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Extrauterine Leiomyomatosis Peritonealis Disseminata: A Malignant Mimicker of Benign Disease

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Abstract

Leiomyomatosis Peritonealis Disseminata (LPD), a form of extrauterine leiomyomatosis, is characterized by multiple subperitoneal benign nodules of smooth muscle origin. It is shown to be associated with conditions of excess gonadal steroids and hence, is most common in women of reproductive age group. However, cases of LPD have also been reported in postmenopausal women and men. LPD is a benign disease but mimics malignancy by its alarming appearance at surgery or radiological imaging. A high degree of clinical suspicion is thus warranted, to avoid radical surgeries in these patients. The majority of the cases of LPD are asymptomatic and detected incidentally while others present with symptoms like abdominal pain, menstrual irregularities, and palpable lump. No clear treatment guidelines have been established so far for LPD management, but currently available options include surgery, hormone therapy, and in some instances chemotherapy. LPD has been shown to recur. It also has a potential for malignant transformation. These patients need to be kept under observation with regular follow-ups. Other forms of extrauterine leiomyomatosis that have been reported in the literature are Parasitic Myoma (PM), Intravenous Leiomyomatosis (IVL), and Benign Metastasizing Leiomyomatosis (BML). Here, we described a rare case of multiple subperitoneal nodules in a young woman.

Keywords: Aromatase inhibitors; Benign; BML; Endometriosis; Extrauterine leiomyomatosis; GnRH agonists; Leiomyoma; Laparoscopic myomectomy; LPD; Malignant transformation; Parasitic myoma; Morcellation; Recurrent leiomyomas; SERM; Smooth muscle tumor; SPRM; Subperitoneal nodules; tamoxifen

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Background

Leiomyomatosis Peritonealis Disseminata (LPD) is a rare benign disorder. It is characterized by multiple, well-defined subperitoneal nodules [1,2]. It is frequently reported in women of reproductive age group, although some cases have also been reported in perimenopausal and postmenopausal women [3]. A few cases of LPD have been described in men also [4,5]. Most LPDs are asymptomatic and incidentally detected on radiology or at surgery for an unrelated cause [6]. Some patients may present with irregular/heavy menstrual bleeding with or without pain in the abdomen and peritonitis [1]. The tumor is composed of spindle-shaped smooth muscle cells having estrogen and progesterone receptors.

Case Presentation

A 27-year-old nulliparous, premenopausal female presented with complaints of swelling in the left side of the abdomen associated with mild, constant, non-cyclical pain for one year. There was no history of fever, burning micturition, change in bowel habits, bleeding per rectum, loss of appetite, or weight loss.

Two years ago, the patient had a history of dysmenorrhea and heavy menstrual bleeding, for which a six-month trial of oral hormonal therapy was administered. As the symptoms were persistent on hormonal treatment, she underwent laparoscopic myomectomy for multiple subserosal myomas elsewhere (data insufficient). Postoperatively there was transient relief in symptoms, but pain recurred and so, hormonal treatment was resumed after one year of surgery by a gynecologist.

The abdominal examination didn't reveal any swelling on inspection, but on deep palpation a single 4 cm × 4 cm firm, slightly painful, non-mobile, non-pulsatile, parietal wall swelling was felt

