

UTERINE FIBROIDS GOING INTO THE HEART: INTRA-VASCULAR AND INTRA-CARDIAC LEIOMYOMATOSIS: A VERY UNUSUAL PRESENTATION

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ABSTRACT

This case report is of a 50 years old female who presented with vague history of long term abdominal pain, shortness of breath and echocardiographic suspicion of right atrial mass. She was investigated using 128-slice Multidetector Computed tomography (MDCT) scanner in the Department of Radiology. Images of lower chest, entire abdomen and pelvis were taken in venous phase. On CT images of our patient, uterus was significantly enlarged and replaced by multiple contour deforming fibroids, which were involving the right adnexa, invading the right ovarian vessels, and extending into the right ovarian vein, inferior vena cava (IVC) and right atrium of heart. The findings were confirmed on surgery. Surgery also confirmed extension into right ventricle and pulmonary arteries i.e. pulmonary leiomyomatosis emboli. The histological findings were consistent with intravascular leiomyoma. MDCT images play instrumental role for preoperative morphologic assessment of IVL as it can easily identify the precise location, extent and provide a roadmap for the surgeons.

Key Words: Multidetector computed tomography (MDCT), Intravascular leiomyomatosis, Fibroids

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INTRODUCTION

Fibroids are benign smooth muscle neoplasms and common uterine tumors. Very rarely, they exhibit unusual growth pattern, like invasion of vessels called as intravascular leiomyomatosis (IVL)¹. The diagnosis of IVL is often overlooked². It is rare that fibroids invade into the lumen of veins³, which is characterized by fibroids proliferating within the vessel lumen without invading the vessel wall. When exposed to venous blood, they may reach the IVC, right cardiac chambers or pulmonary artery⁴. It is called intracardiac leiomyomatosis and can cause cardiac symptoms, even in some cases, sudden death. This is a very rare finding. Intravascular invasion of fibroids is thought to be due to their viscoelastic properties and hyaluronan component, which promotes invasion during pathogenesis⁵. CT scan is usually not the investigation for diagnosing uterine fibroids. Fibroids are usually seen incidentally on CT scans performed for other reasons. They are seen as bulky contour deforming uterus or a mass abutting or in continuity with the uterus. Fibroids can appear complex when degenerating and may contain areas of fluid density⁶. On contrast enhanced CT scan, intravascular fibroids are seen as hypodense filling defects in the vessel, inferior vena cava and right atrium⁷. This finding is occasionally misdiag-

nosed or diagnosed lately because of its rarity. Preoperative diagnosis of intravascular fibroids depends on a huge suspicion of doubt.

We present a very rare case of uterine fibroids going into the heart, which was diagnosed as right atrial myxoma on echocardiography. We present illustrations of case with a review of the pre-operative CT assessment. It is important that a multidisciplinary approach should be taken for diagnosis and management.

CASE REPORT

Our patient was a 50 years old female with few weeks' history of shortness of breath, vague and long term abdominal pain. She was a previously diagnosed case of multiple uterine fibroids. She did not have any cardiac disease. There was no history of diabetes mellitus, hypertension or liver disease. There was no abnormality detected on physical examination. The laboratory tests also revealed normal results including tumor markers etc. Her transthoracic echocardiography showed echogenic oval mass in right atrium which approximately measured 5.5 cm x 2.3 cm suggestive of tumor. It had smooth margins with wall of right atrium and did not have any stalk. There was moderate tricuspid regurgitation. In addition, abdominal ultrasound showed enlarged uterus containing multiple fibroids.

There was a complex density right adnexal mass with abnormal flow like tortuous vessels in right para uterine region. Due to her long time complaint of abdominal pain, her imaging was done by multidetector CT scan. MDCT was performed on 128 slicer Toshiba scanner and images were assessed on vitrea workstation. Uterus was enlarged and replaced by multiple fibroids. These fibroids were abutting and involving the right adnexa. There were multiple tortuous collateral vessels along right ovarian vessels, some with arteriovenous shunting. Multiple filling defects were seen in right ovarian vein and IVC. CT chest sections revealed serpiginous heterogeneous densities in right atrium and right ventricle of heart (Figures1). Images were reconstructed in multiple planes and 3D images were reconstructed using CT vessel selection tool (Figure 2), which clearly demonstrated connection between the uterine fibroids, all the intravascular filling defects (in right gonadal vein & IVC) and heterogeneous densities in right cardiac chambers. The case was assessed and discussed in intra-departmental consultation conference by a team of qualified consultant radiologists. The filling defects in right gonadal vein and IVC were definitely representing thrombi. But close proximity of these probably thrombosed vessels with uterine fibroids and loss of intervening fats were raising strong suspicion of intravascular leiomyomatosis, which is a very rare entity. These findings were suggest-

ing extension of fibroids into the right gonadal vein and through it into the inferior vena cava and right cardiac chambers. Therefore, the diagnosis of leiomyomas with intravascular and intra-cardiac extension was made. Our patient underwent one stage combined multi-disciplinary treatment including departments of gynecology and cardiovascular surgery. Thoraco-abdominal surgery was done. Pelvic uterine fibroids were excised by gynecological team. Cardiovascular team removed the intravascular tumor. The intravascular tumor / fibroids were almost 25 cm long. The surgical opinion was that fibroids within vessels and heart had well-demarcated borders with the vascular walls and heart. Subsequently the pathologic report also confirmed IVL.

