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CASE REPORT

Disseminated Peritoneal Leiomyomatosis Mimicking Ovarian Torsion

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The presentation of disseminated peritoneal leiomyomatosis (DPL) can be misleading. Herein, we present the case of a 42-year-old nulliparous female who had previously undergone a total hysterectomy and presented with an acute abdomen. A presumptive diagnosis of ovarian torsion was made based on the clinical findings and an ultrasonographic examination. A diagnostic laparoscopy was performed immediately. DPL was subsequently diagnosed based on an intra-operative frozen section during surgical exploration and the final histopathologic examination. This case illustrates an atypical presentation of DPL mimicking ovarian torsion.

Key words: Disseminated peritoneal leiomyomatosis, ovarian torsion, acute abdomen

INTRODUCTION

Disseminated peritoneal leiomyomatosis (DPL), first described in 1952 by Willson and Peale,[1] is a rare disease characterized by multiple benign subperitoneal leiomyomas in the pelvic and abdominal cavities. The etiology of DPL remains uncertain; however, DPL is thought to be highly related to estrogen exposure and predominates during the premenopausal years. DPL has been reported in postmenopausal females[1] and males.[2] DPL is usually asymptomatic and discovered incidentally, and can be confused with intra-abdominal carcinomatosis.[3] The diagnosis of DPL is based on surgical findings and histopathologic examination to exclude possible malignant transformation or other malignancies, such as gastrointestinal stromal tumors (GISTs).[4,5] The acute presentation of DPL mimicking ovarian torsion is presented here.

CASE REPORT

A 42-year-old nulliparous female presented to our ED on referral from a local clinic with abdominal distention and the abrupt onset of lower abdominal pain continuously during the morning hours. She underwent a total hysterectomy for recurrent uterine myomas with menorrhagia 4 years ago. The medical history was otherwise unremarkable. The initial vital signs were as follows: Blood pressure, 111/65 mmHg; heart rate, 81/min; respiratory rate, 17/min; and temperature, 37.5°C. The physical examination showed diffuse pelvic tenderness with rebound pain. A sonographic examination revealed a left adnexal mass with hypoechoic content measuring 10.99 cm × 7.54 cm × 9.51 cm. The color Doppler was done, which blood supply revealed disruption from vessel to tumor. Based on these findings, the patient was admitted to the hospital with a presumptive diagnosis of ovarian torsion.

A diagnostic laparoscopy was performed immediately. But after trocar inserted, we found multiple firm, soft tissue masses with severe adhesion that can't be finished by laparoscopy. So an emergent laparotomy was performed to prevent further complications. At the time of laparotomy, there were four firm, soft tissue masses in the pelvis and abdomen without a uterus. The largest mass, 10 cm × 8 cm in size, was located at the left adnexa and was adherent to the posterior aspect of the urinary bladder [Figure 1]. A second mass, 6 cm × 7 cm in size, arose from the ascending colon and the other smaller masses, 3 cm × 4 cm in size each, arose from the sigmoid colon and rectum. Examination of a frozen section suggested a leiomyoma, which was characterized by benign-appearing smooth muscle proliferation with eosinophilic cytoplasm and marked edematous changes in the soft tissue without evidence of malignancy [Figure 2]. All masses were completely resected during the laparotomy. The ovaries were normal in appearance and left *in situ*.

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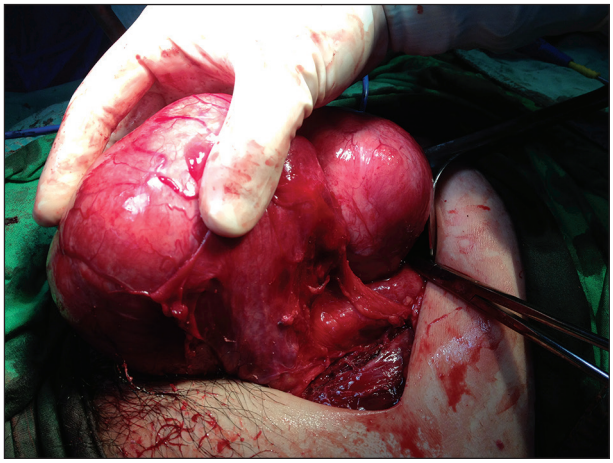