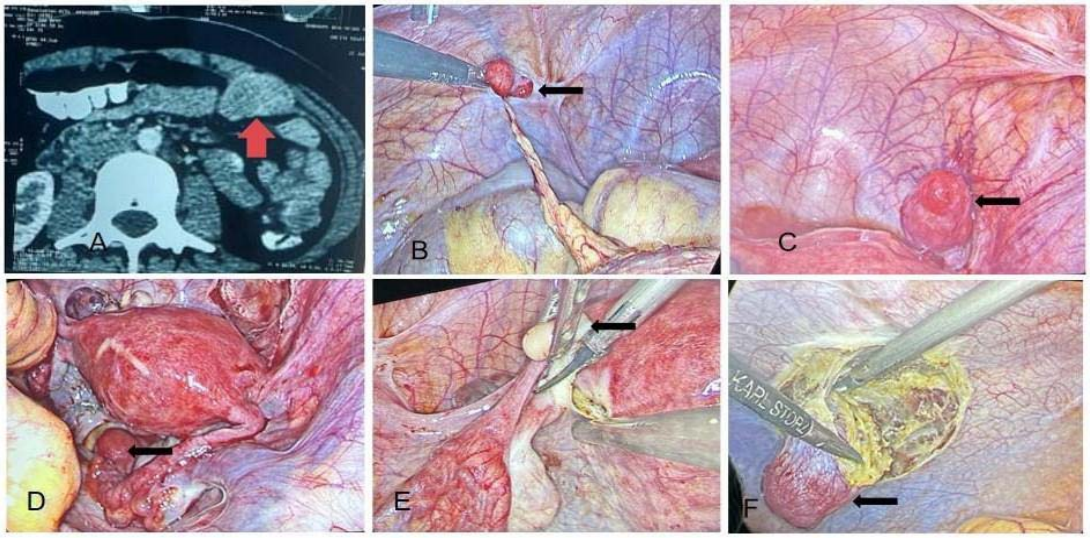

Figure 1: (A) CECT abdomen showing heterogenous swelling at the left anterior abdominal wall in the intramuscular plane with posterior extension into the abdominal cavity with maintained fat planes (red arrow); (B) Laparoscopic view (black arrow): Mesenteric swelling, C) Uterovesical pouch swelling, D) Swelling in the pouch of Douglas, E) Posterior uterine wall swelling, F) Left lumbar anterior abdominal wall swelling.

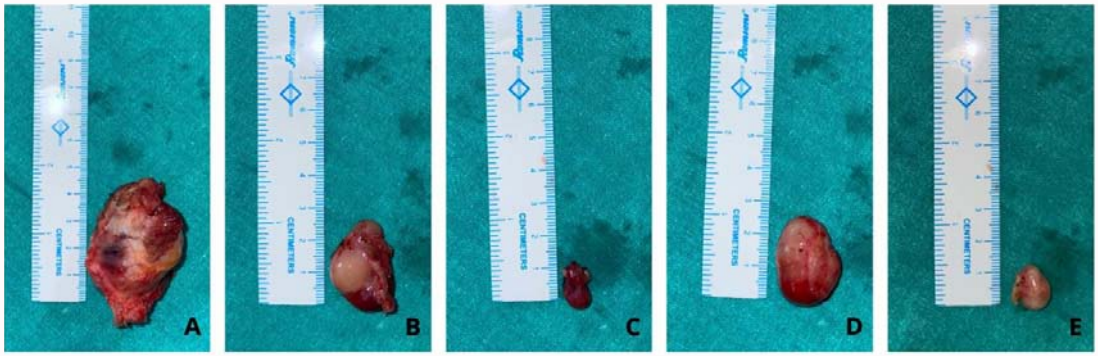

Figure 2: Gross images of excised swellings A) Left lumbar anterior abdominal wall swelling B) Right uterovesical pouch swelling C) Mesenteric swelling D) Swelling in pouch of Douglas E) Posterior uterine wall swelling.

at the left lumbar region. The swelling was well-defined and smooth surfaced but neither palpable bi-manually nor ballotable.

Ultrasound (USG) whole abdomen showed a well-defined, hypoechoic lesion, sized 3.3 cm × 3.9 cm × 4.3 cm. These features were suggestive of a collection at the anterior abdominal wall in the left para-umbilical region.

Contrast-Enhanced Computer tomography (CECT) of the whole abdomen showed multiple swellings. All swellings were well-defined, smoothly marginated, and heterogeneously enhancing with maintained fat planes. The first swelling was sized 2.3 cm × 3.7 × 3.3 cm within the left anterior abdominal wall in the intramuscular plane protruding intraperitoneally. The second swelling was sized 3.7 cm × 2.5 cm × 3.5 cm, within the mesentery of the proximal descending colon. The third swelling was sized 1.4 cm × 2.6 cm × 2.3 cm in the Pouch of Douglas.

Fine needle aspiration cytology of partial wall swelling suggested a benign spindle cell tumor. A preoperative probable diagnosis of the desmoid tumor was made.

