Leiomyomatosis Peritonealis Disseminata in a Preeclamptic Pregnant Woman

Preeklamptik Gebe Bir Kadında Dissemine İntraperitoneal Leiyomiyomatoz

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ABSTRACT Leiomyomatosis peritonealis disseminata (LPD) is characterized by multiple peritoneal nodules of varying sizes on the omentum and peritoneal surfaces, grossly mimicking disseminated carcinoma, lymphoma, tuberculosis and desmoid tumors. The underlying mechanism of LPD development in females is still unknown, but high levels of estrogen and progesterone seem to play a major role. We describe a 25 years old preeclamptic pregnant woman. She was urgently taken to the cesarean section. Multiple nodules were seen on the surface of the omentum and especially the pelvic, uterine, fallopian tube, ovary, bladder peritoneum. Histopathological examination revealed small nodules consisting whorls of desidualized spindle cells. Immunohistochemical study also support the diagnosis. This asymptomatic disease is usually detected unexpectedly at the time of cesarean section or laparotomy. So this entity should be considered in the differential diagnosis of multiple peritoneal nodules especially in women of reproductive age. Diagnosis is often difficult and a histopathological analysis is needed.

Key Words: Leiomyomatosis; pregnancy; pre-eclampsia

ÖZET Dissemine İntraperitoneal Leiyomiyomatoz (DİL); yaygın karsinom, lenfoma, tüberküloz ve desmoid tümörleri taklit edebilen, omentum ve peritoneal yüzeylerde bulunan, değişen boyutlarda çok sayıda peritoneal nodüllerle karakterizedir. Kadınlarda DİL gelişiminin altında yatan mekanizma hala bilinmemektedir, fakat östrojen ve progesteronun yüksek seviyelerinin önemli bir rol oynadığı görünmektedir. Biz 25 yaşında, preeklamptik bir gebeyi tanımlıyoruz. Hasta acil olarak sezaryene alındı. Özellikle pelvik, uterin, fallop tüpü, over ve mesane peritonu ve omentum yüzeyinde çok sayıda nodül izlendi. Histopatolojik incelemede küçük nodüllerin desidualize iğsi hücre sarmalları içerdiği gözlendi. İmmünhistokimyasal çalışma da tanıyı destekledi. Bu asemptomatik hastalık genellikle laparotomi veya sezaryen esnasında beklenmedik bir şekilde tespit edilir. Bu nedenle, bu antite özellikle üreme çağındaki kadınlarda çok sayıda peritoneal nodüllerin ayırıcı tanısında düşünülmelidir. Tanı çoğu zaman zordur ve histopatolojik inceleme gereklidir.

Anahtar Kelimeler: Leiyomiyomatoz; gebelik; pre-eklampsi

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Leiomyomatosis peritonealis disseminata (LPD) is characterized by the presence of multiple peritoneal nodules throughout the peritoneal cavity, grossly mimicking disseminated carcinoma, lymphoma, tuberculosis and desmoid tumors. The condition was first described in 1952 by Wilson and Peale.[1] The lesion usually occurs in women of childbearing age. The incidence and the possible pathophysiology of this condition remain unclear, but the possible causes could be divided into hormonal, sub-

peritoneal mesenchymal stem cells, metaplasia, genetic or iatrogenic. The estrogen and progesterone receptor positivity in LPD support the role of hormones in the pathogenesis.[2,3] LPD is in the majority of cases described in premenopausal women, often associated with pregnancy or oral contraceptive use.[4] Spontaneous regression of LPD can be seen after discontinuation of oral contraceptives or pregnancy. In other way; Sizzi et al. reported 3 cases of LPD among 2,050 women who underwent laparoscopic myomectomy indicating the unusual nature of this condition.[5] In recent years, several authors reported LPD in women with a past history of myoma morcellation. This raises the question whether the condition could occur iatrogenically. There is a relation between LPD and endometriosis. Several authors suggested that both LPD and endometriosis cells might derivate from the same origin, the submesothelial multipotential mesenchymal cells. Occurrence of LPD with malignant transformation to leiomyosarcoma is unusual. Being still a rare disease and not often considered as a differential diagnosis, LPD remains an important issue. We report the case of a 25-year-old pregnant woman suffering from preeclampsia with LPD.

