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Leiomyomatosis peritonealis disseminata complicated by sarcomatous transformation and ovarian torsion: presentation of two cases and review of the literature

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Abstract

Leiomyomatosis peritonealis disseminata (LPD) is a rare disorder usually discovered incidentally in women of child-bearing age and is characterized by multiple subperitoneal smooth muscle nodules. Case reports of two patients with complications related to LPD and a review of the literature are presented. In one case, the patient carried the diagnosis of LPD for 11 years and experienced sarcomatous transformation; this is the first report of the magnetic resonance appearance of this entity. In the second case, LPD was diagnosed after an LPD implant on the ovary-induced ovarian torsion. We also present a patient in whom large, pedunculated uterine leiomyomas mimicked LPD. The clinical presentation, possible pathogenesis, imaging features, and therapeutic options of LPD are reviewed. Because this uncommon condition is being reported with increasing frequency, familiarity with its imaging features and pitfalls is important to suggest the diagnosis in the appropriate clinical setting.

Key words: Uterine neoplasms— Peritoneum—Leiomyosarcoma—MR—CT—US.

Leiomyomatosis peritonealis disseminata (LPD) is a rare disorder characterized by multiple subperitoneal smooth muscle nodules [1]. Since the first report by Willson and Peale in 1952 [2], there have been approximately 60 cases reported in the literature [2–16]. Most cases occur in women in the reproductive age group and are often associated with pregnancy or oral contraceptive use, although there have been a few reports of LPD in postmenopausal women and one report in a man [5, 10, 11, 15]. Although LPD may be asymptomatic and discovered incidentally during cesarean section, laparotomy, or laparoscopy, LPD may occasionally be symptomatic or undergo malignant transformation [11, 14, 16].

Because LPD may mimic peritoneal carcinomatosis on imaging studies and at surgery, it is important that radiologists be aware of this entity and its imaging features, which have not been emphasized in the literature. Even on histologic examination, LPD may be confused with a low-grade leiomyosarcoma [7].

We present two cases of LPD: one complicated by sarcomatous transformation and the other by ovarian torsion. This is the first report of the magnetic resonance (MR) appearance of sarcomatous transformation of LPD. The clinical presentation, possible pathogenesis, and imaging features of LPD are discussed. In addition, we present a case of extensive diffuse leiomyomatosis of the uterus, which mimics LPD.

Case reports

Case 1

A 48-year-old gravida 2 para 2 woman with a diagnosis of LPD presented to the emergency department with shortness of breath and fever. Vital signs were: blood pressure 128/69, pulse 114, respiratory rate 36, and temperature 100.7°F. Physical examination was significant for tachycardia and tachypnea. Routine laboratory analysis revealed a blood urea nitrogen level of 97 mg/dL and a creatinine level of 9.3 mg/dL. Past medical history was significant for a total abdominal hysterectomy performed 11 years before due to symptomatic leiomyomata. Two years after the hysterectomy, physical examination revealed bilateral ovarian masses. The patient underwent bilateral oophorectomy, at which time leiomyomatous ovarian implants were detected, and the diagnosis of LPD was established. Seven abdominal

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and pelvic surgeries were performed over the course of the next 8 years due to recurrent LPD. The pathologic specimens obtained during the multiple surgeries showed no evidence of sarcomatous transformation. The patient had never taken hormonal therapy.

Because of the presumptive diagnosis of urosepsis, renal sonography was conducted to assess for obstruction. The sonogram revealed bilateral hydronephrosis secondary to a 12-cm solid pelvic mass (Fig. 1A,B). Bilateral percutaneous nephrostograms demonstrated extrinsic compression of the ureters by the pelvic mass (Fig. 1C). T1- and T2-weighted images showed multiple, soft tissue masses in the pelvis (Fig. 1D–G).

Laparotomy was performed. However, only four of the pelvic masses could be removed due to extensive adhesions. Gross inspection of the masses revealed that they were white-tan, firm, and focally hemorrhagic. Pathologic analysis demonstrated marked cellular atypia, necrosis, and multiple mitoses. A final pathologic diagnosis of moderate-grade leiomyosarcoma was made. The patient's postoperative course was uneventful. Computed tomography (CT) performed 8 months after laparotomy revealed no change in the size or appearance of the remaining masses as compared with a baseline CT scan performed 6 weeks after laparotomy. Twelve months after laparotomy, the pelvic masses remained stable in size, but multiple lung nodules developed. Biopsy of the lung nodules was not performed. Serial chest CT scans performed over the course of 4 months demonstrated enlargement of one nodule.