Diagnostic laparoscopy showed five well-defined, distinct, sub-peritoneal swellings in the abdomen at the following locations: Left lumbar parietal wall, posterior wall of the uterus, right uterovesical pouch, small bowel mesentery, and Pouch of Douglas. The swellings were of variable size, the largest being 6 cm × 5 cm × 3 cm at the left side of the parietal wall to the smallest being 1 cm × 1 cm at the small bowel mesentery (Figure 1, 2). All swellings were reddish in color, firm, solid, well-defined, and had smooth margins. A small amount of free fluid was noted in the peritoneal cavity, which was sampled and sent for analysis. There was no evidence of endometriosis. The uterus was of normal size without peritoneal adhesions. The abdominal wall swellings were excised laparoscopically and a fresh frozen histopathological examination was carried out. Once the malignancy ruled out in the frozen section, all swellings were excised. The postoperative period was uneventful.

The detailed Histopathological (HPE) examination showed a moderately cellular spindled cell tumor with ovoid to fusiform nuclei with indistinct cytoplasm. These cells were arranged in a haphazard and ill-defined fascicular pattern, intermixed in the collagenous stroma with prominent dilated staghorn-type vasculature with occasional mitotic figures (2/10 high power field). There was no significant atypia or necrosis. Features were suggestive of benign spindle cell lesions, possibly leiomyoma (Figure 3). Peritoneal fluid

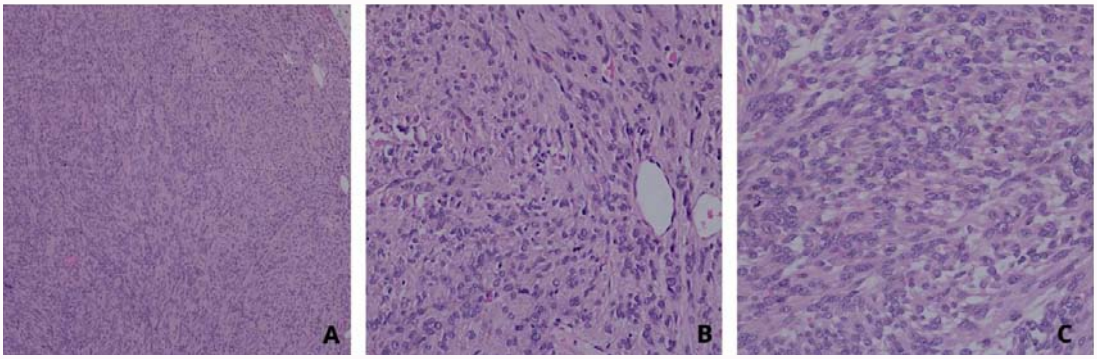

Figure 3: Microphotographs showing a circumscribed thinly encapsulated lesion composed of interlacing fascicles of spindle cells with cigar-shaped nuclei and eosinophilic cytoplasm and indistinct cell borders. No increase in cellularity atypia or mitotic activity is noted. HE A) 100X, B) 200X and C) 400X.

biochemical analysis and cytology were normal.

The patient is kept on radiological surveillance by USG once in six months. In two years of follow-up, the patient is doing well with no evidence of recurrence of pain or swelling.

Discussion

The first known case of LPD was reported in 1952 and a detailed description of the disease was given by Taubert et al. in 1964 [7]. To date, 250 cases of LPD have been reported so far. LPD is usually associated with conditions with excess gonadal steroid levels like pregnancy, hormone-secreting ovarian tumors, long-term oral contraceptives usage, hormone replacement therapy, and endometriosis [8], and hence, most common in women of reproductive age group.

Molecular studies have shown that cytoplasmic estrogen and progesterone bindings of LPD tumors are much greater than receptor concentrations in normal myometrium [9]. The most effective theory for the origin of LPD is that excess estrogen is responsible for the differentiation of sub-coelomic totipotent mesenchyme into fibroblast-like cells, which eventually differentiate into smooth muscle-like cells under the influence of both estrogen and progesterone [6]. However, this theory fails to explain LPDs in postmenopausal women, women with no known hormonal stimulus, and men. A second etiology proposed was local peritoneal implants from myometrial tissue, as few cases of LPD have been observed after laparoscopic hysterectomy/myomectomy that uses uncontained morcellation techniques [10]. Another etiology proposed was estrogen receptor modulation by drugs like tamoxifen [11].

