



## Leiomyomatosis Peritonealis Disseminata Mimics As A Metastatic Carcinoma: A Diagnostic Dilemma in Pregnancy

### KEYWORDS

leiomyomatosis peritonealis disseminate, endomyometriosis, metaplasia of subcoelomic mesenchyme

#### Dr. Vina Kumari

M.D. Department of Obs & Gyn,  
Tata main Hospital, Jamshedpur

#### Dr.J.Sreedevi

Dept.of pathology, Tata main  
Hospital, Jamshedpur

#### Dr. Manas Kabiraj

M.D. Department of Obs & Gyn,  
Tata main Hospital, Jamshedpur

#### Dr.Radhika Narayan

Dept.of pathology, Tata main Hospital, Jamshedpur

#### Dr.Minakshi Mishra

Dept.of pathology, Tata main Hospital, Jamshedpur

#### Dr.R.Mohanty

Dept.of pathology, Tata main Hospital, Jamshedpur

#### Dr.Farah

Dept.of pathology, Tata main Hospital, Jamshedpur

### ABSTRACT

*Leiomyomatosis peritonealis disseminata (LPD) is a rare benign disease presenting as multiple peritoneal nodules of smooth muscle cells, mimicking peritoneal carcinomatosis. LPD is most often found in reproductive and pre/post-menopausal age group. Herein, we report a case of LPD coexisting with endomyometriosis in pregnant woman and review the pathogenesis of this lesion. A 34-year old woman, G4P1L1A2, 19 weeks pregnancy with complex uterine & adnexal mass and was advised for termination of pregnancy. However she wished to continue the pregnancy and underwent LSCS at 36 weeks. Histopathological examination (HPE) revealed of LPD. IHC showed positivity for SMA and Desmin, along with CD10 which highlighted the smooth muscle and endometrial stromal cells, consistent with LPD with endomyometriosis. A confirmatory HPE report can help recognize this uncommon pathological entity and vastly improve patient care in such cases.*

### Introduction:

LPD is a rare benign disease of unknown etiology in women of reproductive age.<sup>[1]</sup> A possible origin from submesothelial multipotential cells, although it is not clear if the stimulus to smooth cell differentiation is hormonal, genetic or both.<sup>[1,2]</sup> The few reported cases of association between LPD and endometriosis favour origin for both the lesions.<sup>[3]</sup> Most LPD cases are benign and rarely undergo malignant transformation.<sup>[4-5]</sup>

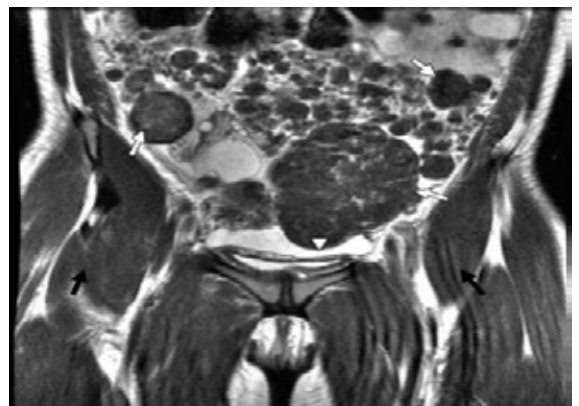
Since then, less than 150 cases of LPD have been reported out of which only six cases with coexistence of endometriosis within LPD lesions have been reported.<sup>[6-7]</sup> Herein, we report another case of LPD coexisting with endomyometriosis within the same lesions.

