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"Disseminated Peritoneal Leiomyomatosis" An Imitator of Disseminated Intra-Abdominal Malignancy

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Conflicts of Interest: Nil

Abstract

Disseminated peritoneal leiomyomatosis (DPL) is a rare condition characterized by the presence of multiple nodules of smooth muscle, fibroblasts and myofibroblasts on the visceral and/or parietal peritoneal surfaces of the abdominal and pelvic cavities in women of reproductive age. [1] Lesion may not be present in uterus at the time of diagnosis, as in our case. It usually causes a great dilemma for clinician, radiologist and pathologist due to the rarity of this condition as well as its resemblance to disseminated intra-abdominal malignancy and no test is available for its preoperative diagnosis. [2] We report a case of DPL in a 37 year old nulliparous woman, in whom correlation of preoperative and pathological findings together with clinical history, made the diagnosis possible and it was confirmed by immunohistochemistry.

Keywords

Disseminated peritoneal leiomyomatosis, Peritoneum, Morcellation, Myoma, Subperitoneal mesenchymal stem cell.

Introduction

Disseminated peritoneal leiomyomatosis (DPL) is a condition characterized by the presence of multiple nodules of smooth muscle, fibroblasts and myofibroblasts on the visceral and/ or parietal peritoneal surfaces of the abdominal and pelvic cavities in women of reproductive age. We report a case of DPL due to its rarity and diagnostic challenge offered by it; only less than 150 cases have been reported in the literature till date.

Case history

Thirty seven year old nulliparous woman reported to the gynecology outpatient clinic with complaints of left lower abdominal pain of two months duration. She had past history of laproscopic surgery for multiple subserous leiomyoma of uterus twice at the age of 32 and 34 years. There was no history of exposure to OCP and her menustrual cycle was regular. On examination clinician detected a mass in her left iliac fossa and diagnostic workup showed elevated serum CA 125 value (66.1 U/mL). CECT showed multiple heterogeneously enhancing lesions with central hypo-dense areas in left iliac fossa and omentum, while uterus was free of any lesion. Patient underwent staging laprotomy with a provisional diagnosis of left ovarian malignancy with omental metastasis.

Preoperatively multiple discrete nodular lesions were observed on the peritoneal surface of descending colon, omentum and pelvis. Both ovaries and uterus were normal and there was no ascites. Surgeon removed the affected segment of colon along with infracolic omentum and considered the possibility of disseminated malignancy of colon. On gross examination, peritoneal surface of colon

showed four nodular lesions, largest measured 8 x 5 x 5 cm and smallest measured 1 x1x 1cm, but its mucosal surface was normal. Seven nodular lesions were seen in the omentum also, largest measured 2 x 2 x 2 cm. All nodular lesions were circumscribed, grey white, firm masses, with variable degree of congestion [Figure 1] and showed similar histopathological appearance. All were circumscribed neoplasms within the peritoneal tissue and were composed of spindle cells arranged in interlacing bundles and diffusely. These cells had moderate amount of eosinophilic cytoplasm and spindly, plump, vesicular nucleus with sparse mitotic figures. Stroma showed mast cells, lymphocytes, congestion and proliferating capillaries, but no tumor necrosis [Figure 2 and 3]. On correlating with the past history of recurrent multiple uterine subserous fibroids, we suggested the diagnosis of DPL in this patient which was confirmed by immunohistochemistry. Spindle cells expressed SMA, and ER [Figure 4 and 5] while negative for CD117. Ki 67 value was 1%.

Discussion

Since the original description of DPL by Wilson and Peale in 1952, only less than 150 cases have been reported in the literature till date. [3, 4] Majority of these were incidental findings during caesarian section or laprotomy in women of child bearing age. Usually DPL lesions are distributed randomly on the peritoneal surface of uterus, ovary, intestine, omentum, mesentry, spleen and pancreas, but without invasion into the substance of adjacent structures, ascites or omental adhesions. [5, 6] In spite of the multifocal involvement, cells in DPL nodules show low mitotic rate, little nuclear atypia and lack coagulative necrosis. Nucleus of these cells expresses estrogen and progesterone receptors. [7] Studies have suggested that multicentric subperitoneal mesenchymal stem cell metaplasia may be the possible mechanism of pathogenesis of DPL and hormonal, genetic as well as iatrogenic factors like morcellation of myoma play a role in it. [8]

Key Message

In DPL, lesion may not be present in uterus at the time of diagnosis, as in our case. Correlation of clinical history with pathological and preoperative findings will be helpful in diagnostically challenging diseases like DPL and confirmation can be done by ancillary methods like immunohistochemistry.