The majority of the cases of LPD are asymptomatic, detected incidentally on radiology, at surgery, and in some instances, at autopsy [12]. Some patients present with palpable mass, abdominal pain, and menstrual problems (irregular cycles, dysmenorrhea). LPD can also present as intestinal obstruction, gangrene of the bowel, and peritonitis [1]. These nodules can impinge on intraabdominal structures leading to upstream obstruction resulting in conditions like hydroureter [13] One case of LPD implant on the ovary that caused ovarian torsion was also reported [14]. A few cases of LPD are associated with ascites [15].

Diagnosis of LPD can be made by imaging (USG, CT, MRI) with FNAC of the lesions where possible. If found incidentally at surgery, then excision followed by histopathological examination of the lesion can be diagnostic. On gross examination, these nodules are round, well-circumscribed, and firm. On microscopic examination, they are formed by spindle-shaped smooth muscle cells with less mitotic activity and no cellular atypia or necrosis [6].

Important differential diagnoses of LPD are peritoneal carcinomatosis, Parasitic Myoma (PM), Benign Metastasizing Leiomyoma (BML), and Intravenous Leiomyomatosis (IVL) [12]. LPD, PM, BML, and IVL are types of extrauterine leiomyomatosis. LPD mimics peritoneal carcinomatosis by its alarming appearance in radiology or at surgery. The multiplicity of lesions of LPD, its spread all over the peritoneum, and in rare instances, its association with lymphadenopathy and ascites contribute to its similarity with peritoneal carcinomatosis. But they can be differentiated by histopathology. Parasitic Myomas (PM) are characterized by one or two masses in the abdominal cavity. It is a leiomyoma that is, however, not attached to the uterus and derives its blood supply from surrounding organs and not the uterus itself (hence, named "parasitic") [16]. BML on the other hand, presents as metastasis of leiomyoma to solid organs, most commonly the lung. Other sites of metastasis include the spine, breast, pleurae, brain, ribs and vertebrae, appendix, parametria, heart, vessels, skeletal, muscle, soft tissue, lymph node, and retroperitoneum [17]. Clinical presentation of BML depends on the site of involvement such as respiratory symptoms when lungs are involved. IVL shows intravascular invasion by benign smooth muscle cells, which can proliferate and form a mass. It is clinically staged based on its progression along the blood vessels with early stages showing only involvement in the pelvis and later gradually progressing to involve the iliac veins/IVC, right atrium, and pulmonary artery. IVL can be asymptomatic or present with non-specific symptoms like pelvic mass, menstrual changes, phlebothrombosis, and right atrial mass, depending on the disease progression [18].

To date, there are no clear guidelines for treatment or follow-up of LPD. Historically, radical surgeries such as total abdominal hysterectomy, bilateral salpingo-oophorectomy, omentectomy, etc. were performed [1]. This could be owed to the alarming malignant appearance of the disease during surgery. Contrarily, few cases of spontaneous regression of disease have been reported (usually after menopause) [1,6]. This puts into question the relevance of surgical or other therapies in asymptomatic cases with accidental discovery.

Recently, with a better understanding of the disease, LPD has been managed more conservatively. The therapeutic strategy depends upon the patient's profile, reproductive desire, and clinical presentation.

In symptomatic cases of LPD, surgical excision, hormone therapy, or a combination of both can be considered [10]. The type of surgery depends on the patient's age and also the desire for reproduction. A patient in the reproductive age group can be taken up for excision of individual nodules either by laparoscopy or laparotomy. Menopausal and post-menopausal females can additionally undergo hysterectomy and oophorectomy [3].