### Case History:

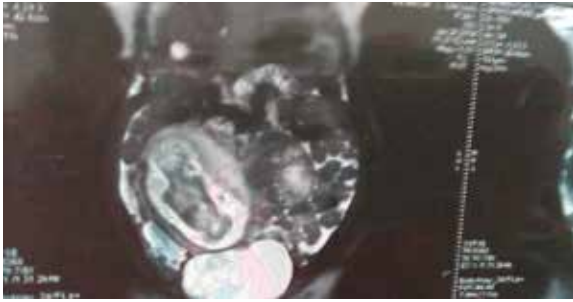
A 34 yrs old G4P1L1A2 presented at 19 weeks gestation with short history of bleeding per vagina. On obstetric history, one previous LSCS was done for premature rupture of membranes, with 2 spontaneous 1<sup>st</sup> trimester abortions. She had high blood pressure during her previous pregnancy. No H/o malignancy in family.

In present pregnancy she had hypertension in 1<sup>st</sup> trimester & on medication. Apart from that she received Inj HCG 5000 IU im per week x 5 doses, micronised progesterone 200mg OD for 2 months and Tab Aspirin x 5 doses, micronised progesterone 200mg OD for 2 months and Tab Aspirin 75mg OD for 1 month for past H/O 2 spontaneous abortions. USG scan report showed the single intrauterine fetus with both ovaries showing cysts and there was a fibroid of 9x 12 mm in posterior wall of uterus. Foetus appeared normal on Anomaly scan. One episode of bleed-

ing per vagina in second trimester, she again advised for USG whole abdomen which showed multiple rounded hypoechoic masses seen throughout abdomen with ascites, uterine scar showed nodular hypoechoic mass 62x 27 mm. MRI Abdomen & pelvis showed the Intrauterine pregnancy with 7 cm size left ovarian mass with solid+cystic components with complex enhancing septa, nodular enhancing areas inseparable from the body of the uterus with multiple rounded enhancing nodules over omentum, mesentery, POD, peritoneal surfaces & in Morrison's pouch with mild ascites[Fig1]. The final impression was Intrauterine pregnancy with left ovarian malignancy with peritoneal metastasis & ascites.



**Fig 1:** MRI Report shows characteristic isointensity with skeletal muscle on T1 and T2 weighted images



**Fig 2 :** Contrast enhanced MRI showed single intrauterine foetus with homogenous Uptake in late phase

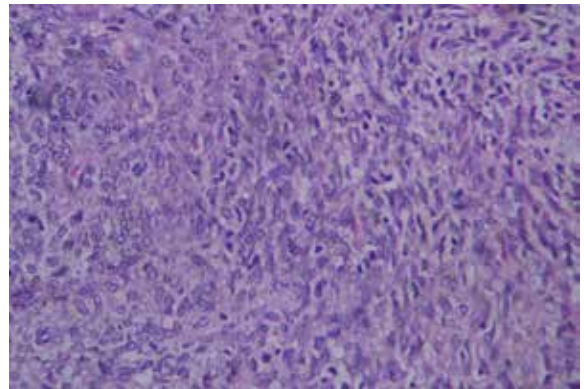
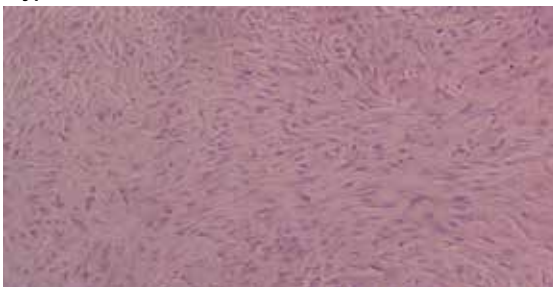
FNAC from abdominal mass was done, which suggested the adenocarcinoma NOS. USG Guided biopsy of peritoneal deposit showed dense fibrocollagenous tissue only. Hence she was advised for termination of pregnancy. But the Patient wished to continue her pregnancy, so she consulted a higher center where tumor marker analysis was done. All of them were within normal limits except raised CA 125. (CA 19.9: <3.0, CEA : 0.52, **CA125 : 236.8**). and also Trucut biopsy report showed the smooth muscle cells only. There was no evidence of malignancy seen. Patient had continued her pregnancy & followed up with routine antenatal check-up. She was planned for elective caesarian section in view of previous caesarean section for chronic hypertension. Patient underwent LSCS, she delivered a live female baby & intraoperative findings showed entire peritoneum, bladder & bowel surface, anterior surface of uterus were studded with large tumour deposits [Fig 3].



