

## LEIOMYOMATOSIS PERITONEALIS DISSEMINATA

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Leiomyomatosis peritonealis disseminata (LPD) is a relatively rare clinical entity which is characterized by the presence of multiple nonneoplastic nodules, mainly due to smooth muscle proliferation over the surface of the abdominopelvic peritoneal cavity. It is usually an incidental finding during procedures such as laparoscopy, cesarean section, laparotomy, and postpartum tubal ligation, and it occurs mainly in women, primarily during the reproductive period. When LPD occurs during pregnancy or during the use of birth control pills, it may regress spontaneously after delivery or discontinuation of the use of the pills. Conservative care is indicated, particularly if fertility is desired. In this study, we report a case of LPD in a pregnant Egyptian woman. To our knowledge, this is the first case reported in an Arab woman.

### Case Report

A 38-year-old pregnant Egyptian woman of 38 weeks' gestation, gravida 4 para 3, was admitted to the Maternity Hospital, Riyadh Medical Complex, for a cesarean section. The previous history of the patient was unremarkable, except for two previous lower segment cesarean sections for cephalopelvic disproportion, with no intraoperative or postoperative complications. The patient had used oral contraceptive pills for a period of one year three years previously, but there was no other history of use of intrauterine contraceptive device or other hormonal treatment. Physical examination on admission revealed temperature of 37°C, blood pressure of 120/80, and mild lower limb edema. Abdominal examination revealed normal pregnancy at term cephalic 4/5.

Within six hours of admission, the patient underwent a cesarean section, and a baby girl was delivered weighing 4.95 kg, with an Apgar score of 9.99. Examination of the abdomen during surgery revealed multiple (about 20-25) small fleshy nodules varying in diameter from 0.1 to 0.5 cm, and scattered over the uterus, fallopian tubes,

omentum, and anterior abdominal wall. No nodules were seen over the bowel surface or the liver. A left ovarian multilocular cystic mass measuring 10x6x6 cm was observed. No ascitis was present and the uterus appeared free of any intramural tumor. With a possible diagnosis of disseminated carcinomatosis, examination of a frozen section specimen from one of the nodules was made, which revealed the mass to be a benign leiomyoma. Bilateral salpingo-oophorectomy and biopsy of another peritoneal nodule was carried out. The postoperative recovery was uneventful and the patient was discharged on the eighth postoperative day. Follow-up ultrasound examination three months later revealed no more nodular masses in the abdominal cavity.

### Pathologic Findings

The left ovary was replaced by a well-defined cystic mass measuring 10x6x6 cm. The right ovary was unremarkable. A cross-section view of the left ovarian mass showed a cystic cavity filled with thick granular yellow material with some hair structures. The two biopsies from the peritoneal nodules measured 0.7x0.5x0.5 cm in diameter each. They were smooth and glistening, firm, and pale gray in color. Microscopic examination of the mesenteric nodules revealed interlacing bundles of smooth muscle fibers which were mixed with interstitial edematous stroma (Figure 1). No clear decidual reaction was observed. No mitosis or atypical changes were present. Trichrome stain showed red-brown color in the cytoplasm of the cells as expected in typical smooth muscle differentiation. Immunohistochemistry markers for desmin and actin were positive (Figure 2). Sections from the left ovarian cyst revealed a classic benign cystic teratoma with areas lined by squamous epithelium with hair and areas showing cartilaginous differentiation.

### Discussion

Since Wilson and Pale<sup>1</sup> first described LPD in 1952, approximately 115 cases have been reported in the literature. LPD is most often found incidentally during laparoscopic examination or surgical exploration for pelvic abnormalities or is discovered at the time of routine examination during cesarean section. This benign condition is frequently clinically confused with disseminated carcinomatosis by the surgeon because of the presence of

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FIGURE 1. A section of the peritoneal nodule showing mature smooth muscle cells arranged in fascicular pattern (H&E, 200x).



