



## DISSEMINATED PERITONEAL LEIOMYOMATOSIS : A CASE REPORT.

## Pathology

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## ABSTRACT

Disseminated peritoneal leiomyomatosis (DPL) is rare clinical condition where multiple smooth muscle nodules or growth of various sizes implant on the peritoneal surface of the abdomen and pelvis. Disseminated peritoneal leiomyomatosis affects women in reproductive age. These diffuse tumors are usually incidentally found during surgery. Surgeons and pathologists face a challenge when dealing with DPL because its macroscopic appearances resembles peritoneal carcinomatosis. Here we report a case of 30 year old female Gravida 2, Para 1 with uncomplicated antenatal course presenting with multiple hard nodules noted over the surface of uterus, ovaries, bladder and omentum intraoperatively during caesarean section. The macroscopic appearance led to suspicion of some proliferative process. However the final diagnosis of DPL was made on histopathology.

## KEYWORDS

Disseminated peritoneal leiomyomatosis, smooth muscle nodules, caesarean section.

## INTRODUCTION

Disseminated peritoneal leiomyomatosis (DPL) also known as leiomyomatosis peritonealis disseminate (LPD) is a rare condition described as multiple smooth muscle tumors disseminated throughout the peritoneum, omentum and pelvic structures.<sup>[1,2]</sup> The first case was reported by Willson and Peale in 1952; however, the entity was clearly described and named by Taubert et al in 1965.<sup>[3]</sup> Less than 200 case reports have been described in the literature, with the majority of cases being diagnosed incidentally at the time of laparoscopy for other reasons.<sup>[4]</sup> The lesions occur most commonly in women of child-bearing age and may have an association with oral contraceptive therapy (OCP) or hormone replacement therapy (HRT), pregnancy and endometriosis.<sup>[5]</sup> Since female gonadal steroids play an important role in the pathogenesis of LPD, it is generally associated with high levels of exogenous and endogenous female gonadal steroids.<sup>[6-9]</sup> The disease is mainly asymptomatic; only a few cases of abdominal pain, urinary tract problems and vaginal or rectal bleeding have been described.<sup>[10]</sup> Etiology is unknown. However, some have hypothesized that DPL occurs due to hormonal stimulation of smooth muscle cells. These diffuse tumors are usually incidentally found during surgery.<sup>[3]</sup> Surgeons and pathologists face a challenge when dealing with LPD because its macroscopic appearances resembles peritoneal carcinomatosis.<sup>[11]</sup>

Here we describe an incidental case of DPL found in a 30 year old female during cesarean section.

## Case Report

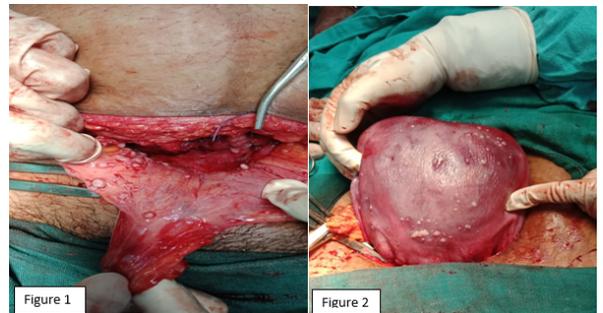
A 30 year old female Gravida 2, Para 1 with previous history of cesarean section was admitted in our hospital for elective caesarean section. The patient had an uncomplicated antenatal course. During caesarean section, after the delivery of foetus, multiple hard nodules were noted over the surface of uterus, ovaries, bladder and omentum. The operating team removed the omental nodules and send it for histopathological examination due to suspicion of proliferative process.

The patient had an uneventful recovery period and was discharged on day 5 in a good condition. These nodules were not detected in any obstetric ultrasound during pregnancy and the patient had no other previous abdominal imaging done prior to pregnancy.

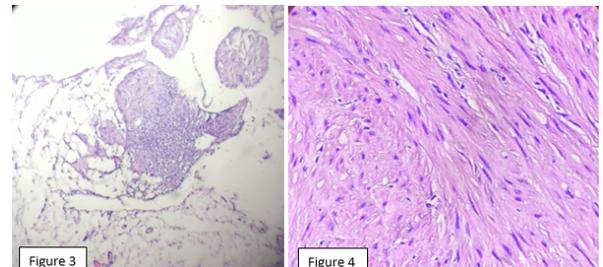
The omental nodule was initially sent for frozen section. We received a small fibrofatty tissue (2cc) with multiple grey white small nodules of various sizes over it, largest measuring 0.7 x 0.6 x 0.2 cm. On frozen section a diagnosis of benign spindle cell lesion was made possibilities

included – leiomyoma and fibrotic nodule. The same tissue was processed in histopathology section of our department and stained by H & E stain. On histopathological sections, nodules were composed of interlacing bundles of smooth muscle cells without cytological atypia and mitosis. Decidual reaction was noted in few nodules. On immunohistochemistry, the spindle cells stained positive for SMA (smooth muscle antigen) and desmin.

