Leiomyomatosis Peritonealis Disseminata: MRI Features Before and After Treatment with GnRH Agonist

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ABSTRACT

Leiomyomatosis peritonealis disseminata (LPD) is a rare disorder characterized by multiple subperitoneal benign smooth muscle nodules. It is usually incidentally discovered during cesarean section or laparoscopy. In this paper, we present pre-and-post GnRH treatment MRI findings of a symptomatic patient with LPD.

Key words: *Leimyomatosis peritonealis disseminata, MRI, GnRH agonist*

Yaygın Leyomatozis Peritonalis: GnRH Analoğu ile Tedavi Öncesi ve Sonrası MRI Bulguları

ÖZET

Leyomyomatozis peritonealis disseminata (LPD) yaygın subperitoneal benign natürlü nodüler lezyonlar ile karakterize nadir bir hastalıktır. Hastalık, genellikle sezeryan sırasında incidental olarak saptandığı için literatürde MR görüntüleme bulgularını anlatan çok az sayıda makale bulunmaktadır. Bu makalede semptomatik bir LPD olgusunun GnRH tedavi öncesi ve sonrası MR bulguları sunulmuştur.

Anahtar kelimeler: *Yaygın leyomatozis peritonalis, MRI, GnRH analoğu*

INTRODUCTION

Leiomyomatosis peritonealis disseminata (LPD) is a rare benign disease characterized by multiple subperitoneal smooth muscle nodules through out the abdominal cavity (1,2). Although these nodules are macroscopically similar to peritoneal carcinomatosis, they are histologically benign in nature (3). The diagnosis of LPD is generally established incidentally during cesarean section, laparotomy, or laparoscopy. Rarely, these nodules can also be detected incidentally in asymptomatic patients during diagnostic cross-sectional imaging studies that are being performed for other purposes. The differentiation of LPD from peritoneal carcinomatosis or other disseminated abdominal malignancies is important since the treatment of these entities are completely different (2). Because of its histological nature, LPD has typical MRI features, and MRI can be reliably used to differentiate this benign entity from malignancy (4, 5). In this paper, we present MRI features of a symptomatic LPD patient.

CASE

A 36-year-old Caucasian woman presented to our institution with heavy vaginal bleeding, shortness of breath and right upper quadrant discomfort. She had two hysteroscopic and one laparoscopic myomectomies in the past due to severe menorrhagia and dysmenorrhea, and had pathological of diagnoses of LPD. She denied any oral contraceptive usage. US showed several solid masses in the pelvis adjacent to the uterus. MRI imaging

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performed with a protocol including multiplanar FRFSE T2-weighted (TR/TE: 2500/86 ms, ETL: 12), multiplanar, 3D, spoiled gradient-echo (TR/TE: 250/4.2, flip angle: 75°) with fat suppression before and after administration of 20 cc of IV gadolinium (Magnevist, Berlex Laboratories, Wayne, NJ) showed a 2.6 x 2.2 cm well-defined, low T2 signal intensity mass within the uterine fundus, most likely representing a submucosal fibroid. In addition to the masses seen on US, MRI showed innumerable, well- defined peritoneal masses within the pelvis and abdomen, the largest (5.6 x 4.6 cm) located in the presacral area. The majority of the masses had low-T2, and intermediate-T1 signal intensity compare to muscle, and demonstrated diffuse enhancement with contrast. Some masses had high T1 signal intensity, and others were heterogeneous on T2-weighted images with signal intensity ranging from homogeneously low signal intensity to central hypointensity with peripheral hyperintensity, to nearly complete hyperintensity (Figure 1 a-d). The ovaries were normal. There were also several subdiaphragmatic peritoneal masses adjacent to the dome of the liver with similar signal intensity and enhancement characteristics to the pelvic ones (Figure 1e). After MRI examination, it was considered that the resection of submucous fibroid would likely improve her menorrhagia, and she underwent another histeroscopic myemoctomy. Due to her some persisting symptoms after myomectomy, she was treated with GnRH and her symptoms had improved. However, she developed hot flashes and diffuse muscular pain secondary to the treatment. Follow-up MRI revealed a response to treatment manifested by decrease size of some of the leiomyomas (Figure 1e-f). In addition, few leiomyomas have developed central T2 hyperintense unenhancing areas consistent with cystic degenerations.

