Case Reports

A case of disseminated peritoneal leiomyomatosis and diffuse uterine leiomyomatosis

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Summary

A rare case of simultaneous uncommon pathologies in the same patient is described: diffuse leiomyomatosis and disseminated peritoneal leiomyomatosis (DPL). The evolution and monitoring of this rare clinical case together with diagnostic and therapeutic procedures are presented.

Key words: Peritoneal leiomyomatosis; Diffuse uterine leiomyomatosis.

Introduction

Disseminated peritoneal leiomyomatosis (DPL) is a rare condition characterized by numerous peritoneal nodules consisting of proliferations of collagen, fibroblasts, and smooth muscle myofibroblasts, and in pregnancy and puerperium, decidual cells. This disorder usually has a benign course and in the literature have been reported only three cases of malignant degeneration [1]. The production of female hormone estrogen plays a major role in the pathogenesis of this disease but there have also been reported cases in menopausal women. Consequently pregnancy, use of oral contraceptives, endometriosis, caudal regression syndrome (Currarino syndrome), uterine myoma, ovary and secretive fibrothecoma have also recently been described: a case report in the course of a Brenner tumor and during therapy with tamoxifen. The diagnosis is made by biopsy. Surgical ablation or therapy with GnRH agonists appear to be valid solutions in case of progression or recurrence of the disease [2-4]. Leiomyoma is described as a neoplasm with different variations according to type of growth [5].

Diffuse leiomyomatosis with a large increase in the uterus consisting of many nodules of various sizes;

– dissecting leiomyoma mimicking an invasive malignant tumor;

– diffuse benign leiomyoma which can cause, metastasis after several years;

– intravenous leiomyomatosis characterized by the development of a benign smooth muscle tumor in the venous system.

The etiology of diffuse leiomyomatosis as is that of DPL is unknown. Some studies confirm hormonal influence since receptors for luteinizing hormone (LH) in nodules of postmenopausal women have been found and it is believed that the etiopathogenesis of this condition depends on increased hormonal levels of LH rather than on stimulation of estrogen. The hormonal influence is considered likely [6]. Moreover, many authors consider pregnancy and exposure to prolonged estro-progesterone therapy as other risk factors [7].

The nodules are random and can reach an inch in diameter, whereas in metastatic leiomyosarcoma they tend to be wider and to invade adjacent tissues. Diffuse leiomyomatosis may be symptomatic or not.

The diagnosis of DPL occasionally occurs during surgery: cesarean section, laparotomy or laparoscopy [8]. Other patients have non specific symptoms: irregular cycles, bleeding, dyspareunia, pollakiuria, and some secondary shown urosepsis and acute abdomen for ovarian torsion. The differential diagnosis of diffuse leiomyomatosis appears to be more difficult with ultrasound (US) than with computed tomography (CT) because it usually presents as a solid mass: for DPL magnetic resonance imaging (MRI) shows masses similar to the uterine parenchyma localized in the pelvis adjacent to the iliac vessels. The final diagnosis is pathological. Therapy should be individualized according to the age of the patient together with the reproductive and hormonal symptoms. There have been reports of favorable reproductive outcomes in women with diffuse uterine leiomyomatosis after hysteroscopic myomectomy with removal of only myoma that invaded the endometrial cavity and leaving the intramural myoma. The uterus was successfully preserved and the normal intensity of menstrual flow was restored [9].

Materials and Methods

A 42-year-old female was admitted to the Division of Obstetrics and Gynecology, with benign leiomyomatosis of the uterus, diffuse leiomyomatosis and associated DPL. There were no problems during clinical preoperative diagnosis for non specific symptoms, and during surgery there were primarily problems of differential diagnoses of the malignant neoplasms. The patient smoked over 20 cigarettes/day and took oral contraceptives for less than five years. She presented symptoms characterized predominantly by slight recurrent metrorragia and abdominal fullness. The patient had undergone surgery for an extrauterine pregnancy and her obstetrical history included a spontaneous birth. Consequently she underwent colposcopy