Case 2

A 44-year-old gravida 3 para 2 abortus 1 woman was in good health until waking suddenly with sharp right abdominal pain. Twelve hours after the onset of the pain, the patient presented to the emergency department. Physical examination revealed right upper and lower quadrant pain associated with rebound. A mass measuring 15 cm was palpated at the site of maximal tenderness in the right midabdomen. The patient demonstrated normal blood pressure, heart rate of 119 beats/minute, and respiratory rate of 20 breaths/minute. Laboratory analysis was normal. Past surgical history was significant for a hysterectomy performed 12 years before due to symptomatic uterine leiomyomata. She had no history of oral contraceptive use. An enhanced CT scan performed in the emergency department demonstrated a heterogeneously enhancing mass arising from the right adnexa and extending into the right side of the abdomen (Fig. 2A,B). A second mass measuring 4 cm was noted in the pelvis immediately posterior to the rectus abdominis muscles (Fig. 2C). Emergency laparotomy revealed a 15 H 21 cm mass arising from the right ovary and extending into the right upper quadrant; this mass was adherent to the peritoneal reflection of the urinary bladder and was torsed on the infundibulopelvic vascular pedicle. An extraperitoneal mass measuring 4 cm and corresponding to the second mass noted at CT was removed. Pathologic examination revealed that the masses were composed of spindle cells, with minimal to no cytologic atypia and very rare mitoses; a diagnosis of LPD was rendered. The patient's postoperative course was uneventful.

Discussion

Definition and clinical presentation

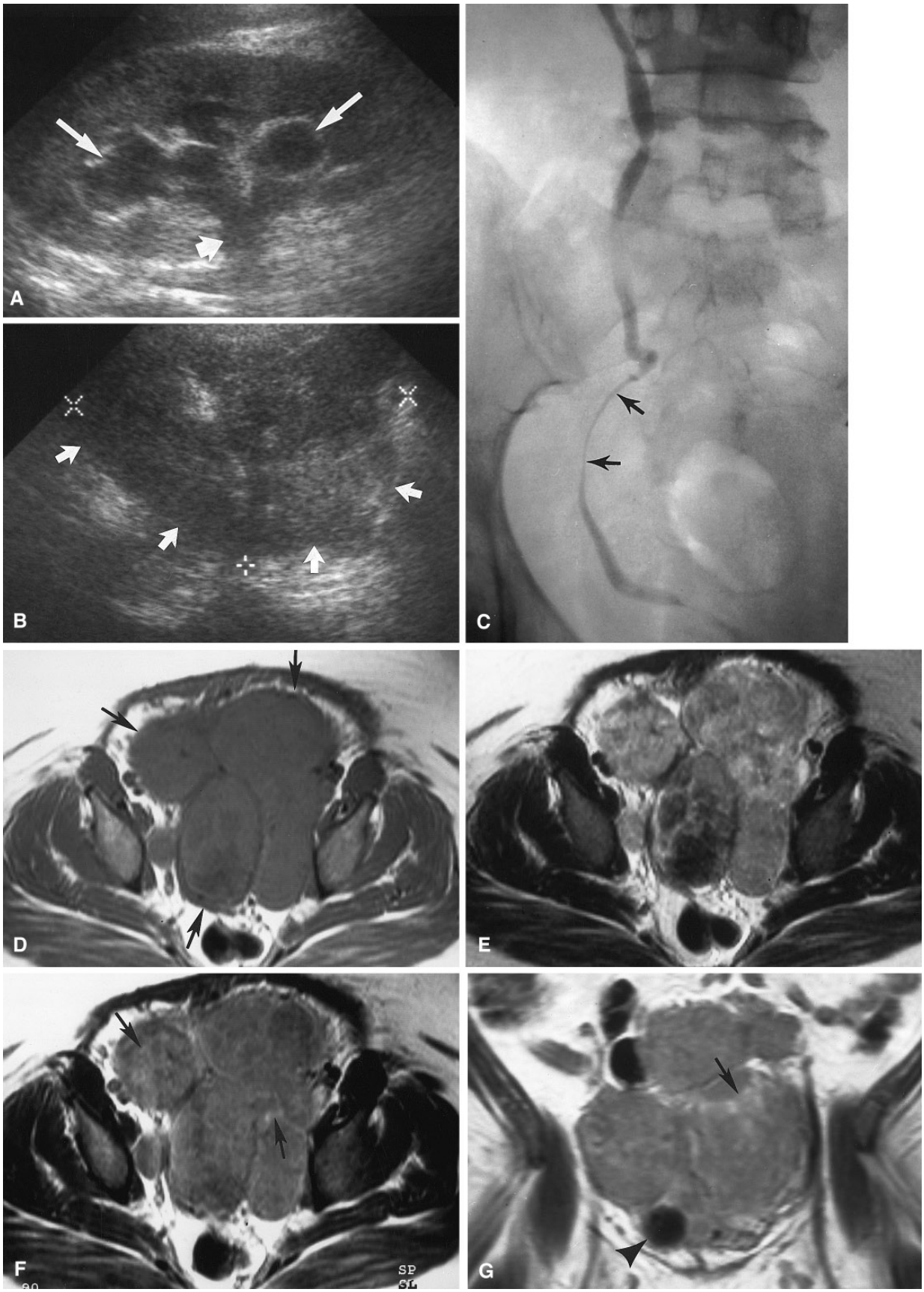
LPD is manifested as multiple, subperitoneal smooth muscle nodules [1]. LPD must not be confused with benign metastasizing leiomyoma, intravenous leiomyomatosis of the uterus, or diffuse leiomyomatosis of the uterus [17]. Benign metastasizing leiomyoma is characterized by a uterine leiomyoma that is associated with a benign smooth muscle tumor in a solid parenchymal organ such as liver or lung. Intravenous leiomyomatosis is defined as gross extension of a uterine leiomyoma into venous channels. Diffuse leiomyomatosis of the uterus represents extensive replacement of normal uterine parenchyma with innumerable benign leiomyomata.

