Simultaneously occurring disseminated peritoneal leiomyomatosis and multiple extrauterine adenomyomas following hysterectomy

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ABSTRACT

Disseminated peritoneal leiomyomatosis (DPL), also known as leiomyomatosis peritonealis disseminata, is a rare condition characterized by multiple benign smooth muscle tumors proliferating along the peritoneal surfaces. In previous case reports, these tumors have been noted to involve the ovaries, round ligaments, bladder, bowel, peritoneum, and mesentery. To date, approximately 150 cases of DPL have been described in the literature. Extrauterine adenomyoma is an even rarer entity, involving benign tumors composed of smooth muscle tissue, endometrial glands, and endometrial stroma arising outside the uterus. Only 22 cases have previously been reported. We describe a woman presenting with both DPL and multiple extrauterine adenomyomas several years after undergoing laparoscopic morcellated hysterectomy.

KEYWORDS Disseminated peritoneal leiomyomatosis; extrauterine adenomyoma; laparoscopic morcellated hysterectomy; leiomyomatosis peritonealis disseminata

Leiomyomas are benign tumors composed of proliferating smooth muscle cells. They are the most common uterine tumors in women of reproductive age, with prevalence estimated as high as 77%.¹ Extrauterine leiomyomas, on the contrary, are quite rare. When multiple discrete extrauterine leiomyomas are found to be distributed along the peritoneal surfaces, the entity is referred to as disseminated peritoneal leiomyomatosis (DPL). Adenomyomas are benign tumors composed of smooth muscles cells, endometrial glands, and endometrial stroma. Extrauterine adenomyomas are even rarer than extrauterine leiomyomas; only 22 cases have previously been reported.² The finding of both types of tumors simultaneously is exceedingly rare and to our knowledge has not previously been reported in the literature. In this case study, we focus on these two rare conditions, briefly review the literature, discuss differential diagnoses, review a few leading theories behind the pathophysiology, and explore treatment options.

CASE DESCRIPTION

A 50-year-old woman presented with complaints of progressive abdominal bloating and indigestion. She had recently noted a palpable left lower-quadrant mass. Her cancer antigen 125 level was mildly elevated. Six years earlier (at age 44), she had had a robotic laparoscopic hysterectomy for large uterine fibroids using the morcellation technique, without utilizing a bag. Pelvic sonogram revealed multiple large pelvic masses. A contrast-enhanced computed tomography (CT) scan demonstrated multiple lobulated heterogeneously enhancing masses filling most of the pelvis and compressing the bladder (*Figures 1a–1c*). Several additional peritoneal masses were present in the bilateral lower quadrants adhering to the bowel. There was a small volume of ascites. Moderate right hydronephrosis secondary to ureteral obstruction by the pelvic masses was also noted (*Figure 1d*). A percutaneous imaging-guided biopsy revealed benign leiomyoma. Because of persistent concern for malignancy, exploratory laparotomy was performed with bilateral

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Figure 1. (a) Axial, (b) coronal, and (c) sagittal contrast-enhanced CT images demonstrate multiple heterogeneous masses filling the pelvis extending into the bilateral lower quadrants of the abdomen and compressing the bladder. The uterus is absent. (d) Coronal image of the abdomen and pelvis shows hydronephrosis of the right kidney caused by mass effect on the distal ureter. A heterogeneous mass can be partially visualized in the pelvis.

Figure 2. Hematoxylin and eosin stains of (a) a leiomyoma (low power), (b) an adenomyoma (low power), and (c) an adenomyoma (high power). The high-power view clearly demonstrates internal endometrial glands and stroma, differentiating the tumor from a leiomyoma.

salpingo-oophorectomy, segmental sigmoid colectomy, and tumor debulking. At the time of laparotomy, multiple masses were found to be adherent to the left ovary, right round ligament, vaginal cuff, bladder, small bowel, and rectosigmoid colon.