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Fig. 1

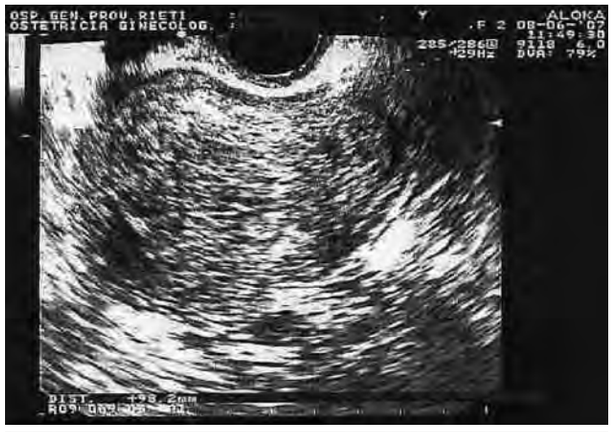

Fig. 2

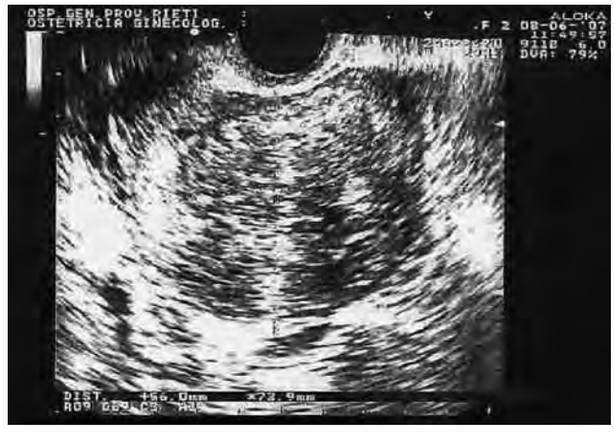

Figure 1. — Longitudinal uterine scans.
Figure 3. — Myoma uterine.

Fig. 3

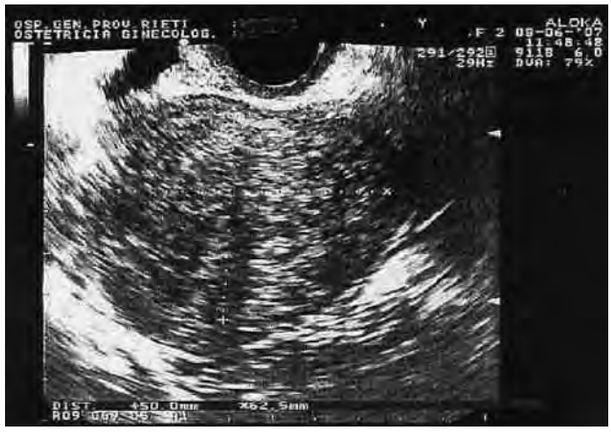

Fig. 4

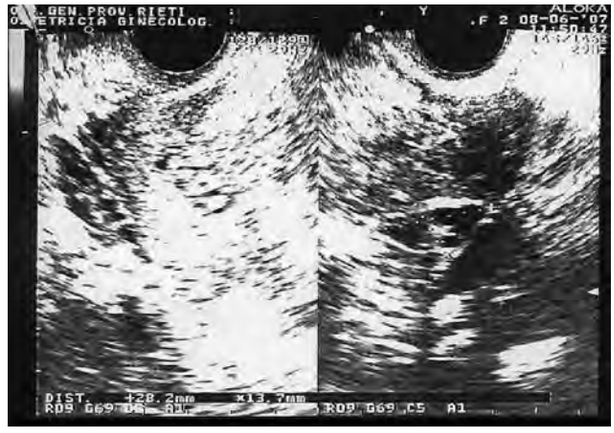

Figure 2. — Trasverse uterine scans.
Figure 4. — Adnexal area.

that revealed a regular mosaic pattern. The pathological endometrial examination revealed endometrium with cytogenetic edematous stroma and small glands in the proliferative phase. There were no urinary symptoms and a pregnancy test was negative. The remaining standard blood tests were within normal range.

Transvaginal ultrasound showed:

– significantly increased uterus size (98 x 56 x 74 mm) (Figures 1 and 2);

– myoma of the uterus (50 x 62 mm) (Figure 3);

– normal adnexal structures (Figure 4).

Because of menstrual irregularities and associated symptoms, surgery was scheduled after discussing the options with the patient. The patient was admitted to our Department and underwent laparohysterectomy with bilateral salpingo-oophorectomy. The uterus was increased in volume, with a nodular appearance on the surface; a cauliflower mass was widespread into the vascularised posterior uterine wall and to the left large ligament. A neoplastic nature was suspected on the basis of the macroscopic intraoperative appearance. The differential diagnosis was performed after laparohysterectomy performed with the typical technique after lysis of the adhesions and removal of the mass from the uterus. Frozen biopsy of the mass was performed and subsequently the mass was removed. The macroscopic examination showed multiple diffused nodules ranging from 0.4 to 1 cm. After careful separation from the bowels the uterine mesenchymal tumor was removed with the left ovary included.

Results

The patient had a good general and the postoperative course was normal. She was discharged four days after surgery.

Histopathology

1) connective tissue, sometimes with mesothelial coating, discretely vascularised, with multiple nodular formations of variable size (from a few millimeters to about discretely one centimeter), consisting of fusocellular elements with epithelial surface characterized by small cytologic atypia, without necrosis and kariokinetic activity (2 x 50 HPF mitosis), there were also signs of endosalpingiosis (Figures 5 a, b, c, d). The findings of a rare condition known as peritoneal leiomyomatosis required follow-up of the patient.

2) Ovary with corpus albicans, follicular and epithelial cysts, caused by coelomic epithelial inclusions, with the fallopian tube free.

3) Ovary with hemorrhagic corpus luteum and follicular cystic.

