Leiomyomatosis Peritonealis Disseminata (LPD) and Meningioma: A Rare Association

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Received: 26 June 2023

Accepted: 17 August 2023

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DOI 10.5001/omj.2025.20

Abstract

A 39 year old, multiparous, with complains of dyspepsia and bloatedness 5 years back was diagnosed as Disseminated Peritoneal Leiomyomatosis on MRI . Her antenatal period was uneventful. She underwent Caesarean section for pathological CTG, biopsy taken from the peritoneal mass confirmed the diagnosis. Postoperatively, she was admitted with Status Epilepticus. CT brain with angiography revealed a right frontal meningioma. With increasing ascites, proceeded with Excision of the tumour and Total Abdominal Hysterectomy with Bilateral salpingectomy.

Keywords: Leiomyomatosis peritonealis disseminata, disseminated peritoneal leiomyomatosis, Meningioma,seizure,status epilepticus

Introduction

Leiomyomatosis peritonealis disseminata is a rare benign gynecological disorder characterized by the dissemination of multiple smooth muscle nodules throughout the peritoneum. It was first described in 1952 by Wilson and Peale[1] and term coined by Taubert in 1965.[2] The pathogenesis is poorly understood hence proper management and prognosis have not been investigated very well. An association between hormones and meningioma risk has been suggested by the increased incidence of post-pubertal disease in women versus men (2:1) with the highest ratio of 3.15:1 during the peak reproductive years, the presence of estrogen, progesterone, and androgen receptors on some meningiomas.[3,4] indicated by the increase in size of the meningioma during the luteal phase of the menstrual cycle, pregnancy and exogenous hormones,[5] and the regression of multiple meningiomas in a patient following cessation of estrogen and progesterone agonist therapy.

Case Report

A 39 year old, multiparous, with complains of dyspepsia and bloatedness 5 years back was diagnosed as Disseminated peritoneal leiomyomatosis on MRI [Figure 1] not willing for further evaluation and treatment. In 2022, she was seen during the antenatal period which was uneventful. No significant medical or surgical history. She underwent Caesarean section for pathological CTG, during which the biopsy from the peritoneal mass was taken which confirmed the diagnosis of Disseminated peritoneal leiomyomatosis. On post operative day 6, she was admitted to the intensive care unit with status epilepticus, with inotropic support. She was under the care of the Multidisciplinary Team (ICU Anesthetists, Obstetrician, Neurosurgeon, General physician, Gyne-oncologist). Her EEG revealed focal epileptiform activity the right fronto-temporo-parietal region for which she was on antiepileptic medications. CT brain angiography [Figure 2] and MRI brain with Gadolinium revealed a right frontal meningioma which did not warrant an immediate intervention as patient was critical. She developed fever, on evaluation, caesarean skin wound swab showed growth of E. coli (E.S.B.L.). Her initial Aerobic and Anerobic blood cultures, High vaginal Swab, Urine culture showed no growth. Sputum culture showed MDRO Scanty

growth of Pseudomonas Aeruginosa and Acinetobacter Baumanii. She was screened for Brucella, Coxiella burnettii, Tuberculosis, TORCH, HIV, Viral and fungal culture were negative. On admission, CT chest, abdomen and pelvis done reported as left lung consolidation/collapse without pleural effusion. Mild ascites. After 1 week, CT revealed left lung consolidation/collapse with left pleural effusion. No obvious Pulmonary embolism. Increasing ascites present. Ultrasound chest revealed moderate amount of left sided pleural effusion with atelectatic left lower lobe. Echo normal. She remained febrile and there was a rising trend in her inflammatory markers inspite of the multiple parenteral antibiotics. Patient developed ascites and abdominal distension and ascitic tapping under ultrasound guidance. The ascitic fluid dint reveal bacterial growth.

