



Case Report: A Rare Case of Disseminated Peritoneal Leiomyomatosis with Unusual Presentation

Authors

**Dr Sivatha Gopinath¹, Dr C Sanjeevi², Dr C.Santhosh³, Dr S.Rajadirajan⁴,
Dr S.Sethurajan⁵, Dr M.Sivakolunthu⁶, Dr M.Adaikappan⁷**

¹Post Graduate RMMCH, Chidambaram, Annamalai University

^{2,3,4,5}Assistant professors, RMMCH, Chidambaram, Annamalai University

^{6,7}Professor, RMMCH, Chidambaram, Annamalai University

ABSTRACT

Disseminated peritoneal leiomyomatosis (DPL) is a rare benign disease characterised by multiple peritoneal smooth muscle nodules growing along the submesothelial tissues of the abdomino pelvic peritoneum. It is associated with increased levels of serum gonadal steroids. Almost all cases of DPL are associated with a gynaecological disorder or mass in the abdominal cavity. We present a case of DPL who manifested with chronic constipation and abdominal distension.

Keywords: Abdominal distension, chronic constipation, disseminated peritoneal leiomyomatosis (DPL)

INTRODUCTION

DPL is a benign condition and a very rare disease, first described in 1952 by Wilson and Peale⁽¹⁻³⁾. It is generally seen in women of child bearing age with estrogen hypersecretion⁽¹⁾. It is thought to be associated with benign smooth muscle proliferation originating from the multicentric metaplasia of the peritoneal surfaces. It rarely occurs in men with no excess hormones and in postmenopausal women. In this study we report a case of diffuse peritoneal leiomyomatosis presenting with complaints of chronic constipation.

CASE REPORT

A 38 yr old female patient Para 1 Live 1 came with complaints of abdominal distension and constipation for the past 6 months. She had no complaints of menstrual problems and there was no history of loss of weight or loss of appetite. There was no previous surgical history or long

term usage of oral contraceptive pills. Tumour markers (CA 125) were normal. Colonoscopy was normal.

IMAGING FINDINGS

Plain Radiograph abdomen shows large soft tissue density lesion displacing transverse colon superiorly and pushing other bowel loops laterally (Figure-1). Ultrasound shows multiple discrete soft-tissue density whorled lesions noted with in peritoneal cavity few of which shows fluid density at the centre. Multi loculated ascites with no central or peripheral calcification (Figure-2). Contrast enhanced CT showed multiple hypodense nodules in peritoneum with moderate enhancement and loculated ascites. Uterus with multiple hypodense lesions noted with soft tissue attenuation of varying size distorting the uterine contour & endometrial cavity (Figure-3). In MRIT2-weighted images the lesions appear

homogeneously low signal intensity. Hyaline degeneration has characteristic diffuse speckled pattern of higher signal foci in low-signal intensity background. Cystic degeneration occurs centrally, and fluid shows angular margins. The solid portion retains its low signal intensity, whorled appearance, and pseudo-capsule (Figure 4).



Figure – 1: X-ray Abdomen shows radio opaque mass displacing bowel loops



Figure – 2: USG shows multiple peritoneal whorled iso to hypo echoic nodules.



Figure – 3: Contrast enhanced CT Scan shows multiple peritoneal hypodense lesions with moderate enhancement.

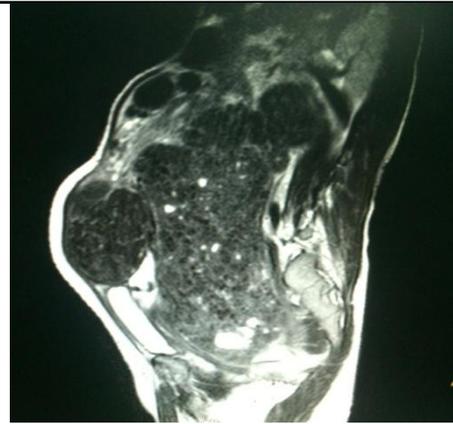


Figure 4 MRI Saggital T2 w image shows multiple peritoneal nodules displacing bladder & rectum.

DISCUSSION

We report a case of disseminated peritoneal leiomyomatosis presenting with complaints of chronic constipation and abdominal distension: Disseminated peritoneal leiomyomatosis (DPL) is a rare benign disease characterised by multiple peritoneal smooth muscle nodules growing along the submesothelial tissues of the abdomino pelvic peritoneum. Female patients with DPL in cases reported in the literature were generally pregnant, using oral contraceptives or had malignant ovarian tumors expressing high estrogen and/or progesterone at the time of diagnosis^(4,5).

USG shows spectrum of features ranging from multiple solid subcentimetric nodules to large solid masses. CT shows masses with homogeneous or heterogeneous attenuation with a variable enhancement pattern. MRI shows multiple masses with signal intensity similar to that of skeletal muscle and smooth muscle on both T1- and T2-weighted images. Homogeneous enhancement following the administration of contrast material. It is difficult to diagnose DPL due to its resemblance to malignant conditions⁽⁶⁾. The main differentials would be Peritoneal carcinomatosis, Peritoneal lymphomatosis, Gastrointestinal stromal tumours (GIST), Leiomyosarcoma.

In Peritoneal carcinomatosis carcinoma from GIT (stomach, colon, appendix, gallbladder and Pancreas) metastasize to the peritoneal surfaces. Patients typically manifest with weight loss, nausea, vomiting, ascites and abdominal pain from

bowel obstruction. Imaging findings includes ascitic fluid with hypoechoic deposits, omental cake, calcifications (nodular with noncalcified component). MRI shows low to intermediate signal in T1 w Images and intermediate to high signal intensity in T2w images.

Peritoneal lymphomatosis occurs secondary to a pre-existing lymphoma. Imaging findings includes splenomegaly, ascites and diffuse retroperitoneal and mesenteric lymphadenopathy, sometimes forming masses causing encasement of superior mesenteric artery and vein producing 'Sandwich Sign'. There will be evidence of peritoneal enhancement and thickening, omental caking and extensive lymphadenopathy of retrocrural region and small bowel mesentery. MRI shows low signal intensity in T1 w and high signal intensity in T2 w images⁽⁷⁾.

GIST are the most common mesenchymal tumour of the GIT. The mass is of soft tissue dense lesion with central areas of low density when necrosis is present. MRI shows the presence of necrosis, haemorrhage and cystic changes. T1w images shows low signal intensity solid component with peripheral enhancement and increased signal intensity solid component T2 w images⁽⁸⁾.

Leiomyosarcoma (LMS) characteristically shows nodules with mitotic activity and cellular atypia. PET is the most useful distinguishing tool to differentiate DPL and LMS.

CONCLUSION

With all these imaging findings, exploratory laparotomy done and histopathological diagnosis confirmed disseminated peritoneal leiomyomatosis. It is important to diagnose DPL because some cases can show malignant transformation with p53 expression⁽³⁾. p53 expression can also be checked in patients with DPL since it is associated with poor prognosis. Most cases present with gynaecological problems however our case had unusual presentation of chronic constipation with abdominal distension. The clinical course is almost invariably benign though close surveillance is mandatory because of rare sarcomatous transformation.

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