DISCUSSION

LPD is characterized by multiple subperitoneal nodules of varying size from a few millimeters to 20 centimeters and grossly resembles peritoneal carcinomatosis. The nodules can seen anywhere in the peritoneal cavity including uterus, fallopian tubes, ovaries, small and large intestines, and mesentery (5, 6). Histologic features of LPD consist smooth muscle cells with little or no pleomorphism, and no mitotic figures (7). Histologic appearances of LPD sometimes may mimic low-grade leiomyosarcoma and requires electron microscopic examination for differentiation (5, 7).

LPD typically seen in women in their reproductive ages although there are few reported case of LPD seen in man or postmenopausal woman (6, 8). The exact etiology of LPD is unknown, but it is thought to be related with hormonal stimulation. Thus, many of the reported cases were presented during or following pregnancy, during oral contraceptive usage, or associated with estrogen-producing granulose cell tumor (3-5). The regression of the LPD after bilateral oophorectomy or after administration of GnRH agonist supports this hypothesis (4-7). Although our patient does not have any cause for hormonal stimulation, showed partial response to GnRH agonist treatment.

Since most reported cases detected incidentally during a laparotomy, imaging findings of LPD had not been described thoroughly. Sonography and CT scan findings are generally nonspecific including multiple, solid nodules with well-defined contours on the subperitoneal surfaces without ascites or other signs of malignancy. Therefore peritoneal carcinomatosis is considered the first in differential diagnosis. But peritoneal carcinomatosis is typically secondary to a known primary, and commonly associated with ascites and other organ metastases. The differentional diagnosis of LPD also includes peritoneal mesothelioma, tuberculosis, desmoid tumours, and lymphoma (4, 7). Our MRI findings were similar to previous reports (4, 5). T2-weighted images are particularly useful to differentiate LPD from other diseases presenting with multiple peritoneal nodules. Due to their fibrous nature, leiomyomas typically demonstrate diffuse, low T2 signal intensity. However as seen in our case, some leiomyomas may show heterogeneous T2 signal intensity, which is attributed to various histopathologic patterns of degeneration such as hyaline, cystic, myxoid, and red degenerations (4, 9). Red or so-called carneous degeneration is due to obstruction of the draining veins resulting in extensive coagulative necrosis that involves the entire leiomyoma, and typically causes severe abdominal pain (9). Leiomyomas undergoing red degeneration appear as high signal intensity mass on T1-weighted images.

There are many treatment options for LPD reported in the literature (5-8). Many investigators have advocated a conservative therapeutic approach without extensive surgery because, in many instances, LPD showed regression after delivery or withdrawal of oral contracep-