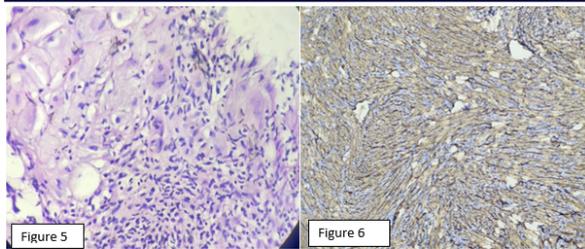
On the basis of these findings along with clinical presentation, we arrived at the diagnosis of DPL (disseminated peritoneal leiomyomatosis). The patient was scheduled for follow up twelve weeks postpartum.



**Intraoperative Findings: Figure 1 & 2- Multiple nodules over peritoneal surface and uterus.**



**Figure 3: Scanner view : Nodules of smooth muscle bundles proliferation. Hematoxylin and Eosin Stain. Figure 4: High power view (40x H&E staining) of leiomyomatous nodules. The spindle cells show no cytological atypia, and no mitoses are identified.**



**Figure 5: High power view (100X, H&E staining) of nodules showing decidual cells surrounded by smooth muscle cells. Figure 6: High power view of immunohistochemical staining demonstrating SMA (Smooth muscle actin) positivity.**

## DISCUSSION

Leiomyomas are benign smooth muscle tumours that arise from the overgrowth of smooth muscle and connective tissue, usually from the uterus, small bowel and the oesophagus. Uterine leiomyomas are the most common gynaecological tumours noted to occur in 20% to 30% of women beyond the age of 35 years. Uncommon growth patterns of leiomyoma include disseminated peritoneal leiomyomatosis (DPL), benign metastasizing leiomyoma, intravenous leiomyoma, retroperitoneal leiomyoma and parasitic leiomyoma.<sup>[3]</sup>

Disseminated peritoneal leiomyomatosis is a rare disorder characterized by the presence of multifocal nodules arising from the proliferation of smooth muscle cells, fibroblasts and myofibroblasts emerging along submesothelial tissue located in the pelvic cavity. Disseminated peritoneal leiomyomatosis affects women in reproductive age.<sup>[10]</sup>

Most of the patients with DPL are asymptomatic, with lesions detected incidentally during surgery (caesarean section, laparotomy or laparoscopy). Others usually present with non-specific symptoms such as abnormal, heavy uterine bleeding and lower abdominal pain or discomfort. Less common presentations include increased frequency of micturition, mass per abdomen and symptoms of obstructive uropathy.<sup>[3]</sup>

Although the aetiopathogenesis is controversial, strong hormonal association is attributed owing to coexisting factors such as pregnancy, long-term use of oral contraceptives and oestrogen-producing tumours.<sup>[12]</sup>

Since, the disease is seen in post menopausal women and in males, the possible causes of it could be divided into hormonal, subperitoneal mesenchymal stem cells metaplasia, genetic, or iatrogenic after morcellation of myoma during laparoscopic surgery.<sup>[13,14]</sup>

Some cases of DPL are associated with surgery of uterine fibroids, especially using a power morcellator. Morcellation is a risk factor of DPL presentation.<sup>[15,16]</sup> One author describes fibroid tissue implantation at the point of laparoscopic trocar entry after laparoscopic myomectomy; the patient was referred because of the easily palpable tumor.<sup>[17]</sup> The potential for malignant transformation and the possible iatrogenic etiology makes close follow up important in patients with DPL.<sup>[5]</sup> Final DPL diagnosis can only be established through a histopathological examination and finding of smooth muscle cells without atypia or necrosis. Differential diagnoses are as follows: parasitic leiomyoma, intravenous leiomyomatosis, peritoneal carcinosarcoma or leiomyosarcoma. Leiomyomatosis must always be differentiated from metastatic leiomyosarcoma. It is believed that leiomyosarcoma nodules are generally fewer in number, bigger and they infiltrate surrounding tissues.<sup>[10]</sup>

Therefore in women who do not wish to become pregnant a total abdominal hysterectomy, salpingo-oophorectomy, omentectomy and debulking surgery is the best option.<sup>[18,19]</sup> However, non- surgical options involve limiting hormonal exposure which have been shown to decrease tumor size.<sup>[2]</sup> These include avoiding oral contraceptives (OCP) and hormone replacement therapy (HRT), administering gonadotropin releasing hormone agonist aromatase inhibitor therapy and avoiding pregnancy. Without proper treatment, DPL can cause complications such as pain, bleeding, chronic constipation and infertility.<sup>[5]</sup>

