DISSEMINATED PERITONEAL LEIOMYOMATOSIS

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Disseminated peritoneal leiomyomatosis (DPL) is a very rare condition that almost always occurs in young women subjected to an altered hormonal milieu, usually pregnancy. To the surgeon, DPL may appear as metastases; the possibility of DPL should be considered whenever peritoneal nodules are encountered in a young woman, particularly if she is pregnant. Here we present the case report of a 35-year-old woman with DPL, spontaneously regressing after total hysterectomy and bilateral salpingo-oophorectomy.

Keywords • disseminated peritoneal leiomyomatosis • leiomyoma

Introduction

Disseminated peritoneal leiomyomatosis (DPL) involves the pelvic and abdominal peritoneum as well as the omentum, and is characterized by the presence of numerous nodules scattered over the peritoneal surface.1

The notable point with this condition, which occurs in association with pregnancy in 70% of cases, is that its macroscopic appearance can be mistaken for peritoneal carcinomatosis.2 Postpartum spontaneous regression is the rule.3

The condition is almost always discovered incidentally and is usually asymptomatic. The disease has recently been reported in a postmenopausal woman not on hormone therapy.4 Recently, clonality and cytogenetic analyses have suggested that the pathogenesis of diffuse peritoneal leiomyomatosis is similar to that of leiomyomas but failed to differentiate between a metastatic or multifocal in situ origin for the disease.5

Case Report

A 35-year-old woman was admitted to the Gynecology Clinic of Mirza Koochak Khan Hospital, because of dysfunctional uterine bleeding for six months without desirable response to prior medroxyprogesterone.

A dilatation and curettage was performed and the tissue sample was sent to a pathologist who reported proliferative endometrium and uterine leiomyoma. The responsible gynecologist prepared the patient for total abdominal hysterectomy. In operating theater the gynecologist faced multiple small nodules of various sizes all over the peritoneal surface, over the uterus, and both adnexa without any adhesions or ascites.

A nodule was taken for histologic evaluation without further interventions. Microscopic examination showed that the nodule was composed of bland spindle cells with cigar-shaped nuclei in interlacing bundles.

The diagnosis was leiomyomas and considering the patient’s history and physical exam, disseminated peritoneal leiomyomatosis was suggested.

During the second operation, abdominal hysterectomy, bilateral salpingo-oophorectomy, and biopsy of a few peritoneal nodules (leaving behind the myriad of other nodules) were performed. Microscopically, multiple leiomyomas of ovaries (Figures 1 and 2), uterus and peritoneal nodules were apparent (Figure 3).

Immunohistochemical staining showed positivity for actin (Figure 4), myosin, and desmin. After ten months, a follow-up laparoscopy surprisingly showed smooth peritoneal surface without any nodules.
Discussion

DPL is a rare and typically self-limiting tumor-like condition of the peritoneal subcoelomic mesenchyme manifesting as myriad nodules of smooth muscle cells, up to 2 cm in diameter, mimicking metastatic carcinoma.

So far, DPL has been described in 48 cases. It is associated with multiple small nodules of mature smooth muscle, distributed throughout the omentum and both visceral and parietal layers of the peritoneum.

It should be distinguished from well-differentiated leiomyosarcoma. Its malignant transformation is very rare, and has been reported in literature in only 3 cases. Steroid hormone receptors have been detected in the proliferating cells of DPL; in addition, it has been reported to be associated with estrogen-secreting ovarian fibrothecoma. One-half of DPL patients are pregnant and many others are taking oral contraceptive agents at the time of presentation. It affects the ovaries in one-quarter of cases. In most instances, spontaneous regression of the nodules occur.

The notable point with our case was her presenting with almost all features, described in literature, for DPL including multiple peritoneal nodules mimicking metastatic carcinoma, consumption of a progesterone agent, uterine and ovarian leiomyomata, and the spontaneous regression of all nodules postoperatively.

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References