Hormonal therapy for LPD has been reportedly effective due to the positive expression of hormone receptors on LPD nodules. Hence, therapies like Selective Estrogen Receptor Modulators (SERM), Selective Progesterone Receptor Modulators (SPRM), Aromatase Inhibitors (AI), and GnRH agonists (GnRH-a) have been attempted. Yang et al. and Nassif et al. showed that the use of Goserelin acetate (GnRH-a) as primary treatment in LPD resulted in a decrease in both volume and number of nodules along with symptomatic relief [9,19]. Leuprolide, also a GnRH-a, has been used in LPD by Benlolo et al. with satisfactory results [20]. Ando et al. and Takeda et al. described successful use of aromatase inhibitors like letrozole in the treatment of LPD with regression in the size of nodules and prevention of recurrence of new nodules [2,11]. SERMs like tamoxifen and raloxifene were not only found to be insufficient in controlling LPD but in fact, contributed to the growth of these lesions [11,21]. Ulipristal acetate, an SPRM, was used by Benlolo et al. and Verguts et al. in LPD with remission of the disease [20,22].

Quaranta et al. reported a case of refractory LPD that was unamenable to surgical, hormonal, or radiotherapy [23]. Xiao et al. reported a case of recurrent LPD with endometriosis that was not amenable to surgical excision and multiple cycles of hormonal therapy, necessitating Cytoreductive Surgery (CRS) with Hyperthermic Intraperitoneal Chemotherapy (HIPEC) [24]. Drugs like sirolimus and systemic chemotherapeutic agents like doxorubicin and dacarbazine have also been used in recurrent cases of LPD [25,26]. Therefore, it is ideal to keep such patients in close follow-up with periodic clinical examinations along with radiological (USG/CT/MRI) or diagnostic laparoscopic examinations.

Although a benign disease with a good prognosis to begin with, LPD has been reported to show malignant/sarcomatous transformation with less than 10 cases reported so far [4,5,7,14,24].

Conclusion

A high level of clinical suspicion is warranted in LPD to avoid radical procedures thereby significantly reducing the morbidity. The LPD can be managed more conservatively with individualized approaches. Medical management can be used as primary treatment or as an adjunct to surgical excision. There is always the possibility of recurrence and the potential for malignant transformation, so strict follow-up of the entity is recommended [1,24].

