. (27) Endometriosis with infiltration of the pelvic floor in a 43-year-old woman with chronic right back pain. (a, b) Coronal (a) and axial (b) T2-weighted FSE MR images show a nodule (arrow) with low signal intensity and irregular margins infiltrating the horizontal portion of the right levator ani muscle (ileo-coccygeus muscle), the obturator internal muscles, and the presacral nerves. (c) Sagittal T2-weighted FSE MR image shows the retroperitoneal location of the nodule (arrow). (d) Sagittal gadolinium-enhanced fat-saturated T1-weighted three-dimensional MR image shows enhancement of the nodule (arrow).
Conclusion
Extrapelvic endometriosis is rare and can be difficult to diagnose because of the great variability in location and clinical manifestations. The catamenial nature of the symptoms can help suggest the diagnosis and guide the clinical investigation, especially in women of reproductive age.

Late diagnosis is common in patients with pelvic endometriosis and is even more common in those with extrapelvic disease. Treatment of endometriosis usually requires a multidisciplinary approach. Surgical excision of all lesions is the best approach for definitive treatment.

References


