A Rare Case of Leiomyomatosis Peritonalis Disseminata

Abstract

Leiomyomatosis Peritonalis Disseminata (LPD), also known as disseminated peritoneal leiomyomatosis is an extremely rare disease in women and men where there is a rapid increase in peritoneal and subperitoneal nodules mainly of smooth muscle cells. Because its diagnosis can mimic or degenerate into peritoneal leiomyosarcoma, it is a benign condition however, diagnosis is tricky. In addition, clinical manifestations can be very nonspecific.

We discuss the case of a 38-year-old single nulliparous lady who presented to the clinic with a 2-year history of lower abdominal swelling as well as a 5-month history of severe menorrhagia. Intraoperative findings showed enlarged uterus with some subserosal fibroids and some deposits on the serosa and intestines. Our patient had Total hysterectomy and bilateral-salpingoophorectomy. Malignant transformation has been reported to almost 10 years from initial diagnosis of the condition so patients should be monitored by repeat ultrasound scans.

Keywords: Leiomyomatosis Peritonalis Disseminata, subserosal fibroids, malignant transformation, estrogen

Introduction

Leiomyomatosis Peritonalis Disseminata, also known as disseminated peritoneal leiomyomatosis is an extremely rare disease in women and men [1, 2, and 6]. Patients with this condition have multiple leiomyomas in the peritoneal cavity which appear malignant but in reality, are benign [3, 4]. Patients are usually in their reproductive age, however there have been reports of this condition after menopause [1, 5]. Cases are discovered incidentally, especially during pelvic surgery or cesarean section or at autopsy [3, 5]. Medical literature also suggests that prolonged estrogen may be responsible and can stem from pregnancy, oral contraceptives or hormone replacement [2, 4]. Based on our search, there were only five documented cases of this condition in Nigeria [1, 2, 3, 4, and 5]. Because many cases are unlikely to be reported, it is possible that the reported incidence for Nigeria is inaccurate.

Case Presentation

This is the case of a 38-year-old single nulliparous lady who presented to the clinic with a 2-year history of lower abdominal swelling as well as a 5-month history of severe menorrhagia. According to her gynecological history, her menses lasted 10 days compared to her previous menses that lasted 5 days. Additional history included weakness, dizziness and shortness of breath on mild exertion.

Positive findings on general examination included- mild respiratory distress [20 cycles/min] and pallor. On examination of her abdomen, there was a 22-week lower abdominal mass described as firm, non-tender, nodular arising from the pelvic region

On Vaginal exam, there was a soaked sanitary pad with vulvar smeared with blood. Speculum exam revealed blood coming from the uterine cavity with a healthy looking ectocervix. Bimanual exam showed a bulky uterus that moved away from the examination finger with no adnexal tenderness.

Investigations showed that hemoglobin was 6.5gm/dl while her electrolytes and urea as well as liver function tests were all normal.

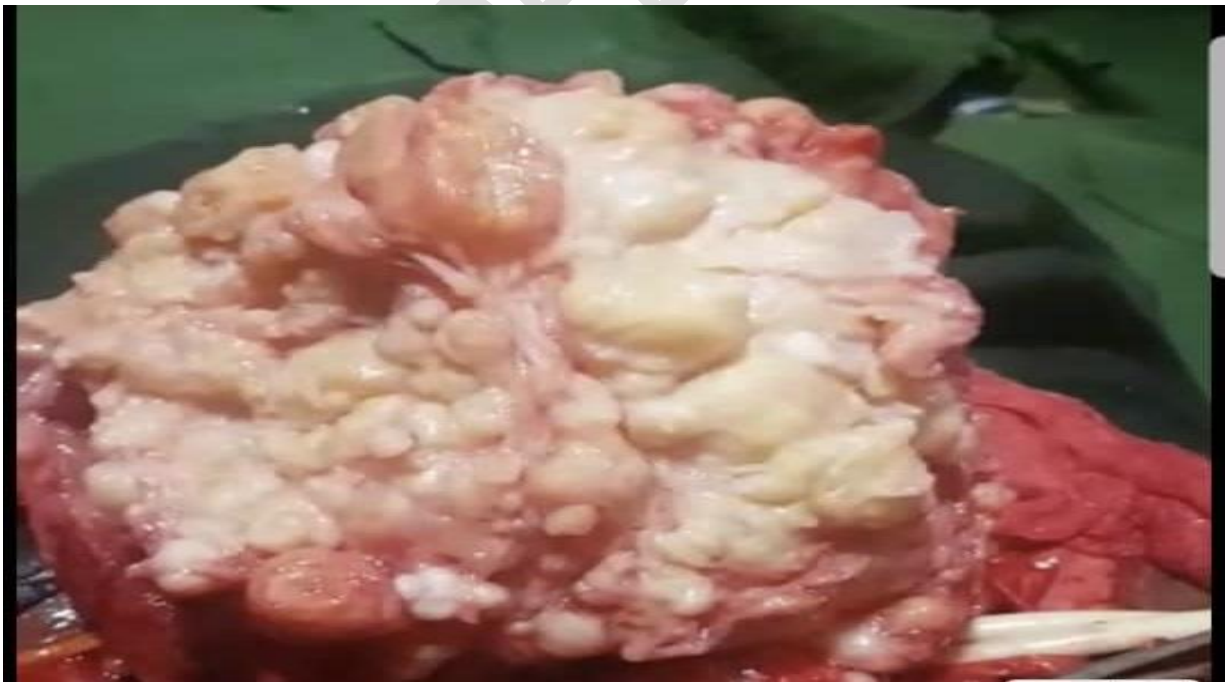
Abdominal ultrasound revealed a very bulky non-gravid uterus with heterogeneous myometrial echo pattern. Endometrial thickness is not appreciated because of multiple myometrial masses. There are multiple masses seen mostly micro-myoma with highest in size measuring 70 x 58cm [post-submucous], 56 x 40cm [ant submucosal, 40 x 34cm [endometrial] & other micro-myometrial masses.

