CASE REPORT

Recurrent Diffuse Peritoneal Leiomyomatosis Following Leiomyomata Uterus

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Introduction

Leiomyomatosis peritonealis disseminata (LPD) is a rare disease in which multiple smooth muscle or smooth muscle-like nodules develop subperitoneally in any part of the abdominal cavity. These nodules, though histologically benign, cannot be distinguished macroscopically from peritoneal carcinomatosis and gastrointestinal stromal tumors. Only about 140 documented cases were found in the English language literature [1]. However, there are occasional case reports of recurrent LPD. We present here the unique case of LPD in a female who presented over a period of 16 years with benign clinical course and morphology.

Case Report

A 40-year-old female presented to the gynecology OPD in the year 1997 with history of pain in the abdomen and

menorrhagia for the past 6 months. Ultrasound of the pelvis showed multiple intrauterine echogenic lesions suggestive of fibroid uterus. The patient underwent hysterectomy. Histopathologic examination of the resected specimen revealed leiomyomata uterus (Fig. 1). Five years later, in the year 2002, the patient presented again with history of pain in the abdomen with sudden onset, vomiting, diarrhea, and inability to pass flatus. CT scan of the abdomen showed a well-defined, solid lesion measuring $12.3 \times 5.6 \times 7.6 \text{ cm}^3$ seen in the pelvis extending below the lower pole of the kidney. A single mass of enhancement was seen over the ileum measuring $1 \times 1 \text{ cm}^2$. Multiple sections from the adnexal mass and peritoneal nodule over the ileum were diagnosed as benign peritoneal leiomyomatosis. No evidence of mitosis or malignancy was seen in the multiple sections studied. The tumor nodules showed positivity for masson trichrome. Immunohistochemistry for desmin, vimentin, ER, and PR was positive in both the adnexal mass and the peritoneal nodule, thereby confirming the diagnosis. Ten years later, in the year 2012, the patient presented again with history of colicky abdominal pain and altered bowel habits. CECT of the abdomen revealed a $6 \times 3 \text{ cm}^2$ lesion adherent to the small bowel and ascending colon and numerous peritoneal small nodules. Right hemicolectomy with lymph node dissection was performed. Clinical differential diagnosis of gastrointestinal stromal tumor and LPD was made. Gross examination of the resected specimen revealed a $4 \times 2.5 \times 1.5 \text{ cm}^3$ gray white mass attached to the serosal aspect of the ileum. Another small nodule measuring

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Fig. 1 H&E stain showing proliferating spindle cells arranged in whorls suggestive of leiomyoma (H&E $\times 400$)

$0.5 \times 0.3 \times 0.2 \text{ cm}^3$ was present on the ascending colon. The wall of the ileum and colon was intact. Microscopically, the tumor consisted of proliferating spindle cells arranged in whorls. No areas of cellular atypia or mitosis were seen. No areas of hemorrhage and necrosis were found within the tumor nodules. An immunohistochemistry panel for desmin, vimentin, ER and PR, and c-kit was put up and nodules showed positivity for desmin (Fig. 2), vimentin, ER, and PR and negativity for c-kit. A final diagnosis of Leiomyomatosis peritonealis disseminata (LPD) was given.

Discussion

LPD is a rare disease and only a relatively small number of cases have been reported so far. Most of the cases are in females, as in our case. Most patients with LPD present without specific symptoms and many documented cases of LPD have been discovered incidentally during surgery (cesarean section, laparotomy, or laparoscopy). The etiology of LPD is unknown, but it is thought to originate from metaplasia of submesothelial, multipotent mesenchymal cells. The developing leiomyomatous nodules probably arise from Muller's epithelium, which is distributed throughout the subperitoneal mesenchyme. In cases of individual predisposition and hormonal stimulation, müllerian derivatives proliferate along lines of myofibrous differentiation [2].

Diagnosis may be confused with peritoneal carcinomatosis and gastrointestinal stromal tumor. Peritoneal carcinomatosis,

Fig. 2 Immunohistochemistry showing positivity for desmin (IHC $\times 400$)

however, is often associated with omental cake, ascites, and liver metastases, which have not been reported with LPD. It is important to differentiate diffuse peritoneal leiomyomatosis from gastrointestinal stromal tumor since management in both the cases is different. Gastrointestinal stromal tumor presents with CD117-positive cells; hence, CD117 should be included in the immunohistochemistry panel for LPD along with vimentin, SMA, ER, and PR.

There are no established guidelines regarding the management of LPD. However, therapy needs to be individualized according to the patient's age, hormonal and reproductive status, and symptomatology. Different drugs (gonadotropin-releasing hormone agonist, megestrol acetate, danazol) have been considered in some cases, but with poor results. If intestinal and bladder mass effect symptoms are prominent, a surgical approach is indicated. The management for malignant LPD is more aggressive. Hence, we advise close follow-up of patients with LPD to detect early sarcomatous change.

References

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