CASE REPORT

We describe a 25 years old pregnant woman at the 38 1/7 week of gestation. She was admitted to the hospital because of the contractions. Blood pressure was 180/100 mmHg and the urine test showed (++) proteinuria. She had a severe headache and upper abdomen pain. She also had hypertension history during pregnancy. The patient underwent laparoscopic ovarian simple cyst excision four years ago and ceserean section two years ago in her medical history. There was nothing about LPD in her history and also previous laparoscopy and cesarean records. She never used any kind of hormonal contraceptive methods. In her first pregnancy; there was no history about gestational hypertension and preeclampsia. She was urgently taken to the cesarean section because of severe preeclampsia. A 2850 gram boy with APGAR 8/9 was born. During the operation; the surgeon observed multiple nodules on the surface of the omentum, pelvic, uterine, salpingeal peritoneum after delivery. The intraoperative findings appeared like an excessive malignant or infectious process with multiple 4-5 mm abdominal nodules. There were no uterine leiomyomatous nodules in uterus. The surgeon decided intraoperatively to perform an partial omentectomy to reduce the tumor masses and to obtain representative tissue for histological examination and then performed multiple biopsies of these lesions and also partial omentectomy.

On gross examination of the omentectomy and these biopsy specimen showed 4-5 mm solid white coloured nodular lesions. Histopathological examination revealed small nodules consisting whorls of desidualized spindle cells which were focally positive with Smooth Muscle Actin (SMA) and negative with S100, CD34, CD117. Histopathological analysis revealed benign tumors classified as leiomyomatosis (Figure 1, 2).

Postpartum a spontaneous regression of the preeclampsia was marked as known. The laboratory parameters normalized and so did the patients symptoms. Six months post operatively the patient is still wellbeing with no hypertension. Sonographic images as well as the follow up computerized tomography and magnetic resonance images did not reveal any sign of progressing leiomyomatosis lesions.

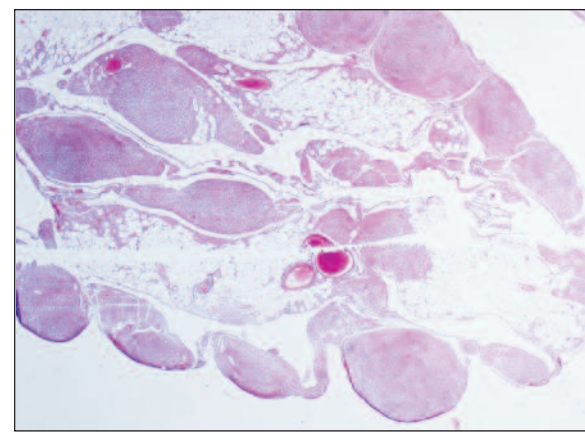

FIGURE 1: Well circumscribed multiple peritoneal nodules (HE, x20).
(See color figure at http://www.turkiyeklinikleri.com/journal/turkiye-klinikleri-journal-of-case-reports/1300-0284/tr-index.html)

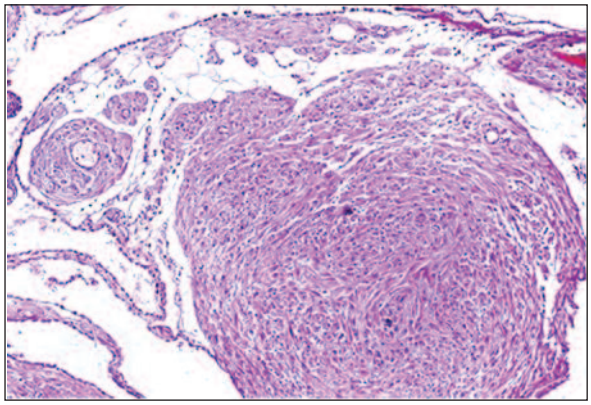

FIGURE 2: The nodules are composed of fibroblasts and bland smooth muscle cells (HE, x100).