The majority of LPD cases have been described in premenopausal women, many of whom were pregnant or taking oral contraceptives at the time of diagnosis [1–3, 6, 8, 18]. However, LPD has been diagnosed in postmenopausal women [5, 10, 11] and in a man [15]. Although many of the documented cases of LPD have been discovered incidentally during surgery, patients may present with symptoms such as discomfort, urinary frequency due to mass effect on the bladder, and gastrointestinal bleeding and peritonitis following erosion of the LPD implant into the bowel wall [7, 11, 14, 16, 18]. In our two cases, both patients experienced symptoms directly related to LPD: urosepsis secondary to obstruction of the ureters by the large pelvic mass and an acute abdomen due to ovarian torsion.

Pathologic features and proposed pathogenesis

Grossly, the LPD nodules are firm, well circumscribed, and tan-white with whorllike trabeculations on the cut surface. The nodules range in size from 0.5 mm to 20 cm and involve a number of structures including the omentum, mesentery, parietal peritoneum, broad ligaments, and surface of the ovary and uterus [1–3, 5]. There may be tens to hundreds of small nodules, and at first inspection, the appearance may mimic peritoneal carcinomatosis [6]. However, the nodules of LPD are located beneath the peritoneum rather than on the peritoneal surface, as occurs with peritoneal carcinomatosis. The histologic features of LPD are characteristic of benign uterine leiomyoma in that they are composed primarily of spindle cells and demonstrate a high degree of cellularity, hyalinization, and rare mitotic activity [1–3]. Decidual cells have been detected within the nodules in pregnant and postpartum patients [1]. However, some cases of LPD may present a diagnostic challenge to the pathologist and require electron microscopic examination to distinguish regressing LPD from sarcoma [7]. In some cases, such as our case 1, LPD undergoes sarcomatous transformation that is characterized by cytologic atypia and increased mitotic activity [14–16].