**Fig 3:** Intraoperative photograph show multiple tumour deposits on peritoneal, bladder, and bowel surfaces and anterior surface of uterus.

Biopsy had taken from multiple peritoneal deposits. Microscopy showed mostly benign smooth muscle cells arranged in bundles and fascicles. Nuclear atypia, mitoses and necrosis were not seen [Fig 4,5].

**Fig 4 :** H&E showed uniform oval to spindle shaped cells arranged in bundles & fascicles without nuclear atypia/mitosis.



**Fig 5 :** H&E (400X) Showed at focal places endometrial stromal cells without gland formation

So diagnosis of LPD was made. IHC on the biopsy sections showed smooth cells positive for SMA and Desmin, along with CD10 positivity which highlighted the endometrial stromal cells. Overall appearances were consistent with a diagnosis of Endomyometriosis. Post operative period was uneventful.

#### Follow up

Followed up with USG report which showed bulky uterus with fibroid in posterior wall measuring 7x11mm, ovaries adherent to bowel loops, rounded nodular lesion seen in POD, mild ascites upto renal angle. After one month, CT Scan report showed the normal Uterus & ovaries, follicles in both ovaries, tiny enhancing peritoneal deposits in pelvis, minimal ascites, cystic lesion in posterior uterine wall. There was significant reduction in size of lesions as compared to previous sonography report. Patient was comfortable, asymptomatic and had not received any treatment during lactation & postpartum period. She was kept on follow up since the size of her tumor is regressing.

#### Discussion:

First case was reported in 1952 by Wilson and Peale,<sup>[8]</sup> LPD was first clearly delineated and named by Taubert et al in 1965.<sup>[9]</sup> Two theories are proposed to explain the etiology of the LPD (a) mullerianosis – misplaced endometrial/mullerian tissue are possible explanation for the extrauterine uterus-like mass, as a developmental entity.<sup>[10]</sup> (b) a metaplasia of subcoelomic mesenchyme. The condition is associated with high levels of exogenous and endogenous female gonadal steroids (e.g. pregnancy, prolonged exposure to oral contraceptives and/or combined hormonal replacement therapy, granulosa cell tumours of the ovary) indicating that oestrogens and progestins play an important role in the pathogenesis of LPD.<sup>[11]</sup> Genetic analysis suggests its origin from peritoneal implantation of single uterine leiomyoma and subsequent proliferation. This might be the etiology in our patient because her first trimester USG report showed the presence of a fibroid. Multiple reports of LPD revealed laproscopic myomectomy where morcellator has been used was being the genesis of lesion. There was no evidence of urogenital abnormality in our patient, so a mullerian duct fusion defect was unlikely. Malignant conditions that mimic LPD are peritoneal carcinoma, Metastatic gastrointestinal & ovarian cancer, Mesothelioma. Peritoneal carcinomatosis, however, is often associated with tumour cake, ascites and liver metastases, which have not been reported with LPD. The ascites in our patient could be due to chronic hypertension associated with pregnancy.

In present study, there was marked discordance between radiological and pathological findings during antenatal period which patient had difficulty in taking a decision regarding continuation of pregnancy due to complex uterine and adnexal mass, effect of pregnancy on disease, risk of patients life for baby. Eventually she delivered a healthy baby & exact characterisation of lesion was done with support of IHC.

### Conclusion

LPD is the potential misdiagnosis of disseminated malignancy, Low incidence & possible unfamiliarity to the medical community. To sum up, the present case reinforces endometriosis with LPD in pregnant woman as a distinct clinicopathological entity & exemplifies diagnostic challenge associated with it and necessitates application of various IHC markers for its correct diagnosis.

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