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CASE REPORT

Uterine Intravenous Leiomyomatosis with Intracardiac Extension and Pulmonary Benign Metastases

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ABSTRACT

Uterine intravenous leiomyomatosis is an uncommon benign tumour characterized by intravascular proliferation of a histologically benign smooth muscle cell tumour.This benign pathological tumour may permeate into the venous system causing intravenous leiomyomatosis of the uterus (IVL). IVL may extend along the pelvic veins to the inferior vena cava, occasionally reach the right cardiac chambers and the main pulmonary artery and producing benign pulmonary metastases and cause signs of right sided congestive heart failure and sudden death. Herein, we describe an unusual case of uterine intravenous leiomyomatosis with intracardiac extension and pulmonary benign metastases. We should suspect intravenous leiomyomatosisin premenopausal women with known history of uterine leiomyoma, presenting with cardiovascular symptoms and evidence of a free-floating mass within the right cardiac chambers. Imaging plays important role in defining its anatomical origin and relations. Urgent surgical treatment with radical excision or embolization of pelvic and intravenous leiomyomatosis yields favourable outcomes and excellent prognosis.

Keywords: Intravenous leiomyomatosis, Intracardiac leiomyomatosis, Uterine leiomyoma, Pulmonary metastases.

INTRODUCTION

leiomyomas accounts for the leading indication for hysterectomy worldwide. Uterine leiomyomas derives from a benign monoclonal proliferation of smooth muscle cells of the uterine myometrium. Intravenous leiomyomatosis is a separate pathological entity which shows growth of benign leiomyoma tissue within vascular wall[1]. The pathological mechanism of the Intravenous leiomyomatosis was not clear, and currently 2 explanations were existing. One is the Knauer theory, which believed IVL originated from vascular wall[2]. The other is the Sitzenfry theory, which believed IVL occurred when uterine leiomyoma invading blood vessels[3]. First cases were reported by Birsh-Hirshfeld in 1896 and Knauer in 1903. Fewer than 300 cases have been reported in the medical literature, with about 100 cases reported with direct intracardiac tumorextension[4]. The lesion is characterized by the abnormal growth of a benign smooth muscle cell tumor arising from the uterine venous system, which may subsequently propagate further to greater caliber veins and occasionally reach the cardiac chambers with pulmonary benign metastases. Althoughhistopathologicallybenign[5], this type of neoplasm has highly aggressive component and tendency to invade the venous system.

Herein, we describe the case of a nulliparous woman in premenopausal age, presenting with signs and symptoms of right sided congestive heart failure and a previous history of uterine fibroids, providing a review of the literature with regards to intravenous leiomyomatosis of the uterus with cardiac and pulmonary involvement.

CASE DESCRIPTION

A 32-year-old nulligravida, nulliparous woman presented to Radiology department on 13th February 2020 for USG abdomen and pelvis for abdominal pain and vaginal bleeding. USG showed Bulky uterus with heterogenous echotexture with multiple fibroids with thickened endometrium. Hysteroscopy with HPE done which showed benign findings and then Mirena IUCD insertion done. On 7th October 2020, Patient presented with complaints of mild chest discomfort and underwent a two-dimensional Doppler transthoracic echocardiography, which showed mobile mass in right atrium protruding into right ventricle. Followed by cardiac MRI was performed, it showed an irregular mobile T2 hypo and T1 iso to hypointense lesion measuring 5.5 x 1.55 cms seen in right atrium extending into IVC and protruding into right ventricle through tricuspid valve. Diagnosis given as Right Atrial Thrombus extending into IVC and protruding into RV (figure 1).

The patient again presented on 17th September 2021 for USG abdomen and pelvis for abdominal swelling, pain and vaginal bleeding. USG showed Large well defined lobulated heterogenous hypoechoic mass lesion of ~size 18 x 10 cm showing cystic areas within noted in pelvis predominantly on left side extending into left adnexa, pouch of douglas and minimal extension to right adnexa. The lesion is pushing the uterus towards right side with loss of endomyometrial interface. Engorged vessels noted in left parametrium. Superiorly the lesion is reaching upto left lumbar region (figure 2).