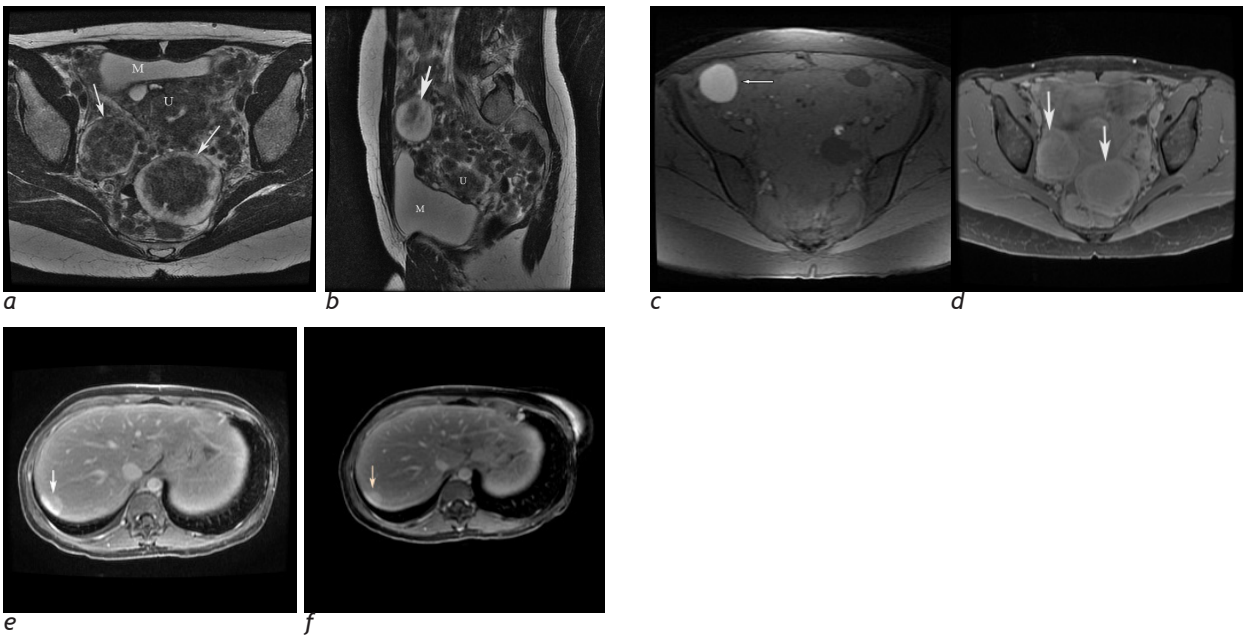

***Figure 1 a-f**. Axial (a) and sagittal (b) T2-weighted MR images show numerous, well-defined, extrauterine masses in the pelvis. While the small masses are homogeneously low in signal intensity, some large masses (arrows) show central hypo, peripheral hyperintense signal. Axial, fat-suppressed, unenhanced (c) and enhanced (d) T1-weighted spoiled gradient echo MR images obtained cranial to the images on Figures 1a and b show that the masses are isointense with muscle, excluding the one in the right lower quadrant (arrow on c), which demonstrates hyperintense signal likely due to red degeneration, and enhance diffusely (d) with gadolinium. Axial, fat-suppressed, enhanced T1-weighted spoiled gradient-echo MR images at the level of the liver before (e) and after (f) treatment with GnRH anolog show a diffusely enhancing subdiaphragmatic leiomyoma (arrows) adjacent to the liver. Note decrease size following treatment, which suggests treatment response.*

tives or castration without completes resection of all implants. Because of strong association between these tumors and increased hormonal production, recent studies have used GnRH as a therapeutic measure. TAH with unilateral or bilateral salpingo-oopherectomy is only recommended for symptomatic relief in cases with completed families.

Although LPD is a benign tumor of smooth muscle cells, malignant transformation can rarely occur. In the literature, six cases of malignant transformation to leiomyosarcoma have been described (6). Malignant transformation in these patients occurred 4-12 months after the initial diagnosis. For this reason, patients with LPD need to be followed up with imaging, preferentially with MRI. MRI examinations are also very useful method for following response to GnRH agonist treatment.

In summary, early diagnosis of LPD is very important to initiate appropriate medical treatment and to prevent unnecessary surgery. US examination may fail to reveal extent of the disease whereas MRI is the best imaging method to diagnose LPD noninvasively, and follow the respond to the treatment.

REFERENCES

1. *Sharma P, Chaturvedi KU, Gupta R, Nigam S. Leiomyomatosis peritonealis disseminata with malignant change in a post-menopausal woman. Gynecol Oncol 2004;95: 742-5.*
2. *Halama N, Grauling-Halama SA, Daboul I. Familial clustering of Leiomyomatosis peritonealis disseminata: an unknown genetic syndrome? BMC Gastroenterol 2005;33: 1-5.*
3. *Nappi C, Di Spiezio Sardo A, Mandato VD, et al. Leiomyomatosis peritonealis disseminata in association with Currarino syndrome? BMC Cancer 2006; 6:127.*

4. Ruscalleda N, Eixarch E, Pages M, et al. Leiomyomatosis peritonealis disseminata. Eur Radiol 2006;16: 2879-882.

5. 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: 640-4.

6. Bekkers RL, Willemsen WN, Schijf CP, Massuger LF, Bulten J, Merkus JM. Leiomyomatosis peritonealis disseminata: does malignant transformation occur? A literature review. Gynecol Oncol 1999;75: 158-63.

7. Papadatos D, Taourel P, Bret PM. CT of leiomyomatosis peritonealis disseminata mimicking peritoneal carcinomatosis. AJR Am J Roentgenol 1996;167: 475-6.

8. Kokcu A, Alvur Y, Baris YS, Kuskonmaz I. Leiomyomatosis peritonealis disseminata. Acta Obstet Gynecol Scand 1994;73: 81-3.

9. Ueda H, Togashi K, Konishi I, et al. Unusual appearances of uterine leiomyomas: MR imaging findings and their histopathologic backgrounds. Radiographics 19: 1999;131-45.