(See color figure at http://www.turkiyeklinikleri.com/journal/turkiye-klinikleri-journal-of-case-reports/1300-0284/tr-index.html)

DISCUSSION

LPD is a rare lesion characterized by multiple subperitoneal nodules of benign smooth muscle. These myomatous nodules vary in size and can be found on omental or any of the peritoneal surfaces, especially the pelvic, uterine fundal, colonic mesenteric, ovarial, splenic and pancreatic ones.[6] Grossly DPL appears as multiple, hard solid nodules and microscopically, the nodules have an appearance of typical leiomyoma and in pregnant women they may also consist of decidual cells.[7] When the lesion is composed of highly desidualized cells immunoreactivity with SMA may decrease. Usually nuclear pleomorphism and mitotic figures are absent.[8] Often additional uterine leiomyomas may be present. No clear correlation between LPD and uterine leiomyomas could be ascertained so far and cases without uterine leiomyomas have also been reported.[9] In our case, there was no uterine submucosal or intramuscular leiomyoma.[10]

The clinical course of LPD is usually asymptomatic. In symptomatic cases; ultrasonography, computerized tomography-scan and magnetic resonance may be useful for differential diagnosis of malignancies and infectious diseases. And also serologic markers or special tests (like purified protein derivative of tuberculin [PPD]) can be used for detection of other LPD mimicking lesions. The disease is usually detected unexpectedly at the time of a cesarean section or laparotomy. Female gonadal steroids play a major role in the pathogenesis, but occasional reported cases in postmenopausal women do indicate other causative factors.[10] So the possible causes could be divided into hormonal, subperitoneal mesenchymal stem cell origin, metaplasia, genetic or iatrogenic. Imaging features are often characteristic and additional findings such as uterine involvement and ovarian endometrioma support the diagnosis. The clinical course is almost invariably benign; however, sarcomatous transformation has been reported.[11,12] Though rare, systemic metastases have been reported and should not preclude diagnosis of LPD. Surgery remains mainstay treatment for LPD. Favorable response can be achieved with advent of systemic chemotherapy in patients developing unresectable or metastatic disease.[13,14]

Hormonal exposure seems to play a major role in LPD development. Diagnosis is always difficult and histopathological analysis is needed. The disease is rare and that is probably why it is hardly considered as a differential diagnosis in young females. This is because in pregnant women extremely high levels of hormones such as estrogen and progesterone are reached. So far no standard therapy (surgical, GnRH agonists) is available in pregnancy. The only causal therapy in pregnant women is to prolong pregnancy as long as possible and perform a cesarean section before the patient or the unborn get into difficult situations. Inspite of this, LPD remains a diagnostic and therapeutic challenge.[15]

Our case was pregnant and diagnosed as LPD after the histopathologic examination of milimetric peritoneal nodules which were examined during the ceserean section. These nodules did not progress after the pregnancy period. As the patient had no symptoms of LPD and progressive lesion, there was no need for further laparoscopic or laparotomic examination. As conclusion LPD should be considered in the differential diagnosis of multiple peritoneal nodules especially in women of reproductive age. We found no data about LPD and preeclampsia togetherness in the literature. In our

case; we tought that preeclampsia was co-incident, but the patient's first cesarean delivery and laparoscopy records suggest that there were nothing about LPD. It is interesting that LPD developped in this hypertensive-preeclamptic pregnancy period after one pregnancy and two surgical procedures with no-use of hormonal therapy or contraceptive methods.

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