References

1. Gaichies L, Fabre-Monplaisir L, Fauvet R, Alves A, Mulliri A. Leiomyomatosis peritonealisis disseminata: Two unusual cases with literature review. J Gynecol Obstet Hum Reprod. 2018;47(2):89-94.
2. Ando H, Kusunoki S, Ota T, Sugimori Y, Matsuoka S, Ogishima D. Long-term efficacy and safety of aromatase inhibitor use for leiomyomatosis peritonealis disseminate. J Obstet Gynaecol Res. 2017;43(9):1489-92.
3. Edwards J, Katali H. A novel case of fibroids after menopause. Case Rep Obstet Gynecol. 2018;2018:9104719.
4. Yamaguchi T, Imamura Y, Yamamoto T, Fukuda M. Leiomyomatosis peritonealis disseminata with malignant change in a man. Pathol Int. 2003;53(3):179-85.
5. Yu RS, Wang ZK, Sun JZ, Chen LR. Computed tomography of pancreatic implantation with malignant transformation of leiomyomatosis peritonealis disseminata in a man. Dig Dis Sci. 2007;52(8):1954-7.
6. Li J, Dai S. Leiomyomatosis peritonealis disseminata: A clinical analysis of 13 cases and literature review. Int J Surg Pathol. 2020;28(2):163-8.
7. Xu S, Qian J. Leiomyomatosis peritonealis disseminata with sarcomatous transformation: A rare case report and literature review. Case Rep Obstet Gynecol. 2019;2019:3684282.
8. Mabrouk M, Arena A, Raimondo D, Parisotto M, Caprara G, Seracchioli R. Leyomiomatosis peritonealis disseminata associated with ovarian endometriosis in a patient submitted to hysteroscopic myomectomy. Fertil Steril. 2019;111(6):1259-61.
9. Yang JW, Hua Y, Xu H, He L, Huo HZ, Zhu CF. Treatment of leiomyomatosis peritonealis disseminata with goserelin acetate: A case report and review of the literature. World J Clin Cases. 2021;9(19):5217-25.
10. Chen X, Liu H, Shi H, Fan Q, Sun D, Lang J. Leiomyomatosis peritonealis disseminata following laparoscopic surgery with uncontained morcellation: 13 cases from one institution. Front Surg. 2021;8:788749.
11. Takeda T, Masuhara K, Kamiura S. Successful management of a leiomyomatosis peritonealis disseminata with an aromatase inhibitor. Obstet Gynecol. 2008;112(2 Pt 2):491-3.
12. Kim H, Cheong H. Leiomyomatosis peritonealis disseminata: An incidental finding at autopsy and review of literature. Am J Forensic Med Pathol. 2023;44(2):e10-e12.
13. Urman B, Misirlioglu S, Aksu S, Ata B, Taskiran C. Leiomyomatosis peritonealis disseminata that caused hydroureter in association with deep infiltrating endometriosis. J Minim Invasive Gynecol. 2015;22(6S):S130.
14. Fulcher AS, Szucs RA. Leiomyomatosis peritonealis disseminata complicated by sarcomatous transformation and ovarian torsion: Presentation of two cases and review of the literature. Abdom Imaging. 1998;23(6):640-4.
15. Nappi L, Sorrentino F, Angioni S, Pontis A, Barone I, Greco P. Leiomyomatosis Peritonealis Disseminata (LPD) ten years after laparoscopic myomectomy associated with ascites and lymph nodes enlargement: A case report. Int J Surg Case Rep. 2016;25:1-3.
16. Quaranta M, Mehra G, Nath R, Culora G, Sayasneh A. A rare case of refractory disseminated leiomyomatosisperitonealis complicated by cauda equina compression. Eur J Obstet Gynecol Reprod Biol. 2018;229:205-7.
17. Wang J, Liu G, Yang Q. Parasitic myoma after transabdominal hysterectomy for fibroids: A case report. BMC Womens Health. 2023;23(1):310.
18. Barnaś E, Książek M, Raś R, Skręt A, Skręt-Magierło J, Dmoch-Gajzlerska E. Benign metastasizing leiomyoma: A review of current literature in respect to the time and type of previous gynecological surgery. PLoS One. 2017;12(4):e0175875.
19. Tan Y, Han J, Wang Z, Yan J, Dong L, Liu R. Different clinical diagnoses end up in the same pathological diagnosis of intravascular leiomyomatosis: Two case reports. Medicine (Baltimore). 2024;103(2):e36887.
20. Nassif GB, Galdon MG, Liberale G. Leiomyomatosis peritonealis disseminata: Case report and review of the literature. Acta Chir Belg. 2016;116(3):193-6.
21. Benlolo S, Papillon-Smith J, Murji A. Ulipristal acetate for disseminated peritoneal leiomyomatosis. Obstet Gynecol. 2019;133(3):434-6.
22. Bayya J, Minkoff H, Khulpateea N. Tamoxifen and growth of an extrauterine leiomyoma. Eur J Obstet Gynecol Reprod Biol. 2008;141(1):90-1.
23. Verguts J, Orye G, Marquette S. Symptom relief of leiomyomatosis peritonealis disseminata with ulipristal acetate. Gynecol Surg.

2014;11(1):57-8.

24. Xiao X, Wang C, Zhang Y, Li F, Zhang H, Ma R. Combination therapy of cytoreductive surgery and hyperthermic intraperitoneal chemotherapy for recurrent leiomyomatosis peritonealis disseminata with endometriosis: A case report. Heliyon. 2023;9(9):e19794.

25. Zhang G, Fan R, Yang H, Su H, Yu X, Wang Y, et al. Safety and efficacy of sirolimus in recurrent intravenous leiomyomatosis, pulmonary benign metastatic leiomyomatosis, and leiomyomatosis peritonealis disseminata: A pilot study. BMC Med. 2024;22(1):119.

26. Lin YC, Wei LH, Shun CT, Cheng AL, Hsu CH. Disseminated peritoneal leiomyomatosis responds to systemic chemotherapy. Oncology. 2009;76(1):55-8.