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Case Report

Leiomyomatosis peritonealisis disseminata: Two unusual cases with literature review

L. Gaichies [a,*], L. Fabre-Monplaisir [b], R. Fauvet [a,c], A. Alves [c,d], A. Mulliri [d]

[a] Gynécologie–obstétrique et médecine de la reproduction, CHU de Caen, 14000 Caen, France
[b] Anatomie et cytologie pathologique, CHU de Caen, 14000 Caen, France
[c] Pôle des formations et de recherche en santé, université de Caen-Normandie, 2, rue des Rochambelles, 14000 Caen, France
[d] Chirurgie digestive, CHU de Caen, 14000 Caen, France

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ABSTRACT

Background. – Leiomyomatosis peritonealis disseminata (LPD) is a rare benign disease characterized by numerous smooth muscle-like cell nodules disseminated among the abdominal cavity. The pathogenesis of LPD is not well-known; one hypothesis, widely reported, is parasitic LPD (after uterine myoma surgery). The role of hormonal status has been raised without any evidence yet confirmed. Lesions of LPD can mimic carcinomatosis and lead to an inappropriate treatment. Total and spontaneous regression of the nodules is sometimes reported. This benign disease is also associated with leiomyosarcoma or invasive lesions, leading to mechanical complication or death.

Cases and review. – We report two additional cases. The first case occurred in a 78-year-old woman with a history of total hysterectomy for uterine myoma, and was revealed by a small bowel obstruction. Second case occurred in a 50-year-old non-menopausal woman suffering from obesity (BMI 61,7), with an incidental diagnostic of LPD during a surgical procedure and spontaneous full regression of the disease. We analyzed 165 articles. We fond 16 menopausal women with LPD, 5 patients with intestinal obstruction or peritonitis due to LPD and 5 cases with spontaneous regression of LPD lesions. Six cases of recurrence as leiomyosarcoma were found.

Conclusion. – Cases of LPD have been reported without uterine myoma history and could be from extrauterine origin. Hysterectomy and oophorectomy should not be performed in first line. Peritoneal cytology should be done in case of ascites. Recurrences as leiomyosarcoma are reported and occurs early and justify a close follow up the first years.

Introduction

Leiomyomatosis peritonealis disseminata (LPD) is a rare disease characterized by the dissemination of multiple smooth muscle nodules throughout the peritoneal surface.

Nowadays, 193 cases have been reported. The first case was described in 1952 [1], it was designated as LPD 10 years later [2].

LPD occurs preferentially in non-menopausal women. History of uterine myoma morcellation and hormonal influence have been incriminated in the physiopathology. One recent publications highlights a link between laparoscopic open morcellation of uterine myomas and LPD incidence (0.12 to 0.95% LPD incidence after morcellation) [3].

LPD lesions often involve the pelvis, especially the Douglas pouch, but the disease can reach the entire abdomen cavity: omentum, mesentery, and become invasive with non-extricable lesions reaching for example ureters and bladder, retroperitoneal space, liver, small bowel or mesentery [4–9].

Most of the time, the lesions look like numerous infracentimetric white-grey firm nodules. Those nodules can be mixed with a solid and cystic component and sometimes it can be hemorrhagic.

The association of ascites and adenopathy can lead to confusion with the diagnosis of peritoneal carcinomatosis. Furthermore, malignancy evolution to leiomyosarcoma, with a high mortality rate, is now well-described [10–18].

Those elements suggest that a reliable diagnosis and an appropriate follow-up are essential to provide patients the best therapeutic options.

However, reported therapeutic options and patient follow-up are very heterogeneous. Epidemiological features, preoperative diagnostic and patient follow-up options remain poorly known.

We report two unusual cases. One occurred on a post-menopausal woman and was revealed by a bowel obstruction.

* Corresponding author.
E-mail address: leo.gaichies@gmail.com (L. Gaichies).

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Table 1
Characteristics of the two reported cases.

Case	Age	G-P H S	Myoma history treatment	Clinical features at diagnosis	Involvement	Surgical procedure	Follow-up (month)	Recurrence Follow-up (month)
1	78	1–1	Type 2 myoma	Small bowel obstruction	Entire abdomen	Bowel resection	12	0
2	50	0 M−	0	Asymptomatic	Small Bowel stenosis Ascites Mesentery retraction	Biopsies	48	12 (Total regression) 48

G-P: gravida-para; H S: hormonal status, M+: menopausal, M−: non menopausal; TH: total hysterectomy.

The other case seems to be an invasive disease with total and spontaneous regression of the lesions.

We reviewed the literature to define appropriate therapeutic options according to the similar reported cases.

Cases report

Table 1 summarizes the characteristics of the two reported cases.