Figure 1: The largest pelvic mass about 10 × 8 cm in size, was located at the left adnexa and adherent to the posterior aspect of the urinary bladder

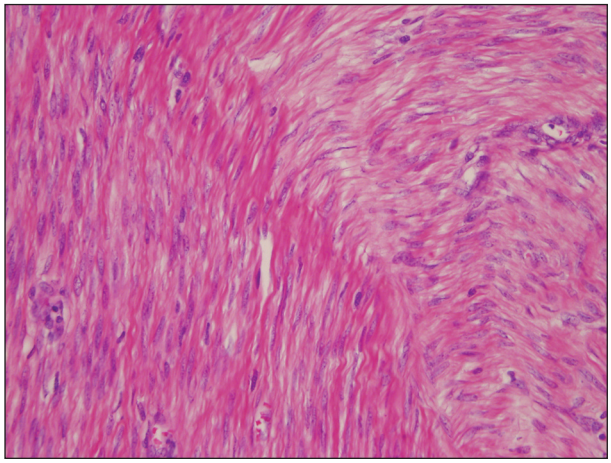

Figure 2: The pathologic finding was benign-appearing smooth muscle proliferation with eosinophilic cytoplasm and marked edematous changes in the soft tissue without evidence of malignancy. H&E stain, 400×.

The resected specimens were transported to the Anatomic Pathologic Division for examination. No mitoses, nuclear atypia, or necrosis was noted microscopically, and the Ki-67 ratio did not indicate an increased proliferation index. Immunostaining was positive for actin expression but negative for CD117, CD34, and S-100. Tumor markers, including CA125 and carcinoembryonic antigen, were within normal ranges. A final diagnosis of DPL was made. The patient had an uneventful postoperative course and discharged 2 days later.

DISCUSSION

Although the pathogenesis of DPL is unknown, pregnancy, prolonged oral contraceptive use, hormone replacement therapy, endometriosis, and estrogen-secreting ovarian tumors have been implicated.[6] Estrogen exposure is regarded to be an etiologic factor. In the current case, the patient underwent a total hysterectomy without oophorectomy, which implied that hormone stimulation may play a role in the progression of DPL. Miyake *et al*.[7] suggested that DPL found during the second or third surgeries results from implantation and proliferation of benign smooth muscle cells of uterine myomas from the initial surgery. A history of recurrent myomas, as in our patient, can be assumed to be a predisposing factor.

In our patient, a false-positive diagnosis of ovarian torsion was made and prompt surgery was indicated to prevent functional loss of the ovary.[7] Ovarian sonography with color Doppler that was done at Emergency Department is strongly recommended in patients suspected to have a torsion.[8] The image with blood supply revealed disruption from vessel to tumor, but image wasn't saving at that time.

An acute presentation of DPL is extremely rare.[9] The nodules of DPL range in size from 0.5 mm to 20 cm and involve a variety of structures in the abdominal and pelvic cavities.[10] An inadequate blood supply due to overgrowth of a leiomyoma results in acute ischemic pain, which may explain our patient's symptoms mimicking an ovarian torsion. But, unfortunately, there is no evidence of pathology can confirmed it. Maybe the disease progressed still in the beginning.

It is impossible to distinguish DPL from all possible conditions, such as malignancy or GIST, based on macroscopic findings alone; an intra-operative frozen-section analysis and postoperative histopathologic studies are essential. A higher mitotic index, nuclear atypia, tumor necrosis, infiltrative patterns, and a high Ki-67 ratio are the characteristics of leiomyosarcomas and other malignancies, unlike our patient.[4] GISTs express CD117 and CD34, which were both negative in our patient, and usually do not have smooth muscle cells.[4] S-100 protein is a common characteristic of neural crest-derived tumors, such as poorly differentiated schwannomas, which was also not expressed in our patient.[11]

Disseminated leiomyomas occurring after laparoscopic morcellation of a myoma is a rare but reported entity. The incidence of sarcomatous transformation in benign leiomyoma is 0.13-0.81%. No imaging method can enable a reliable preoperative diagnosis of leiomyosarcoma versus leiomyoma. Herein, it is reported a case in which disseminated peritoneal leiomyosarcoma developed after laparoscopic myomectomy and removal of the ''myoma'' via morcellation.[12] But in our case, there was no morcellation used during last surgery.