Leiomyomatosis consisting of diffuse proliferation of small nodules range in diameter from a few millimeters to a centimeter, made up of fusocellular elements of smooth muscle, with small cytologic atypia, without necrosis and

Fig. 5a

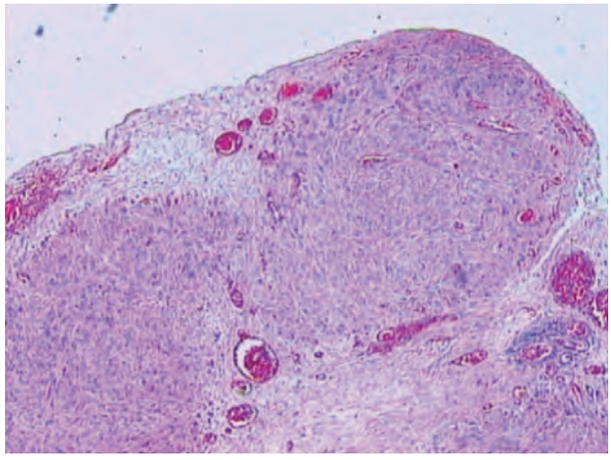

Fig. 5b

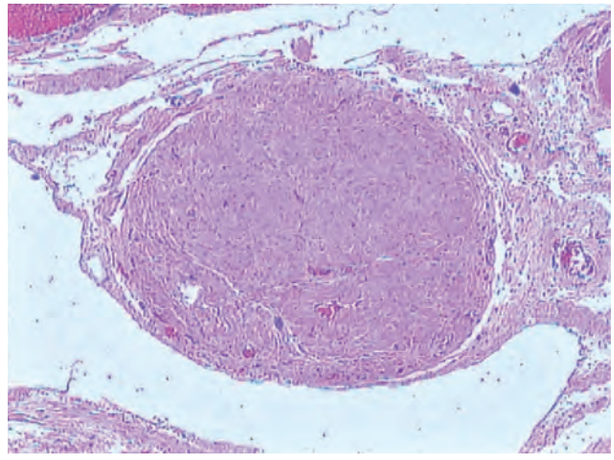

Fig. 5c

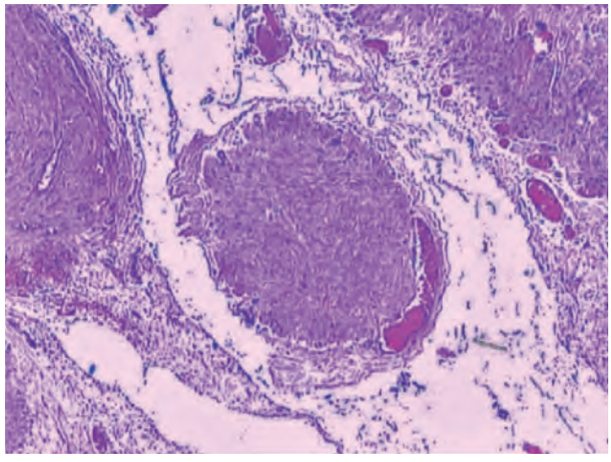

Figure 5. — **a)** Low power photomicrograph illustrates multiple small nodules of smooth muscle cell (H&E - original magnification 4x). **b)** and **c)** Nodules of spindle-shaped smooth muscle cells in the peritoneum (H&E - original magnification 4x).

kariokinetic activities (1/50 mitoses), and myometrum with adenomiosis. There was chronic cervicitis squamous superficial and immature metaplasia and proliferative endometrium.

One month later the patient was checked by CAT scan of the complete abdomen and chest and was disease free.

Discussion

This rare case of contemporaneous diseases leads us to consider the correct role and modality of the intraoperative diagnosis for the best management of the diseases. The peritoneal leimyomatosis was asymptomatic and associated with diffuse uterine myomatosis. The diagnosis of DPL isoften incidental; therefore once the lesion is detected it differential diagnosis of an DPL with other diseases is important because the risk is high. Only the biopsy and pathological examination give us a precise and proper surgical approach. The laparotomy approach is indicated when the leiomyomatosis is infiltrating and diffuse.

Conclusion

On the basis of the literature results on this pathology, clinical history associated gynecological pathology and good diagnostic assessment with ultrasound, CT and MRI permit a precise diagnosis, with the identification of early lesions in a disease that can remain asymptomatic for a long period. Although there is no standardized treatment and no set guidelines regarding the management of the DPL, it is believed that therapy should be personalized. Different therapies including GnRH agonists, danazol and the megestrol acetate have been used but when there are symptoms of bleeding and the effects of compression on the bladder and on the intestine, surgery treatment choice is the best. We believe that total hysterectomy with bilateral salpingo-oophorectomy and removal of the peritoneal neoplasm was the only appropriate treatment for this case.

Surgical therapy allowed radical excision of the masses and adhesiolisis with great benefit for our patient.

Removal of the lesion must be complete with a single

surgical operation even in cases of multifocal spread to avoid the risk of recurrence.

Biopsies need to be multiple and enough for a complete diagnosis. Last but not least, a critical analysis of the management is needed, to be sure that the best choice for the patient has been made with a correct and complete differential diagnosis of the disease.

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