Case 1

A 78-year-old woman was referred to the general surgery department of the CHU of Caen (Caen, France) in July 2016 for an acute intestinal obstruction. This gravida 1 para 1 (caesarean section) Caucasian patient had a history of type 2 uterine myoma (according to the 2009 FIGO classification), treated by abdominal total hysterectomy without salpingo oophorectomy in 1988. Her BMI was 17.5 kg/m^2; she was treated for arrhythmia and suffered from sigmoiditis without surgical treatment. She took an oral contraceptive (panovlar) before menopausal and never had hormonal treatment afterwards.

The patient remained asymptomatic before an acute abdominal syndrome due to a small bowel obstruction. She had intense abdominal pain especially in right lower belly area. An abdominal CT Scan after injection of intravenous contrast showed a small bowel obstruction with junction in right iliac area (Fig. 1). A 53 × 51 mm cystic and solid mass was developed the right iliac area (Fig. 2). Numerous peritoneal nodes, especially omentum nodes, were seen. No ascites, no lymph node enlargement, and no visceral anomalies were noted.

The serum carbohydrate antigen 125 (CA 125) level was 65.8 U/mL (normal range: 0–35 U/ml); carbohydrate antigen 19–9 was within the normal range (0.6 U/mL); Carcinoembryonic antigen (ACE) was 2.5U/mL (normal range: 0–5,99 ng/mL). Blood test showed no anomaly except inflammatory syndrome and moderate malnutrion.

Injected abdominal CT scan showing the cystic mass and small bowel occlusion.

The patient had an explorative laparotomy. She was treated in emergency context with mechanical occlusion requiring surgery because of severe abdominal pain and gravity signs on CT scann. Ovarian or intestinal carcinoma with peritoneal extension or a pseudomyxoma peritonei were strongly suspected. An explorative laparoscopic procedure was therefore debatable in first line. However, because of the strong intestinal distention, surgical history and probable carcinomatosis, a laparotomy seemed more cautious.

During this procedure, numerous grey-red lesions were identified, measuring 1–20 mm in diameter, involving the mesentery, greater omentum, peritoneum, Douglas's pouch, serosal surface of the small and large bowel. Both ovaries, as well as the appendix, were normal in appearance and size (especially no ovarian cyst). There were no ascites or lymph node enlargement.

A mechanical obstruction of the last ileal loop was noted, due to an epiploic node involving bowel serosa.

Considering a peritoneal extension of a digestive tract carcinoma, no frozen-section analysis was done during surgery. The patient underwent a small bowel resection with cystic mass resection and ileostomy to cure the obstruction. A partial omental resection and excision of some of the disseminated lesions were performed.

The final histological examination confirmed the diagnosis of LPD.

The cystic mass was a necrosed epiploic fringe. Nodules consisted of non-encapsulated spindle-shaped mature smooth muscle cells. Mitotic figures were very infrequent throughout the tumors. There was not any histological feature of malignancy, necrosis, mitotic activity or nuclear atypia. The immunohistochemical test for Desmin was positive and negative for CD34 and CD117. This profile was concordant with the diagnosis of leiomyomatosis, defined as a smooth muscle proliferation.

Desmin and SMA (smooth muscle actin) are two markers present in smooth muscle cells, classically positive in leiomyomatosis. CD34 is a low specific vascular marker, and is also positive in gastrointestinal stromal tumors (GIST) and solitary fibrous tumors for exemple. This marker enables to eliminate potential differential diagnosis of leiomyomatosis. It should be negative in cases of leiomyomatosis.

CD117 reflects an activating mutation of C-KIT receptor gene in most of GIST. It can sometimes be positive in other fusiform tumors

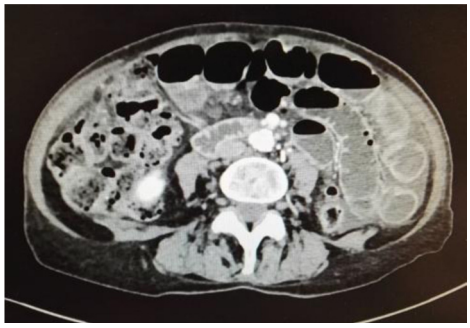

Fig. 1. Small bowel obstruction.

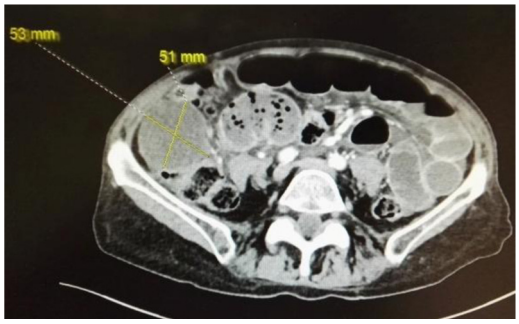

Fig. 2. Cystic mass.

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