CONCLUSION

Disseminated peritoneal leiomyomatosis is a rare benign entity in the differential diagnosis of multiple peritoneal

masses with absence of ascites or constitutional symptoms, as are found in malignancies.[3] Although there are no accepted guidelines for the management of DPL, surgical intervention is the mainstay of treatment in symptomatic patients. Close follow-up imaging is suggested due to the possibility of recurrence and malignant transformation in DPL.[13,14]

CONFLICT OF INTEREST STATEMENT

The authors have no conflicts of interest to declare.

REFERENCES

1. Danikas D, Goudas VT, Rao CV, Brief DK. Luteinizing hormone receptor expression in leiomyomatosis peritonealis disseminata. Obstet Gynecol 2000;95:1009-11.
2. Halama N, Grauling-Halama SA, Daboul I. Familial clustering of Leiomyomatosis peritonealis disseminata: An unknown genetic syndrome? BMC Gastroenterol 2005;5:33.
3. Papadatos D, Taourel P, Bret PM. CT of leiomyomatosis peritonealis disseminata mimicking peritoneal carcinomatosis. AJR Am J Roentgenol 1996;167:475-6.
4. Bayrak S, Pasaoglu E, Cakar E, Bektas H, Colak S, Sevinc MM, *et al.* Disseminated peritoneal leiomyomatosis with chronic constipation: A case report. J Med Case Rep 2014;8:114.
5. Bisceglia M, Galliani CA, Pizzolitto S, Ben-Dor D, Giannatempo G, Li Bergoli A, *et al*. Selected case from the Arkadi M. Rywlin international pathology slide series: Leiomyomatosis peritonealis disseminata: Report of 3 cases with extensive review of the literature. Adv Anat Pathol 2014;21:201-15.
6. Al-Talib A, Tulandi T. Pathophysiology and possible iatrogenic cause of leiomyomatosis peritonealis disseminata. Gynecol Obstet Invest 2010;69:239-44.
7. Bar-On S, Mashiach R, Stockheim D, Soriano D, Goldenberg M, Schiff E, *et al.* Emergency laparoscopy for suspected ovarian torsion: Are we too hasty to operate? Fertil Steril 2010;93:2012-5.
8. Niafar F, Mirfendereski S, Rostamzadeh A. Diagnostic efficacy of sonography for diagnosis of ovarian torsion. Zahedan J Res Med Sci 2014;16:37-9.
9. Tan CH, Ho BC, Shelat V, Tan CH. Leiomyomatosis peritonealis disseminata presenting as omental torsion. Singapore Med J 2012;53:e71-3.
10. Fulcher AS, Szucs RA. Leiomyomatosis peritonealis disseminata complicated by sarcomatous transformation and ovarian torsion: Presentation of two cases and review of the literature. Abdom Imaging 1998;23:640-4.
11. Weiss SW, Langloss JM, Enzinger FM. Value of S-100 protein in the diagnosis of soft tissue tumors with particular reference to benign and malignant Schwann cell tumors. Lab Invest 1983;49:299-308.
12. Anupama R, Ahmad SZ, Kuriakose S, Vijaykumar DK, Pavithran K, Seethalekshmy NV. Disseminated peritoneal leiomyosarcomas after laparoscopic "myomectomy" and morcellation. J Minim Invasive Gynecol 2011;18:386-9.
13. Bekkers RL, Willemsen WN, Schijf CP, Massuger LF, Bulten J, Merkus JM. Leiomyomatosis peritonealis disseminata: Does malignant transformation occur? A literature review. Gynecol Oncol 1999;75:158-63.
14. Sharma P, Chaturvedi KU, Gupta R, Nigam S. Leiomyomatosis peritonealis disseminata with malignant change in a post-menopausal woman. Gynecol Oncol 2004;95:742-5.