An impression of multiple myometrial masses was made. The patient was counselled on the diagnosis and treatment [myomectomy, possible subtotal hysterectomy]. She was admitted and worked up for surgery. She received 3 units of blood before the surgery and her post transfusion.

Intraoperative findings showed enlarged uterus with some subserosal fibroids and some deposits on the serosa and intestines. No areas of cleavage between the multiple fibroids nodules involving the whole uterine muscles and cavity. In addition, there were beaded fallopian tubes. Total hysterectomy and bilateral-salpingoophorectomy were done.

The uterus and the ovaries were sent for histology which features were suggestive of Leiomyomatosis Peritonalis Disseminata

Fig 1. Morphology of Leiomyomatosis Peritonalis Disseminata



Discussion-

This uncommonly documented condition is known to be common in reproductive aged women. It was first reported by Wilson and Peale as multiple peritoneal leiomyomas and the term leiomyomatosis peritonealis disseminata (LPD) was established in 1965 [6, 7 and 8].

The medical literature suggests that exposure to female sex hormones especially for women who are premenopausal and so estrogen and progesterone receptors are implicated. On the other hand, another school of thought suggests that iatrogenic etiology is responsible especially after laparoscopic myomectomy with morcellation especially considering that the procedure might result in myoma tissues that are residual. This school of thought cannot explain our patient's condition. At the time of writing this report, only five cases in Nigeria were identified in the medical literature [1-5]. While this condition can mimic a malignant condition, it is a benign disorder where the patients have numerous myomas in the peritoneal cavity [2-4]. Some patients may have a coexisting fibroid. Our patient was in her reproductive age bracket; this condition is not common in women who have become menopausal. No cause has been identified in the literature. Medical literature implicates long term exposure to estrogen plus or minus progesterone. Pregnancy, hormone replacement therapy and oral contraceptive pills belong to this category of exposure to estrogen [3-5, 10]. Many patients do not have symptoms, however, for those who have symptoms, they present with menorrhagia. Others have abdominal pain, abdominal swelling and/demonstrable abdominal masses [1, 2, 9]. Investigating this condition includes imaging such as ultrasound scan or CT scan. Definitive diagnosis is made by histology. Treatment includes surgery or medical and is dependent on hormones, reproductive status, age and the symptoms. Our patient had Total hysterectomy and bilateral-salpingoophorectomy. Malignant transformation has been reported to almost 10 years from initial diagnosis of the condition so patients should be monitored by repeat ultrasound scans [2].

Conclusion

LPD is an uncommon and benign disease which can be confused for malignancy in the peritoneum. In a patient exposed to estrogen and progesterone or a patient with a history of hysterectomy who presents with abdominal mass, this condition should be suspected. Even though malignant transformation has been rarely reported, it is a possibility. Patients with malignant transformation will possibly include: postmenopausal patients, no history of myomectomy with morcellation and male patients. Differentials for this condition include: disseminated leiomyosarcoma, peritoneal carcinomatosis and tumors of the gastrointestinal system. Treatment depends on different factors

Consent

As per international standard or university standard, patient(s) written consent has been collected and preserved by the author(s).

Ethical Approval:

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

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