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Figure 1. Gross: Multiple discrete circumscribed grey white nodules with variable degree of congestion on the peritoneal surface of colon and omentum.

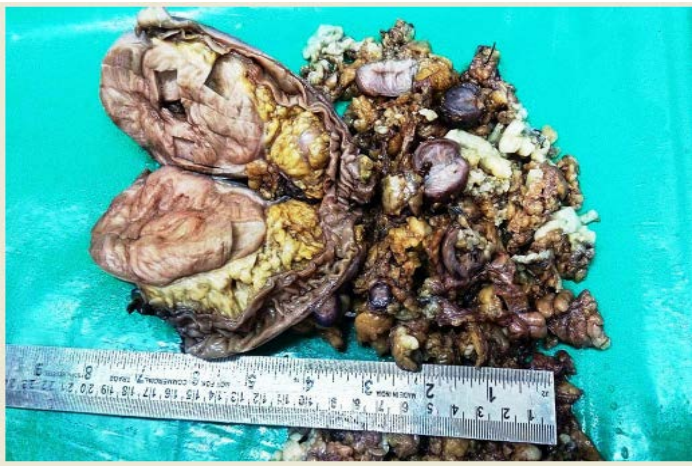

Figure 2. Microscopy: Neoplasms composed of spindle cells arranged in interlacing bundles [H & E stain x10].

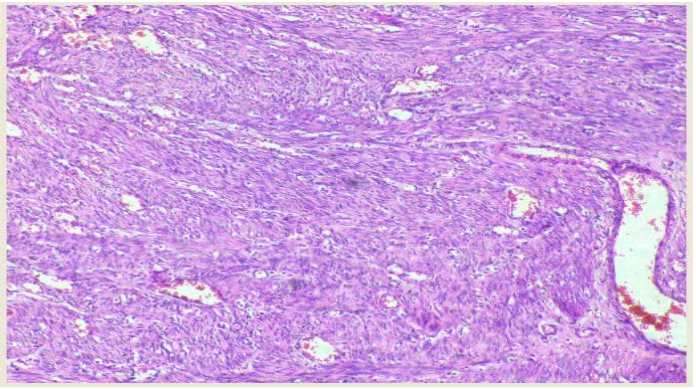

Figure 3. Microscopy: Spindle cells with moderate amount of eosinophilic cytoplasm, elongated plump vesicular nucleus and sparse mitotic figures [H & E stain x 40].

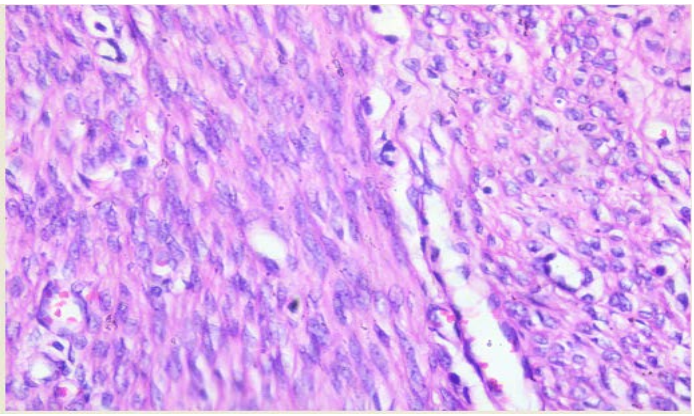

Figure 4. Microscopy: Spindle cells with positive cytoplasmic staining for smooth muscle actin [immunohistochemical staining x 20].

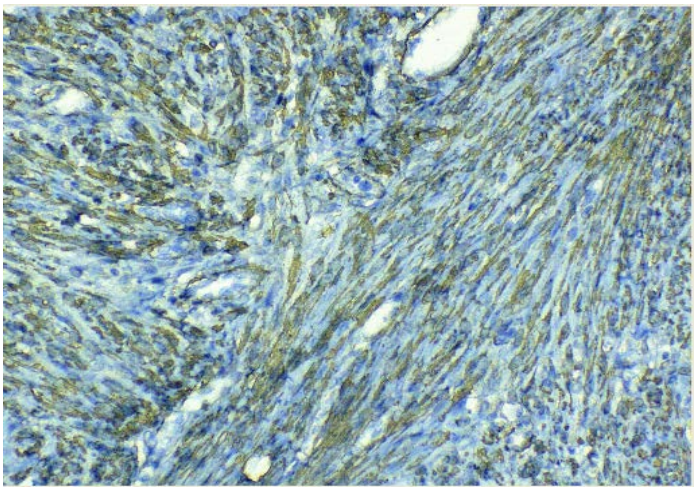

Figure 5. Microscopy: Spindle cells with positive nuclear staining for estrogen receptor [immunohistochemical staining x 20].