Leiomyomatosis Peritonealis Disseminata and Meningioma: A Rare Association

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Abstract

A 39-year-old multiparous woman underwent emergency caesarean section after pathological findings in her cardiotocograph. Biopsy taken from the peritoneal mass confirmed the diagnosis of disseminated peritoneal leiomyomatosis (LPD). Post-operatively the patient developed status epilepticus. Computed tomography (CT) image of brain with angiography revealed a right frontal meningioma. With increasing ascites, excision of the tumor and total abdominal hysterectomy with bilateral salpingectomy were performed.

Keywords: Leiomyomatosis Peritonealis Disseminata; Meningioma; Seizure; Status Epilepticus; Oman

Introduction

Leiomyomatosis peritonealis disseminata (LPD), also called disseminated peritoneal leiomyomatosis, 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 was coined by Taubert in 1965.[2] The pathogenesis of LPD is poorly understood, and its management and prognosis have not been investigated adequately.

Meningioma is a tumor formed in the meninges. An association between hormones and meningioma risk has been suggested as it occurs in post-pubertal women twice as much as in men. The female-male ratio of meningioma patients rises to 3.15:1 during the peak reproductive years. The presence of estrogen, progesterone, and androgen receptors on some meningiomas also points to the hormonal association.[3,4] Other pointers include a tendency for the tumors to increase in size during the luteal phase of the menstrual cycle, pregnancy and exogenous hormones,[5] and the regression of multiple meningiomas following cessation of estrogen and progesterone agonist therapy.

We present a rare case of association of LPD and meningioma in a female patient.

Case Report

A 39-year-old multiparous woman underwent an emergency Caesarean delivery after an antenatal cardiotocography (CTG) revealed a fetal pathology. Biopsy from the peritoneal mass confirmed a diagnosis of LPD. The same diagnosis had been made five years earlier by magnetic resonance imaging (MRI) [Figure 1] for which she had refused further evaluation or treatment. The patient had no other significant medical or surgical history.

Six days after the caesarean section, the patient developed status epilepticus and was admitted to the intensive care unit (ICU) with inotropic support, under the care of a multidisciplinary team comprising ICU anesthetists, an obstetrician, a neurosurgeon, a general physician, and a gynecologic oncologist. Her electroencephalogram (EEG)

revealed focal epileptiform activity in the right fronto-temporo-parietal region for which she was put on antiepileptic medications. CT brain angiography [Figure 2] and MRI brain with gadolinium revealed a right frontal meningioma, but an immediate intervention was ruled out in view of her critical condition. She developed fever, and a swab culture from the caesarean skin wound showed growth of *Escherichia coli* with extended-spectrum beta-lactamases (ESBL). Her initial aerobic and anerobic blood cultures, high vaginal swab, and urine culture showed no growth. Sputum culture revealed scanty growth of multidrug-resistant (MDRO) *Pseudomonas aeruginosa* and *Acinetobacter baumannii*. Tests yielded negative for Brucellae, *Coxiella burnetii*, tuberculosis pathogen, TORCH pathogens, human immunodeficiency virus (HIV), and other viral and fungal pathogens. CT of chest, abdomen, and pelvis revealed left lung consolidation/collapse without pleural effusion. Mild ascites was present. One week later, repeat CT revealed left lung consolidation/collapse in addition to left pleural effusion, with no obvious pulmonary embolism. The ascites was increasing. The ultrasound image of the chest revealed moderate left-sided pleural effusion with atelectatic left lower lobe. Echocardiography was normal.

Despite being on multiple parenteral antibiotics, the patient remained febrile with rising inflammatory markers. The ascites had resulted in abdominal distension; ascitic tapping was done under ultrasound guidance. The ascitic fluid did not reveal any bacterial growth.

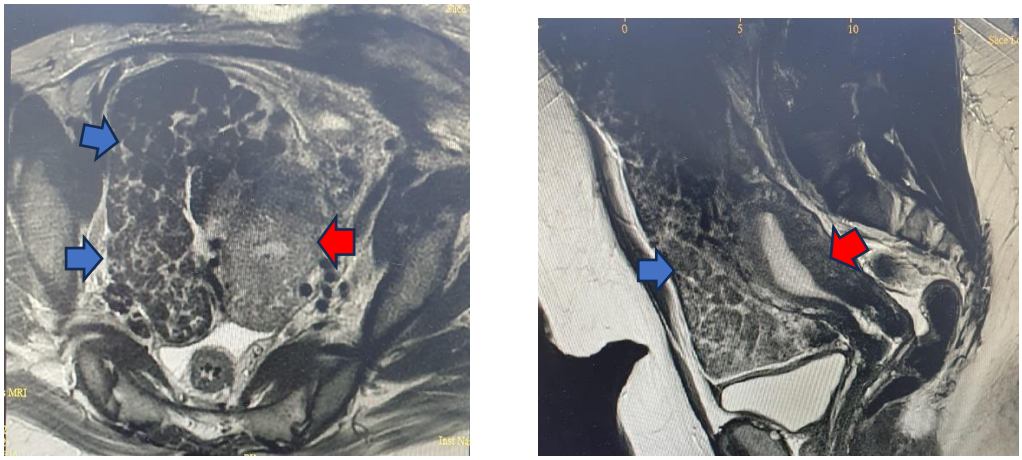

Figure 1: Magnetic resonance images (MRI) of abdomen using gadolinium-based contrast media. Left: Axial view. Right: Sagittal view. (Red arrow: uterus; Blue arrow: leiomyomatosis peritonealis disseminata.)