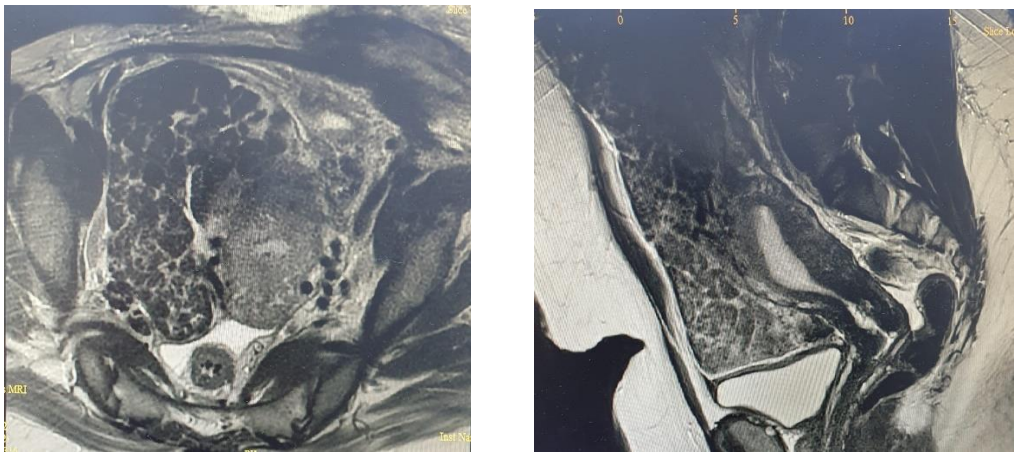

Figure 1: MRI abdomen with gadolinium.

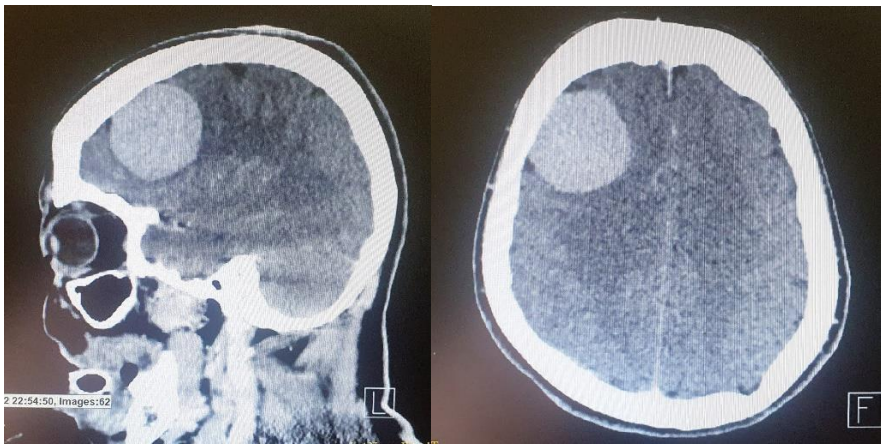

Figure 2: CT brain with angiography.

As no improvement in the clinical condition of the patient and increasing ascites, proceeded with Excision of the tumour and Total Abdominal Hysterectomy with bilateral salpingectomy under ventilator support. Uterus of 12 weeks size with subserosal fibroid extending to involve the entire peritoneum, bladder serosa and omentum. The mass weighed 8kg. Ascitic fluid culture and sensitivity showed no growth. Histopathology showed diffuse benign leiomyomatosis with signs of degeneration with no evidence of infection or atypia [Figure 3 and 4]. A intraperitoneal drain was placed.

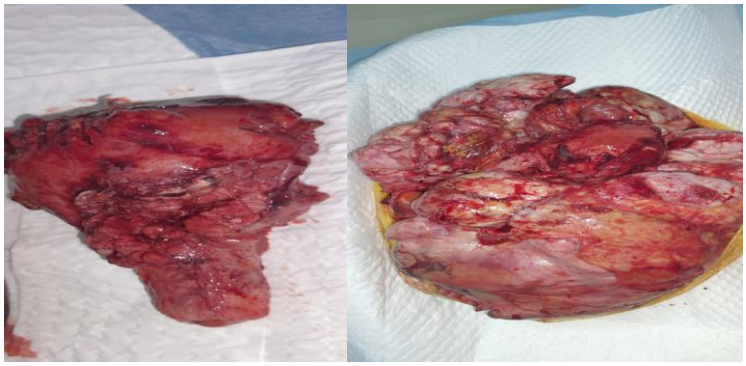

Figure 3: Hysterectomy specimen with the peritoneal leiomyomatosis.