The pathogenesis of LPD remains uncertain. Some have postulated that LPD results from a predisposition of subperitoneal mesenchymal stem cells to differentiate into smooth muscle cells [1]. Because many cases of LPD have been detected in patients who are pregnant or taking oral contraceptives and because LPD often regresses after pregnancy, cessation of oral contracep-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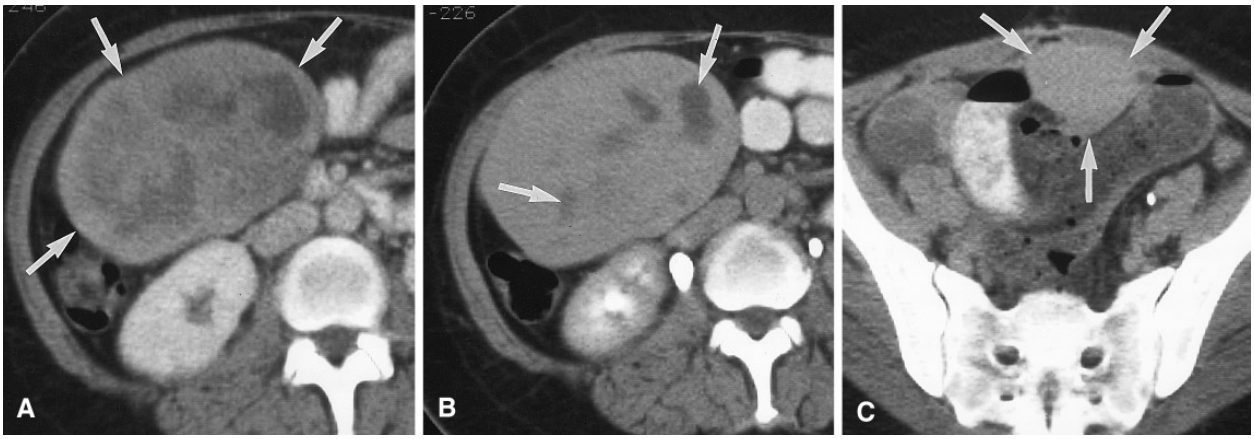

Fig. 2. A Enhanced axial CT shows a heterogeneous soft tissue mass (*arrows*) in the right mid abdomen. **B** Delayed axial CT obtained at a level similar to A reveals that portions of the mass do not enhance (*arrows*). **C** Enhanced axial CT of the pelvis demonstrates a 4-cm mass (*arrows*) lying posterior to the rectus abdominis muscles.

tives, or surgical castration, a hormonal influence is possible [1, 5, 7, 8]. However, other cases of LPD have been diagnosed in postmenopausal patients who have not taken hormonal therapy and in whom the disease has progressed despite surgical castration and removal of recurrent tumors, as occurred in our case 1 [9, 11]. Such cases suggest an increased tissue sensitivity to normal and diminished levels of estrogen and progesterone.

Imaging features

Atlhough the literature addressing the pathologic diagnosis of LPD is relatively extensive, little emphasis has been placed on its radiologic features, perhaps because many of the previous cases of LPD were diagnosed incidentally at laparotomy without any imaging performed prior to surgery. More recently, there have been reports of preoperative detection of LPD. The imaging findings will depend on the size and number of subperitoneal nodules. The difficulty in detecting small peritoneal implants in patients with peritoneal carcinoma-

