Leiomyomatosis peritonealis disseminata and pregnancy: a case report

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Summary

Leimyomatosis peritonealis disseminata (LPD) is a benign tumor of smooth muscle tissue. It is rare and is characterized by the development of multiple peritoneal nodules mimicking peritoneal carcinomatosis. We report a case of LPD diagnosed in a 35-year-old patient, G4/P1, without any major gynecological history. The patient underwent an elective cesarean section at 42 weeks, during which numerous peritoneal nodules ranging in size from 0.1 to 0.5 cm were found. Microscopic examination showed a proliferation of smooth-muscle cells without mitosis or atypia or necrosis.

Key words: Leiomyomatosis peritonealis disseminata; Leiomyoma.

Introduction

Leiomyomatosis peritonealis disseminata (LPD) is a benign tumor of smooth muscle tissue and is characterized by the proliferation of multiple peritoneal nodules. Nodules of variable size are found throughout the peritoneal surface, on the omental apron simulating a peritoneal carcinomatosis, and often coexist with uterine fibroids [1] although no clear correlation between LPD and uterine fibroids has been established [2]. We present the second reported case in the Ivory Coast since the one published in 2006 [3].

Case Report

A 35-year-old woman (G4P1) was admitted to the maternity ward of Cocody University Hospital (Abidjan, Ivory Coast) for an elective cesarean section indicated due large fetus size to in October 2009. Her first child had been delivered vaginally. She had irregular cycles between 28 and 32 days and menstrual duration was four to five days. She had never used contraceptives and had no history of uterine fibroids. She also had had two terminations of pregnancy with an uneventful postoperative course. On admission, we noted a good general condition: blood pressure = 11/7; weight = 77 kg; height = 161 cm. Uterine size was estimated at 37 cm. The fetal heart sounds were regular at 150 bpm. The uterus appeared regular and soft to the touch. Speculum examination showed a vagina with no abnormalities. On vaginal examination, the inferior segment of the uterus was small and the cephalic presentation repressed. Other examinations were unremarkable. Obstetric ultrasound revealed a fetal weight superior to 4,400 g. Laboratory tests as part of the preoperative evaluation were normal. Cesarean section allowed an delivery with macrosomia, weight: 4,600 g, Apgar score 8-9 at 1 and 5 min. At cesarean section, multitude nodular formations ranging in size from 0.1 to 0.5 cm scattered throughout the pelvic cavity were revealed. These nodules were visible on the right broad ligament, the bladder-uterine peritoneum, and the pelvic colon on the omentum (Figures 1 to 4). Before closing the abdomen, biopsy was performed for nodular histological examination. The exam highlighted a proliferation of smooth muscle fibers without mitosis, atypia, or necrosis. The postoperative course was uneventful and the patient was releals from hospital on the 7th postoperative day. No special treatment was required in the postoperative course, and the postoperative consultation six weeks after surgery was normal.

Discussion

Frequency

LPD is a rare disease. Since the first case reported by Wilson and Peale in 1952, a broad review of the Anglo-Saxon literature has identified to date 132 cases reported by more than 80 teams, including 113 women in the reproductive group age, seven postmenopausal women, six cases in males and one case in an animal (a horse) [4]. Despite the abundance of literature on LPD, the etiopathogenesis, diagnosis and treatment are still unclear.

Etiopathogenesis

The intimate mechanism of development of LPD is still today a mystery; however *the role of estrogen* in the genesis of this disease has been discussed in numerous publications [3, 5, 6].

Animal studies have shown that high rates and prolonged elevated levels of estrogen can induce metaplasia of mesenchymal stem cells into fibroblastic, leiomyocytes or endometrial stroma [7]. These animals developed disseminated leiomyomatous peritoneal lesions similar to LPD. High levels of estrogen and progesterone were found on LPD cells suggesting a real potential hormonal responsiveness of these tumors. Finally the high frequency of LPD during genital activity including pregnancy (our observation) validates the idea of involving hormonal activity in the genesis of LPD. Thus the temptation is great to introduce hormonal therapy in the management of LPD.

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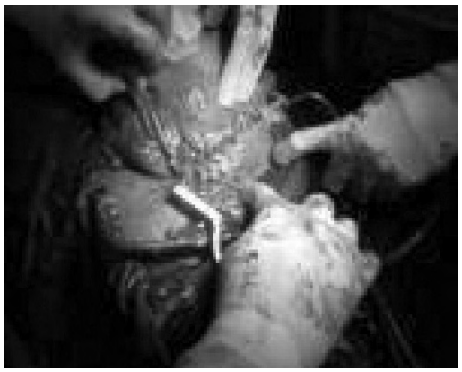
Fig. 1



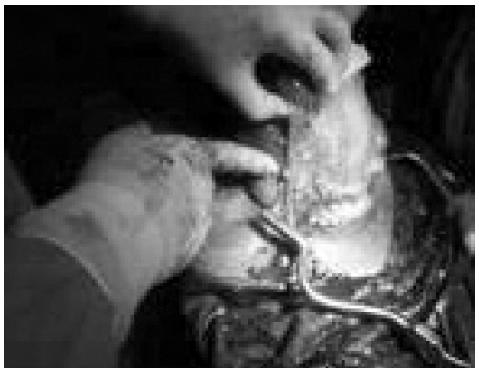


Fig. 2

Fig. 3

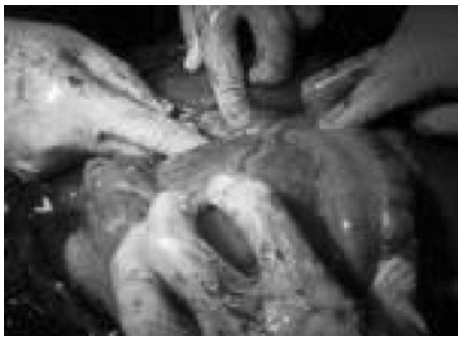


Fig. 4

Figure 1. — Multiple nodules overlying the bladder-uterine peritoneum.
Figure 2. — Right broad ligament dotted with nodules.
Figure 3. — Nodule in the transverse colon.
Figure 4. — Nodules covering the entire uterine surface.

