

Leiomyomatosis peritonealis disseminata: An exceptional case

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Abstract

Leiomyomatosis peritonealis disseminata is a rare clinical disorder characterised by proliferation of smooth muscles spread all over the peritoneal cavity. History and examination do not help in establishing a proper diagnosis, which is possible only by histopathology. Although LPD is a benign disease which regresses spontaneously, it may degenerate to malignancy, so proper follow up is mandatory. A 38-year-old lady presented with lower abdominal pain on 27th day of her menstrual cycle. Due to suspicion of ovarian cyst accident, laparotomy was performed during which innumerable nodules were found scattered all over the peritoneal cavity. Histopathology revealed leiomyomatosis peritonealis disseminata. The patient was followed up for two years without any treatment and she remained asymptomatic.

Keyword: Leiomyomatosis peritonealis disseminata, Benign, Carcinomatosis, Nodules, Fibroma.

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Introduction

Leiomyomatosis peritonealis disseminata (LPD) is typically a benign and rare disorder found in females during their reproductive age, although some cases have been reported in postmenopausal females and males.^{1,2} LPD is characterised by the proliferation of smooth muscles leading to gross nodularity all over the peritoneal cavity.³ Although high circulating oestrogen and progesterone levels play a significant role, exact etiopathogenesis is not clear along with its definitive treatment strategy.¹ The condition is found incidentally during surgery or on imaging in asymptomatic patients. LPD may degenerate to malignancy and may mimic carcinomatosis.³ To our knowledge, this is the first case reported in Pakistan.

Case Report

A 38-year-old woman presented in the emergency department with history of acute abdominal pain for the

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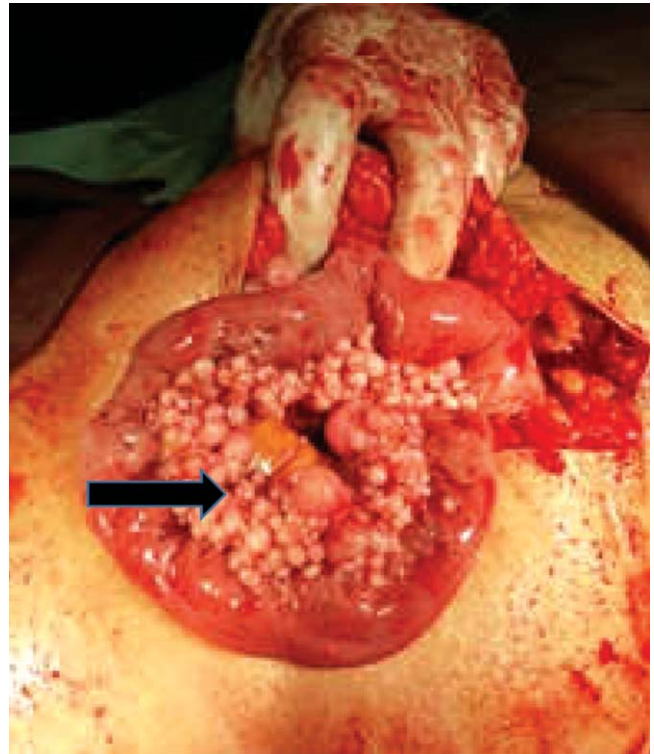


Figure-1: Numerous Nodules in the mesentery of small intestine.

last two days which started on 27th day of her menstrual cycle. She had four children, all delivered vaginally and the youngest child was seven years old. She had never taken any contraceptive pills or any other type of hormone replacement therapy. On examination, the abdomen was soft; tenderness and rebound tenderness was positive in the left iliac fossa. Her routine blood tests were normal and pregnancy test was negative. Ultrasound revealed ovarian cyst of 5×5 cm with mild free fluid, due to which haemorrhage or torsion was suspected. She underwent laparotomy, which revealed that the peritoneal cavity was full of innumerable nodules ranging from few millimetres to few centimetres. These nodules were along the mesentery of small gut, large gut, posterior abdominal wall and under the surface of diaphragm, while the whole pelvis was occupied with nodularity. It was difficult to identify uterus, ovaries and tubes separately (Figure-1 and 2). One of the nodules 3×4 cm from the abdominal wall was taken for biopsy.