Histopathology showed nine discrete masses, six consistent with leiomyomas (*Figure 2a*) and three with adenomyomas (*Figures 2b and 2c*). The leiomyomas were found adherent to the left ovary, right round ligament, bladder (two separate masses), rectosigmoid colon, and small bowel. The adenomyomas were found adherent to the rectosigmoid colon, vaginal cuff, and descending colon. The masses ranged in size from 6 mm to 14.5 cm, in total weighing 705 g. All specimens were negative for malignancy. The patient was discharged home 5 days after surgery in good condition. Her indigestion and abdominal bloating resolved after surgery, and she has experienced no further symptoms in the 6 months since her surgery.

DISCUSSION

Though uterine leiomyomas and adenomyomas are relatively common benign tumors, the appearance of these tumors in extrauterine locations is exceedingly rare.

The pathogenesis behind extrauterine proliferation of leiomyomas and adenomyomas is not well understood, in part because of the rarity of the conditions. The most widely reported theory is that of parasitic spread of the tumors after seeding during hysterectomy or myomectomy.³ In particular, multiple cases of DPL have been reported following morcellated hysterectomy/myomectomy—a laparoscopic procedure in which the myometrial tissue is divided into multiple small fragments in situ using a power morcellator prior to extraction.^{3–5} This is due to the fact that morcellation carries the risk of leaving small fragments of fibroid or even unsuspected malignant tumor behind, thereby seeding the pelvis and laying the groundwork for tumor proliferation. In April 2014, the US Food and Drug Administration recommended that manufacturers of laparoscopic power morcellators include a warning label on the devices to inform patients and surgeons of the risk of potential abdominopelvic seeding of occult uterine cancer with morcellation procedures.^{6,7} Of note, however, not all patients with extrauterine leiomyomas or adenomyomas have a history of their intrauterine counterparts. In our patient, there was a history of uterine fibroids but not of adenomyoma or adenomyosis.

Another theory suggests that there may be an association of the tumors with estrogen and progesterone levels. This is

based on multiple case reports noting the occurrence of these tumors almost exclusively in premenopausal women, as well as an increase in size and number of the masses during pregnancy, with contraceptive use, and in association with granulosa cell tumors of the ovary.^{3,8,9}

Typically, patients are asymptomatic and the masses are found incidentally on imaging or during surgery, such as cesarean section. However, as in our case, several patients have presented with symptoms including abdominal pain, gastrointestinal symptoms, and urinary obstruction. Due to their striking appearance on imaging, these tumors are often initially mistaken for disseminated malignancy. Metastatic leiomyosarcoma and peritoneal carcinomatosis are the main differential considerations. On imaging (with CT or magnetic resonance imaging being the modalities of choice), the lack of omental fatty infiltration of tumor, significant ascites, or solid organ metastases can help differentiate DPL and extrauterine adenomyomas from malignant conditions; however, histopathologic examination is required for definitive diagnosis.

Treatment methods include both pharmacological and surgical approaches. A few cases of complete regression of DPL have been described with bilateral oophorectomy, essentially removing the body's primary source of estrogen.³ For symptomatic cases, as in our patient, laparotomy and tumor debulking usually provide excellent symptomatic relief. Though surgery is the mainstay of therapy, gonadotropin-releasing hormone agonists and aromatase inhibitors were shown in a case study of five patients with DPL to be beneficial in inoperable cases or cases in which the patient wanted to avoid the risks of surgery.¹⁰ In addition, a case report of two patients with extrauterine adenomyomas showed benefit of long-term treatment (4–10 years) with gonadotropin-releasing hormone agonists following surgical resection.¹¹

Of note, tumor recurrence following surgery is not uncommon with DPL.¹⁰ In addition, development of leiomyosarcoma in the setting of DPL has been reported.¹⁰ It has thus been recommended that patients be followed periodically with imaging for several years after diagnosis of DPL, although there is no clear consensus on the precise timeline for follow-up.

In conclusion, both extrauterine leiomyomas and adenomyomas are exceedingly rare entities, and their pathogenesis is not well understood. They are often difficult to diagnose, because their imaging appearance mimics that of leiomyosarcoma metastases or peritoneal carcinomatosis, which are far more common conditions. Histopathologic examination

is required for definitive diagnosis. The simultaneous presentation of disseminated extrauterine leiomyomas and adenomyomas, as presented in this case study, to our knowledge has not been previously reported.

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