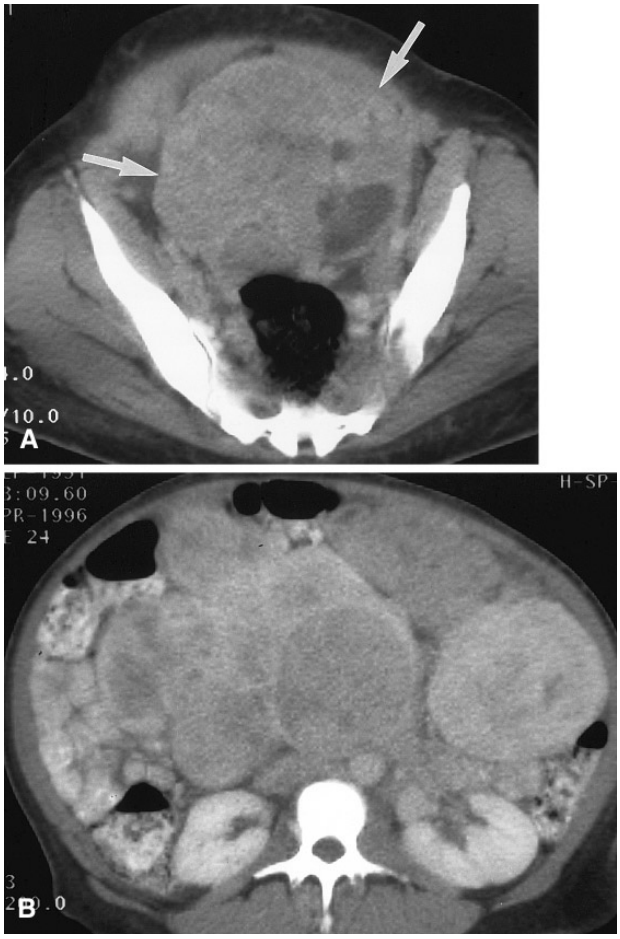

Fig. 3. A Enhanced axial CT of the pelvis shows a heterogeneous mass (*arrows*) arising from a uterus involved by diffuse leiomyomatosis. **B** Enhanced axial CT obtained at the level of the kidneys demonstrates multiple, apparently separate masses resembling LPD. At laparotomy, these masses were found to be pedunculated uterine leiomyomas.

Fig. 1. A Coronal sonogram of the right kidney demonstrates dilatation of the calyces (*arrows*) and renal pelvis (*short arrow*). **B** Transverse sonogram of the pelvis reveals a solid heterogeneous mass (*arrows*) filling the pelvis and mimicking a leiomyomatous uterus. **C** Contrast material introduced through a percutaneous nephrostomy tube opacifies the right ureter (*arrows*), which is displaced and partly obstructed by the pelvic mass seen on sonography. Similar changes involve the left ureter (not shown). **D** Axial T1-weighted image shows multiple pelvic masses (*arrows*) that are similar in signal intensity to skeletal muscle. **E** Axial T2-WI demonstrates slight heterogeneity of the pelvic masses. **F,G** Axial and coronal T1-weighted images obtained after the intravenous administration of gadolinium show heterogeneous enhancement of the masses (*arrows*). An air-filled balloon of a Foley catheter (*arrowhead*) is seen in the collapsed urinary bladder.

tosis is well recognized, and the same likely holds true in cases of LPD with numerous small nodules. Sonographic and CT findings in our cases and those of others reported in the literature include nonspecific, solid, and complex soft tissue masses that are often large and mimic a leiomyomatous uterus [4–6, 9, 14–16]. In some cases, the masses enhance in a fashion similar to that of normal uterine parenchyma, whereas others demonstrate heterogeneous enhancement. There may be confusion with peritoneal carcinomatosis if the masses are present diffusely throughout the abdomen and pelvis. Peritoneal carcinomatosis, however, is often associated with omental tumor cake, ascites, and liver metastases, which have not been reported with LPD. If the masses are located in the pelvis adjacent to the iliac vessels, they may be confused with lymphadenopathy.

MR findings include masses similar in signal intensity to skeletal muscle or uterine parenchyma on unenhanced T1-weighted images, which show variable degrees of enhancement following the administration of gadolinium. The masses of LPD remain low in signal intensity on T2-weighted images due to their smooth muscle components. In our case 1, in which sarcomatous transformation occurred, the MR features of the LPD implants did not differ significantly from the features reported for nonsarcomatous LPD implants.

When making the diagnosis of LPD, it is important to remember that in some cases, multiple, pedunculated leiomyomas arising from the uterus may mimic LPD implants (Fig. 3A,B).

Therapeutic options

Because in many instances LPD regresses after delivery, withdrawal of oral contraceptives, or castration without complete resection of all implants, many have advocated a conservative therapeutic approach. When LPD is encountered during laparotomy, some investigators have not advocated complete surgical excision of all implants [1, 7, 8, 12]. Because hormonal stimulation seems to play a role in progression of the disease, further use of oral contraceptives or additional pregnancies are not recommended after diagnosis of LPD. There has been one report of regression of LPD in a patient treated with a gonadotropin-releasing hormone agonist [18].

Although most cases of LPD follow benign courses, there have been reports of patients requiring multiple resections for recurrent disease, and there are at least five reports of malignant transformation [13–16]. These findings have led some to recommend close surveillance or aggressive resection in postmenopausal patients or in patients in the reproductive age group not desiring additional pregnancies.

Conclusions

Although LPD is a rare disorder that is often discovered incidentally, this entity is being reported with increasing frequency. For this reason, it is important for radiologists to be familiar with its clinical presentation, imaging features, and differential diagnostic possibilities so that LPD may be considered in the appropriate setting. In most instances LPD is uncomplicated, but in some cases LPD may undergo sarcomatous transformation or induce ovarian torsion.

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