MRI Abdomen and pelvis was performed for the patient on 30th September 2021. It showed a large lobulated heterogeneously T2 hyperintense lesion with multiple dilated vessels within noted in lower abdomen and pelvis, measuring approximately 22 x 19 x 9.6 cms. Multiple tortuous dilated vessels noted within the lesion predominantly on left side with the dilated left common iliac and proximal internal iliac vein. Uterine body and fundus appear to be in continuity with the above mentioned lesion. The lesion extends from right iliac fossa to left iliac fossa, anteriorly extending to anterior abdominal wall and posterior to presacral region (figure 2). Diagnosis given as Uterine Leiomyomatosis.

Non contrast CT chest was performed as patient had cough and it showed multiple variable sized lobulated soft tissue nodules randomly distributed in lung parenchyma bilaterally with largest lesion in right upper lobe anterior segment measuring 11 x 10 mm (figure 3). Biopsy was performed from largest lesion in right upper lobe anterior segment (figure 13). Histopathology report came as Spindle cell neoplasm with no high-grade features and in view of multiple uterine fibroids, this can be viewed as Metastasizing leiomyoma (over) Leiomyosarcoma.

Patient underwent uterine artery embolization (figure4) and was on medical therapy. Post procedure MRI abdomen and pelvis was performed on 8th February 2022, it showed mild to moderate interval decrease in size of the lesion.

DISCUSSION

Intravenous leiomyomatosis is an uncommon, histologically benign neoplasm usually confined to the venous system of the uterus, a condition that only affects women, is histologically confirmed benign smooth muscle tumour within vascular spaces from intrauterine venules to the systemic veins including iliac vein, IVC, even extending into the right heart chamber. It can lead to right-sided heart failure and eventually death.

Intra-cardiac leiomyomatosis is a serious and advanced condition. If diagnosed early and surgically excised, permits a rapid recovery of right cardiac function and may have a good prognosis. IVL mainly involves premenoupasal women - although age is highly variable and ranges from 21 to 80 years old[6]. The clinical picture of patients presenting with intra-cardiac dissemination of IVL usually includes dyspnea, thoracic pain, syncope and lower-extremity edema, menometrorrhagia, abdominal pelvic pain and abdominal swelling.

We described the case of a 32-year-old woman affected by IVL with aintracaval-intracardiac mass in the right atrium and ventricle causing signs and symptoms of right cardiac failure. She presented to the Department of radiology of our hospital with the diagnosis of a free-floating atrial mass, initially mistaken for a thrombus in echocardiography. Then she underwent cardiac MRI and was accurately diagnosed IVL with cardiac involvement.

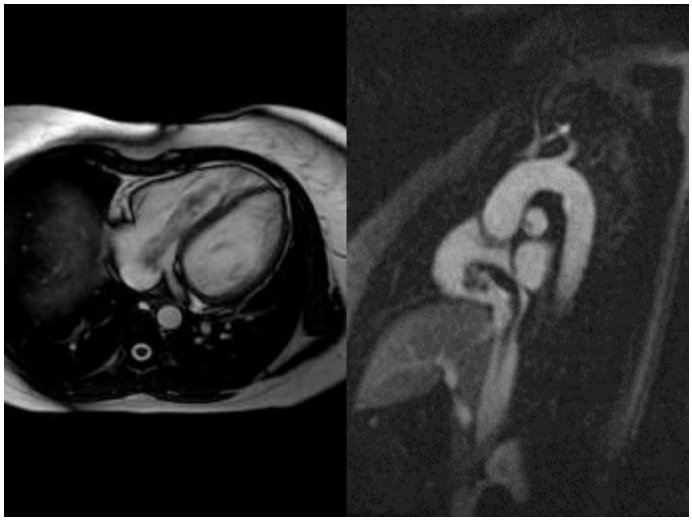

Figure 1: Non contrast and contrast enhanced cardiac MRI axial and sagittal plane showed thrombus in right atrium extending into IVC and protruding into right ventricle through tricuspid valve.