FIGURE 2. Immunoperoxidase staining for actin shows strong positivity (Avidin-Biotin peroxidase, 200x).

multiple nodules of varying sizes, from a few mm to 10 cm in diameter, widely distributed throughout the abdominal cavity and pelvis.<sup>2</sup> Analysis of about 107 previously reported cases of LPD reveals that the age of the patients ranges from 20 to 64 years.<sup>3,4</sup> Most of the cases reported were in their reproductive years and were mostly associated with pregnancy.<sup>4-6</sup> Some of the non-pregnant women were under hormonal therapy,<sup>7-9</sup> or had granulosa cell tumor of the ovary.<sup>1</sup> A few cases were in the menopausal or perimenopausal age groups.<sup>2,10</sup> LPD has been reported more frequently in Caucasians and blacks, but a few cases in Asiatic women have also been reported.<sup>11</sup>

The most common condition of LPD has been leiomyoma involving the uterus,<sup>5,9,10</sup> but associated cases involving the intestines and producing intestinal obstruction have been reported.<sup>12</sup> Williams and Pavlick have suggested that most of the leiomyomata of the uterus are probably additional foci of LPD, rather than uterine leiomyomata, since in most instances they are subserosal in location.<sup>2</sup> Another frequent association is the presence of endometriosis<sup>5,7,9</sup> and occasionally ovarian teratoma,<sup>6</sup> as in the present case. Histologic examination usually reveals bundles of spindle cells with no significant nuclear pleomorphism or mitoses characteristic of a leiomyoma, as it was in this case. Ultrastructural studies clearly demonstrate a leiomyomatous differentiation.<sup>2,9</sup> The lesions are always well defined, and no evidence of infiltration is observed in the surrounding soft tissue. Although decidual reaction has been described in some LPD cases associated with pregnancy,<sup>6</sup> in this case the group of cells were separated by edematous stromal tissue but no definitive decidual reaction was noted.

Regression of LPD has been reported to occur in most patients who have been clinically followed up, but recurrence<sup>11</sup> or persistence during a second pregnancy has been reported.<sup>5</sup> Regression is thought to be a result of bilateral oophorectomy or termination of pregnancy. Although the disease has been shown to regress spontaneously, it may persist for many years. Malignant transformation has been reported in a few cases.<sup>3</sup>

There is evidence to suggest that LPD is a product of a hormonal imbalance, such as its exclusive occurrence in women, the association with pregnancy, hormone administration, granulosa cell tumor and regression after surgical castration, as well as the reduction in the size of the tumor after termination of pregnancy. Furthermore the presence of estrogen and progesterone receptors within the cells of the lesion,<sup>13</sup> and the experimental production of LPD in guinea pigs by the administration of estrogen alone or in combination with progesterone,<sup>14,15</sup> support the hormonal background of this disorder.

Metaplasia of the subcelomic mesenchyme has been proposed as the method of development of LPD which occurs in endometriosis. Embryologically, the celomic mesenchyme gives origin to the peritoneal lining and to the female internal genitalia. Several reports have demonstrated endometrial glandular inclusion within the leiomyomatous nodules in the LPD.<sup>7,9</sup> Endometriosis and LPD have been found in the same patient. The submesothelial mesenchymal cells are multipotential, and may differentiate into endometriosis,<sup>9</sup> or into smooth muscle-causing LPD, or to decidua and later into myofibroblast, which in turn may differentiate into smooth muscle. Although a few studies have demonstrated an imbalance in the hormone status in these patients, the results are contradictory.<sup>2</sup>

It is interesting that although gynecologic hormonal disorders are frequently observed in women, LPD is rarely observed, therefore, the possibility of an unusual sensitivity of the celomic tissue undergoing metaplastic changes observed in these patients remains a possible etiologic factor. Various surgical procedures, including total abdominal hysterectomy, bilateral salpingo-oophorectomy, omentectomy, myomectomy and debulking of the nodules, have been performed at the time of diagnosis. These procedures have either been done alone or in combination, however, treatment of LPD should be conservative since the disease usually regresses after the removal of the hormonal stimulus (e.g., after delivery). Radical excisions are unnecessary. The need for discussion of results of intraoperative frozen section specimen of the leiomyoma

cannot be overemphasized, as it is very helpful in clarifying the benign nature of this condition and thus preventing unwarranted surgery. Follow-up should include a pelvic examination, ultrasound and possibly laparoscopy.

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