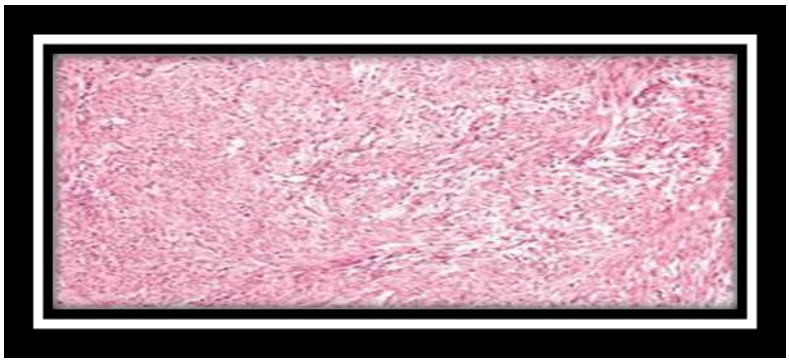

Figure 4: Histology.

Postoperatively, there was initial improvement in the fever and inflammatory markers but later stared to rise. Patient could not be extubated was on inotropic support. On postoperative day 7, CT abdomen, pelvis and chest showed newly developed pelvic abscess collection with air. EEG revealed intermittent Right frontotemporal parietal epileptiform discharges. Head CT repeated Stable right frontal extraaxial lesion representing meningioma. No evidence of abnormal meningeal enhancement.

CT guided abscess drained from pelvis sent for culture reported as MDRO, CRE (Carbapenem Resistant Enterobacteriaceae) Klebsiella pneumoniae(scanty growth).

On postoperative day 10, patient had high grade fever, hypotension on high inotropic support, she arrested CPR done and revived. ABG revealed metabolic acidosis Ph 6.9, high potassium and lactate for which correction given.

Patient arrested again and CPR continued as per ACLS protocol but could be revived.

Discussion

LPD could be caused by metaplasia of mesenchymal cells of the peritoneum and, in susceptible women, residual myoma in the abdominal cavity postoperatively might contribute to the development of LPD.[6,7] Metaplasia and differentiation from mesenchymal stem cells into smooth muscle cells may be promoted by estrogen exposure.[8] Therefore, LPD is often considered a premenopausal benign disease. An association of multiple intracranial meningioma presentation with long standing use of megestrol acetate, a progesterone agonist which was confirmed histologically by the presence of progesterone receptors on the largest tumor. Regression of the tumor was noted with discontinuation of the medication.[8] Studies have shown a stronger association with PR than with ER status.[3]

The PR status seemed to be associated with changes near the *NF2* gene on 22q continues to suggest that hormones are likely to play an important role in either the development or progression of some meningiomas.[3]

The predominance of meningiomas in females and their accelerated growth during the luteal phase of the menstrual cycle and during has led to a number of studies examining the potential role of steroids on the growth of meningiomas. The presence of estrogen receptors(ER), ER-α and ER-β on meningiomas were identified, using reverse transcription and polymerase chain reaction (RT-PCR) Southern blot analysis.[4] A retrospective cohort study conducted by Yen Y et al concluded that women with uterine myoma was at a significantly higher risk of developing meningioma (45%) than among women without uterine myoma.[9] The expression of the PR alone in meningiomas signals a favorable clinical and biological outcome. In female patients, sex hormone receptor status should routinely be studied for its prognostic value, and should be taken into account in tumor grading. The initial receptor status of a tumor may change in progression or recurrence of tumor.[10]

Intracranial leiomyoma may be primary or secondary presentation. Various cases of histology, immunohistochemistry and electron microscopy.[11,12] Allessi et al reported cases of benign metastasizing leiomyoma to skull base and spine.[13]

Conclusion

MRI can establish with a high degree of confidence the definitive diagnosis of diffuse leiomyomatosis which might allow the patient management by a multi- disciplinary team for therapeutic decision-making process. Surgery is the main stay of treatment. Meningioma has an association with disseminated peritoneal leiomyomatosis. Although rare, leiomyoma should be considered in the differential diagnosis of well-circumscribed intracranial lesion.

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