## CONCLUSION

Disseminated peritoneal leiomyomatosis is rare clinical condition where multiple smooth muscle nodules or growth of various sizes implant on the peritoneal surface of the abdomen and pelvis.<sup>[4]</sup> It is usually diagnosed in patients undergoing pelvic surgery where multiple nodules implanted on the pelvic and peritoneal surfaces are found incidentally, these give the appearance of carcinosarcoma.<sup>[20]</sup> This condition occurs mainly in premenopausal patients and has a benign course in the majority of cases, with less than 5% undergoing malignant transformation.<sup>[4]</sup> Histopathology is the confirmatory tool.<sup>[20]</sup> The risk for recurrence, metastasis, colonic obstruction, impaired quality of life, obstetric complications, and possible malignant transformation underlines the importance of accurate diagnosis, facilitating patient specific treatment.<sup>[5]</sup>

## REFERENCES

- Nappi C, Di Spiezio Sardo A, Mandato VD, Bifulco G, Merello E, et al. Leiomyomatosis peritonealis disseminata in association with Currarino syndrome. *BMC Cancer* 2006; 6:127.
- Wu C, Zhang X, Tao X, Ding J, Hua K, et al. Leiomyomatosis peritonealis disseminata: A case report and review of the literature. *Mol Clin Oncol* 2016; 4(6):957-958.
- Hiremath SB, Benjamin G, Gautam AA, Panicker S, Rajan A. Disseminated peritoneal leiomyomatosis: a rare cause of enigmatic peritoneal masses. *BJR Case Rep* 2016; 2: 20150252.
- Kruger P F. Disseminated peritoneal leiomyomatosis: A case report and review of current events in the media and the literature. *South African Journal of Obstetrics and Gynaecology* 2014; 20(3): 91-92.
- Rezai S, Hughes A, Ligorski J, Cheung ML, Lumapas S, et al. Disseminated Peritoneal Leiomyomatosis (DPL); A Case Report and Review of Literature. *Obstet Gynecol Int J* 2017; 7(2): 00240.
- Drake A, Dhundee J, Buckley CH, Woolas R. Disseminated leiomyomatosis peritonealis in association with oestrogen-secreting ovarian fibrothecoma. *BJOG* 2001; 108: 661-4.
- Surmacki P, Sporny S, Tosiak A, Lasota J. Disseminated peritoneal leiomyomatosis coexisting with leiomyoma of the uterine body. *Arch Gynecol Obstet* 2006; 273: 301-3.
- Ramesh L, Edozien L. Not all disseminated intra-abdominal lesions are malignant: A case of leiomyomatosis peritonealis disseminata. *J Obstet Gynaecol* 2005; 25: 409.
- Altinok G, Usubutun A, Küçükalı T. Disseminated peritoneal leiomyomatosis A benign entity mimicking carcinosarcoma. *Arch Gynecol Obstet* 2000; 264: 54-5.
- Ciebierra M, Slabuzewska-Jóźwiak A, Zareba K, Jakiel G. A case of disseminated peritoneal leiomyomatosis after two laparoscopic procedures due to uterine fibroids. *Videosurgery and other Minimvasive Techniques* 2017; 12(1): 110-114.
- Momtahan M, Nemati M, Safaei A. Disseminated peritoneal leiomyomatosis. *Iran J Med Sci* 2011; 36(1): 57-59.
- Drake A, Dhundee J, Buckley CH, Woolas R. Disseminated leiomyomatosis peritonealis in association with oestrogen secreting ovarian fibrothecoma. *BJOG* 2001; 108: 661-4.
- Abulafia O, Angel C, Sherer DM, et al. Computed tomography of leiomyomatosis peritonealis disseminata with malignant transformation. *Am J Obstet Gynecol* 1993; 169: 52-4.
- Al-Talib A, Tulandi T. Pathophysiology and Possible Iatrogenic Cause of Leiomyomatosis Peritonealis Disseminata. *Gynecol Obstet Invest* 2010; 69: 239-44.
- Lete I, González J, Ugarte L, et al. Parasitic leiomyomas: a systematic review. *Eur J Obstet Gynecol Reprod Biol* 2016; 203:2509.
- Rosenbaum L. N-of-1 policymaking – tragedy, trade-offs, and the demise of morcellation. *N Engl J Med* 2016; 374: 986-90.
- Ostrzenski A. Uterine leiomyoma particle growing in an abdominal wall incision after laparoscopic retrieval. *Obstet Gynecol* 1997; 89: 853-4.
- Toriyama A, Ishida M, Amano T, Nakagawa T, Kaku S, et al. Leiomyomatosis peritonealis disseminata coexisting with endometriosis within the same lesions: a case report with review of the literature. *Int J Clin Exp Pathol* 2013; 6(12): 2949-2954.
- Ordu lu Z, Dal Cin P, Chong WW, Choy KW, Lee C, et al. Disseminated peritoneal leiomyomatosis after laparoscopic supracervical hysterectomy with characteristic molecular cytogenetic findings of uterine leiomyoma. *Genes Chromosomes Cancer* 2010; 49(12): 1152-1160.
- Sayed A, Black D, Murshed K, Khaldi HM. Disseminated peritoneal leiomyomatosis: A case report and literature review. *European Journal of Obstetrics and Gynecology and Reproductive Biology* 2019; 234:e193.