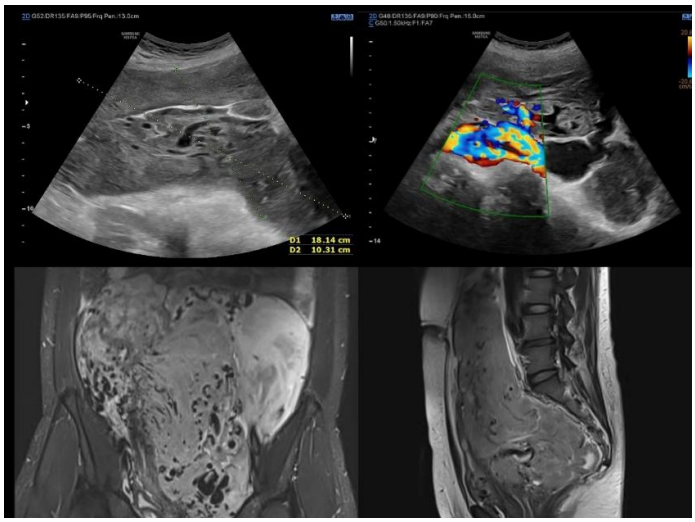

Figure 2: Transabdominal USG showed Large well defined lobulated heterogenous hypoechoic mass lesion with internal vascularity and MRI Abdomen and pelvis in sagittal T1 and coronal T2 fat saturation showed large lobulated heterogeneously T2 hyperintense lesion with multiple dilated vessels within noted in lower abdomen and pelvis.

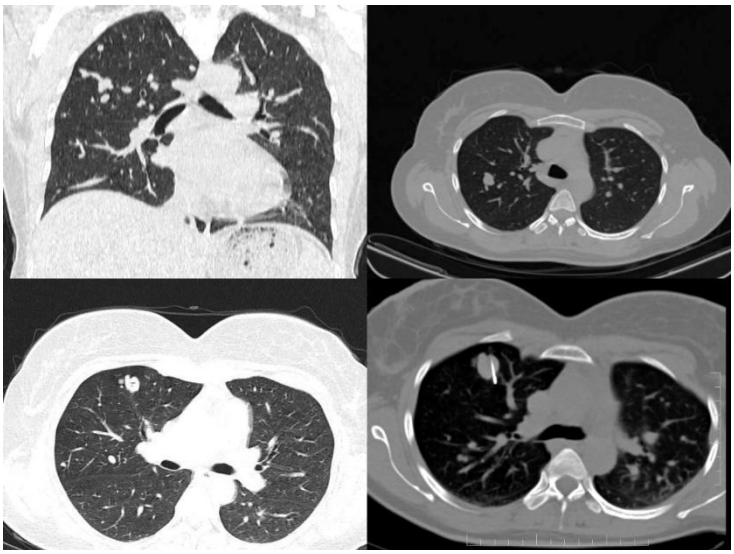

Figure 3: Non contrast CT chest axial lung and mediastinal window and coronal lung window showed multiple variable sized lobulated soft tissue

nodules randomly distributed in bilateral lung parenchyma and Non contrast CT chest axial mediastinal window showed right upper lobe anterior segment nodule with biopsy needle within.

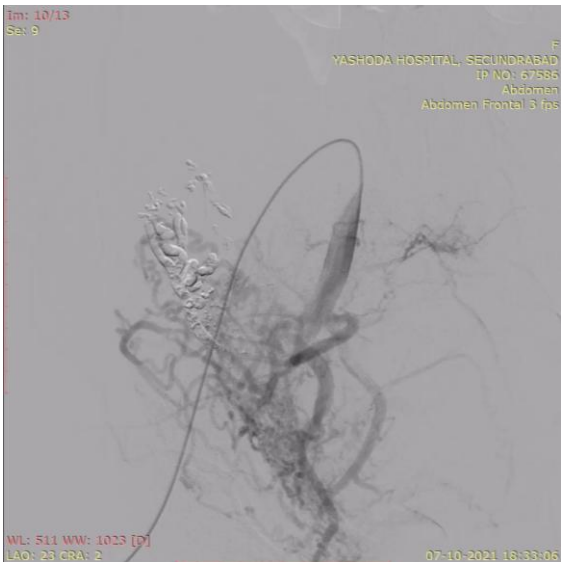

Figure 4: Selective internal iliac artery angiography showed contrast blush and followed by uterine artery embolization done.

CONCLUSION

In conclusion, in presence of signs and symptoms of right-sided heart failure with evidence of a free-floating right atrial mass at imaging, in a premenopausal or nulliparous woman with a history of uterine leiomyomas, intravenous leiomyomatosis should be suspected. Early surgical treatment with radical excision or embolization of pelvic and intravenous leiomyomatosis gives favorable outcomes and prognosis with low recurrence rates, as compared delayed diagnosis and treatment exposes to increased risk of congestive heart failure and sudden death.

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CONFLICT OF INTEREST

No Conflict of interest

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