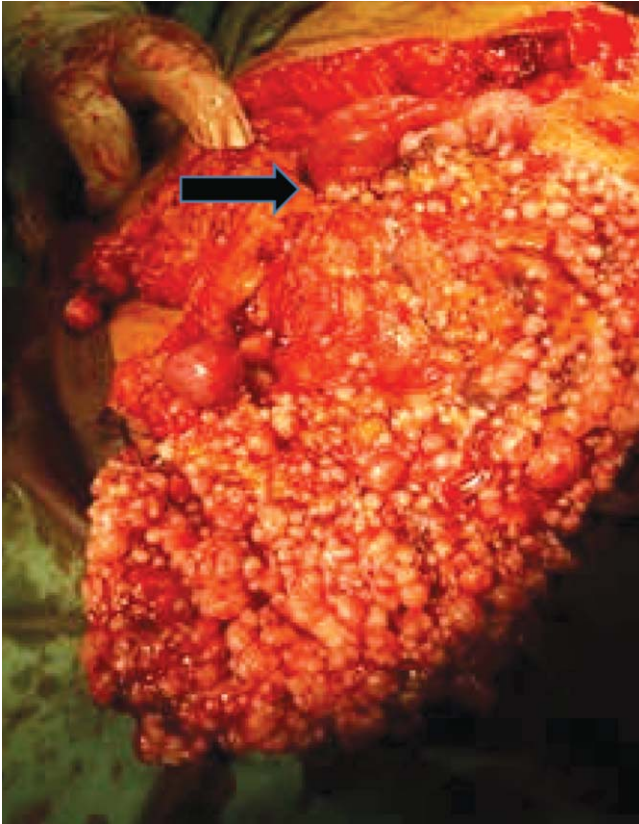


Figure-2: Numerous Nodules in the mesentery.

Histopathology revealed the proliferation of smooth muscles interdigitating fibres with no nuclear atypia. During follow up the patient remained asymptomatic without any treatment for a period of two years.

Discussion

Leiomyomatosis peritonealis disseminata is an extremely rare clinical condition that was first described by Wilson and Peale in 1952,⁴ it was later designated as LPD by Taubert in 1965.⁵ To date, 165 cases have been reported but no criteria for its diagnosis and management has been proposed yet. In Pakistan, it has never been reported previously, probably because of asymptomatic nature of the disease. However, it does not always remain stationary because malignant transformation has also been reported.⁶ LPD mainly presents in women of reproductive age but can occur in postmenopausal women and men.

The aetiology and pathophysiology of LPD is not yet certain,⁷ however, LPD originates from metaplasia of submesothelial, multi potential mesenchymal cells.⁸ Travassoli and Norris hypothesised that unusual and selective sensitivity of subperitoneal mesenchymal stem cells to metaplasia is probably promoted by hormonal

stimulation.⁹ The reason may be high levels of oestrogen and progesterone, which could be endogenous or exogenous oestrogens like prolonged use of contraceptive pills or repeated pregnancies. LPD is reported after laparoscope-assisted myomectomy that may indicate a common pathogenesis between myoma and LPD.¹⁰ Khangar reported the first ever LPD with ovarian leiomyoma.¹¹ Halama suggested that it may be autosomal dominant and may be associated with ascites and endometriosis.

Clinical presentation is non-specific; it may present with abdominal pain, discomfort, bleeding from vagina or rectum or symptoms of intestinal obstruction.⁵

Proper diagnosis is difficult if based on history alone. Computed tomography (CT) and magnetic resonant imaging (MRI) may be beneficial but definitive diagnosis can only be made through histopathology. Good quality ultrasound might be beneficial in making a differential diagnosis. Microscopically LPD is typically characterised by smooth muscle cells proliferation with or without mitosis. However, cytological atypia, tumour cell necrosis and increased mitotic figures are not the features of LPD. In LPD, tumour, both ER and PR receptors are identified.

To date, clinical consensus in the management of LPD has not been established. One option is surgical removal of the tumour if clinically and surgically possible, in cases where small and few nodules are present, along with oophorectomy to reduce oestrogens exposure. Iatrogenic menopause with gonadotrophin releasing analogues¹² or aromatase inhibitors may be effective.¹³ Treatment modalities should be devised according to the patient's age, comorbidities and severity of symptoms. Women who wish to remain fertile should be offered conservative treatment and advice to stop contraceptive pills. Aggressive surgical treatments can be offered to those patients who are at risk of developing malignancy like those who have prolonged exposure to contraceptive pills, history of myomectomy with positive ER and PR receptors in their benign nodules.

Conclusion

LPD is a rare benign disorder prevalent in women of reproductive age with possibility of malignant transformation. Proper diagnosis and follow up must be ensured so that its malignant counterpart is not missed.

Consent: A verbal consent was obtained from the patient regarding the publication of her case because it can be helpful in further research.

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Conflict of Interest: None.

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