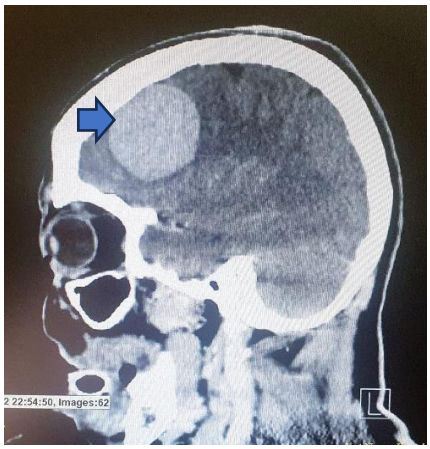

Figure 2: Computed tomography scan of sagittal view of the brain showing the meningioma.

With no improvement in the clinical condition of the patient and increasing ascites, we proceeded with excision of the tumor and total abdominal hysterectomy with bilateral salpingectomy under ventilator support. The excised uterus had the size of 12 weeks gestation, and subserosal fibroid had extended into the entire peritoneum, bladder serosa, and omentum. The mass weighed 8 kg. 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.

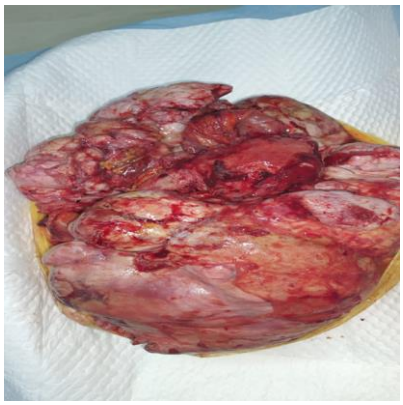

Figure 3: Hysterectomy specimen with the peritoneal leiomyomatosis involving peritoneum and omentum.

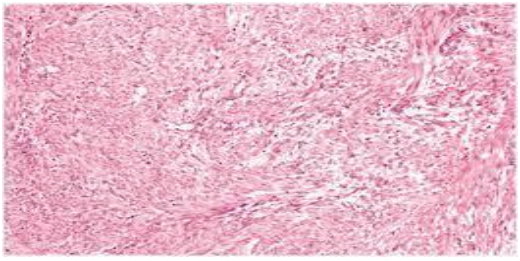

Figure 4: Histopathology: diffuse benign leiomyomatosis with signs of degeneration, with no evidence of infection or atypia.

Postoperatively, there was an initial lowering in the fever and inflammatory markers, but they rose again later. The patient could not be extubated as she was on inotropic support. CT scans of abdomen, pelvis, and chest conducted on postoperative day 7 showed newly developed pelvic abscess collection with air. EEG revealed intermittent right frontotemporal parietal epileptiform discharges. Repeat CT of the brain revealed a stable right frontal extraaxial lesion representing meningioma with no evidence of abnormal meningeal enhancement. The abscess in the pelvis was drained under CT guidance and its culture revealed scanty growth of MDRO carbapenem resistant *Klebsiella pneumoniae*.

On postoperative day 10, the patient had high grade fever, hypotension on high inotropic support, and suffered cardiac arrest. She was revived by cardiopulmonary resuscitation (CPR). ABG revealed metabolic acidosis (pH 6.9), high potassium, and lactate which were corrected. The patient had another cardiac arrest. Though CPR was given as per advanced cardiovascular life support (ACLS) protocol, she could not 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 its development.[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 benign premenopausal 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 progesterone receptor (PR) status than with estrogen receptor (ER) status.[3]

The PR status seemed to be associated with changes near the *NF2* gene on 22q, suggesting 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 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 found women with

uterine myoma to be at a significantly higher risk of developing meningioma (45%) than those 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 considered in tumor grading. The initial receptor status of a tumor may change with the progression or recurrence of the tumor.[10]

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

Conclusion

MRI can establish with a high degree of confidence the definitive diagnosis of diffuse leiomyomatosis. Meningioma is rarely associated with leiomyomatosis peritonealis disseminata, as in the current case. Surgery is the mainstay of treatment. Management by a multidisciplinary team is recommended.

Disclosure

The authors declare no conflicts of interest. Informed consent was obtained from the patient's husband.

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