LPD and uterine fibroids have in common in their genesis a hormonal influence and histologically to develop in smooth muscle. Thus the appearance seems to be the same disease. However, this argument is constantly defeated by the following facts: the occurrence of LPD without intrauterine leiomyomas and the presence of uterine fibroids without LPD, particularly in postmenopausal women [2], and male cases of LPD [8], which led to the idea that LPD could be a new disease entity [4]. The rarity of LPD described in the black African population where there is a higher frequency of uterine fibroids compared to Caucasian women [9], is an additional challenge for the connection of etiopathogenic LPD and uterine fibroids.

Moreover LPD can coexist with a number of pathologies such as Currarino syndrome (in groups of caudal regression syndrome) characterized by multiple congenital abnormalities: anorectic malformation, rectovestibular fistula, ectopic right ureteral orifice, megaureter, hemisacrum [10] or pelvic endometriosis [11].

The hypothesis of peritoneal transplant after laparoscopic myomectomy has been raised. Five cases reported in the literature support this hypothesis. Fragments from the morcellation of myoma during laparoscopic surgery left in the peritoneum would be capable of inducing metaplasia of the peritoneal mesenchymal cells and particularly vulnerable women could develop LPD [4, 12]. According to these authors the need for a good peritoneal toilet after laparoscopic myomectomy, removing the peritoneum of any myoma tissue, could aid in prevention of LPD.

We have already mentioned the coexistence of LPD with Currarino syndrome. *Genetic syndromes* such as leiomyomatosis of the esophagus (X-linked Alport syndrome [13] or the genitourinary tract (multiple cutaneous and uterine leiomyomatosis) are known [14]. The familial characteristics of LPD have been mentioned by Halama *et al*. [2]; they describe a case of a white family in which six members were affected by LPD. In addition they noted the association of LPD with Raynaud's syndrome and nodular prurigo with lichenification of the skin.

Diagnosis

Patients generally exhibit nonspecific abdominal symptoms: abdominal pain or discomfort, or rectal pain, and sometimes bleeding (in the case of an associated uterine fibroid). Clinic examination may reveal abdominal masses with increased abdominal girth. A single case of acute abdomen in relation to bowel obstruction has been described in the literature [15]. In practice, patients are usually asymptomatic and the diagnosis is incidental to the occasion of laparotomy (our case).

The contribution of imaging in the diagnosis is not decisive. Computed tomography and magnetic resonance have shown in some cases images of abdominopelvic diffuse masses [15] suggestive of peritoneal carcinomatosis, which is the main differential diagnosis of LPD. Biopsy and histological study of sampling biopsies are

the keystone of diagnosis. Given the abundant vasculature of peritoneal nodules, the risk of severe hemorrhage can not be excluded at the time of biopsy [2]. For these reasons the procedure should be meticulous and include biopsy. Histological examination had shown a fusiform proliferation of smooth muscle cells without mitosis or necrosis, and without atypia or atypical with low-grade [2, 16]. Hyper-vasculature without evidence of vascular invasion may be noted [2]. Electron microscopy can show an abundance of intracellular contractile fibers, a basement membrane surrounding the cells, mitochondria at the nuclear poles and many pinocytic vesicles on the cell surface [17]. The histological features for some authors are at a stage of traditional development between fibroblasts and mature smooth muscle cells [2]. Immunohistochemical studies have shown that tumor cells co-expressed smooth muscle actin, vimentin, desmin, h-caldesmon, and calretinin [16, 18]. The tumor cells also exhibit positivity for progesterone and estrogen receptors. In our case the changes observed were exclusively represented by a proliferation of smooth muscle cells without mitosis, no atypia or necrosis.

Treatment

In terms of therapy, no treatment is accepted by most authors after a review of frozen section samples to determine the histological nature [1, 3]. The risk of malignant transformation is rare. Indeed eight cases of malignancy have been reported in the literature [16]. According to Yamaguchi *et al*. and Sharma *et al*. in about 2-5% of cases, progression to malignancy can be observed [8, 19], justifying the need for a codified monitoring well. According to Goldberg cited by Halama *et al*. [2], the addition of anti-estrogen, removal of exposure to estrogen, or chemical castration by agonists of LH-RH can lead to the regression of tumorous masses. An aromatase inhibitor such as anastrozole was effective for nodules in the study of Takeda *et al*. [6]. Cases of favorable evolution after surgical removal of nodules have been reported and hormone therapy has been proposed to avoid recurrences, however, no definitive therapeutical concept has evolved thus far [2]. In the case we present, we opted for no treatment with clinical and CT scan monitoring.

Conclusion

LPD is a rare disease whose evolution is generally benign, despite the intraoperative findings that frequently lead to peritoneal carcinomatosis. Although benign, the potential risk of degeneration into leiomyosarcoma imposes clinical and CT scan monitoring.

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