DISCUSSION

Intravascular leiomyomatosis (IVL) is benign smooth muscle tumor within vessels, from intrauterine to the systemic veins⁸. Fewer than 200 cases of IVL have been reported worldwide. Only 14 cases involved intracardiac extension from the IVC. Only a few scattered case reports of IVL exist in the radiology literature⁹. Extra uterine involvement of fibroids occurs in approximately 30% of cases and intracardiac extension accounts for about 10%^{10,11}. Although the pathogenesis remains unclear; one theory suggested that leiomyomas originate from the vessel wall¹² whereas according to another

Figure 1 (a-f): Multiple axial images of Post contrast CT scan showing large contour deforming uterine masses (fibroids). Linear hypodense filling defects (arrows) are seen in right ovarian vein and IVC, extending into the right cardiac chambers.

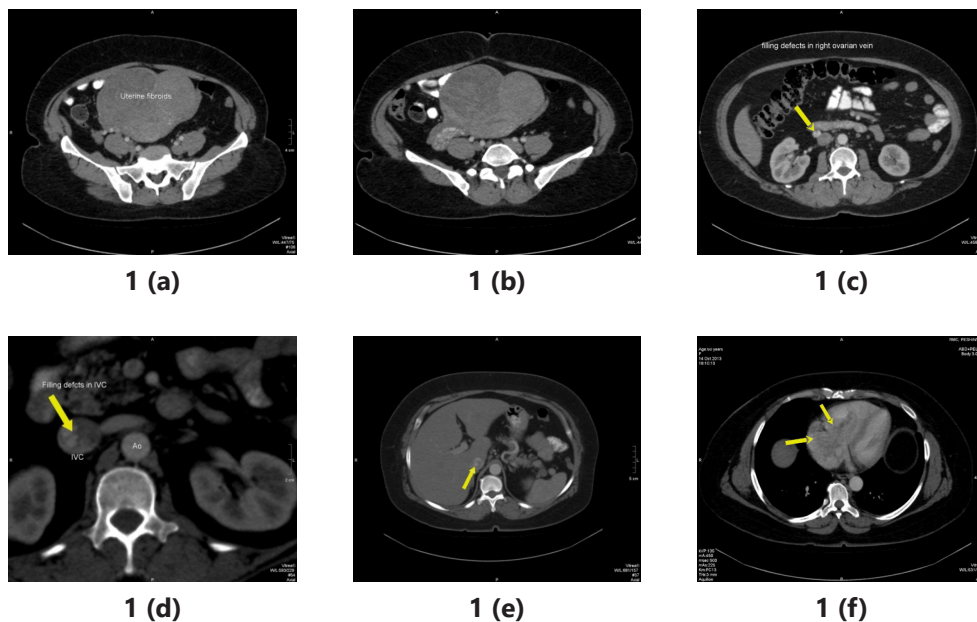
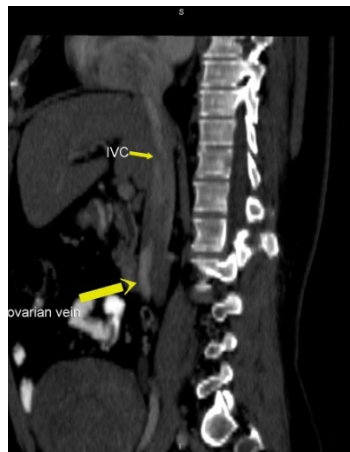


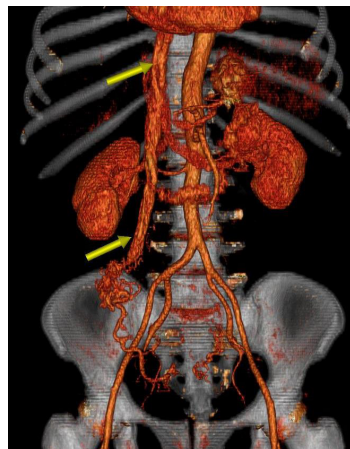
Figure 2 (a-d): Multiplanar and 3D surface shaded images showing serpiginous filling defects (arrows) extending from uterus and right ovarian vein into IVC and right heart.



2 (a)



2 (b)



2 (c)



2 (d)

theory, the uterine fibroids invaded into the uterine vein¹³, which is similar to our case. In our case, there was evidence of uterine leiomyoma invading into the vessels shown by the classic features of CT imaging findings. So far, 90% of reported cases of intravascular fibroid extension have occurred in parous females and 10% had previous pelvic surgery or hysterectomy¹⁴. Incomplete hysterectomy may cause intravascular extension of fibroids¹⁵, or alternatively, sometimes the leiomyomas may arise from the smooth muscles of vessel wall¹⁶. Our patient had a normal pregnancy and was delivered four years earlier via cesarean section. Leiomyomas might invade the uterine or ovarian veins, and can progress into the IVC and right heart¹⁶. Case of uterine fibroids extending into the heart was seen similar to our case originating from huge leiomyoma in the pelvis and extending into the ovarian vein, the IVC, the right atrium, the right ventricle and the pulmonary artery⁵. Post-con-

trast CT plays an instrumental role in diagnosing intravascular leiomyomatosis. MDCT acts as a road map for surgeons especially if done with proper angiography protocol. Recent literature has recommended that diagnosis of intravascular leiomyomas should be considered in females with history of pelvic surgery, partial hysterectomy or uterine fibroids, who present with nonspecific abdominal symptoms or with deep vein thrombosis and CT scan showing a uterine mass with tumor thrombus in IVC or even the right atrium of heart¹⁷.

CONCLUSION

Fibroids going into the heart via vascular invasion or intravascular leiomyomatosis (IVL) should be considered in patients with a history of uterine fibroids or a pelvic mass accompanied by right atrial tumor.

128 slice MDCT with its high resolution and multi-

planar imaging plays an instrumental role in diagnosing intravascular extensions of uterine fibroids and provides a road map for surgeons to plan treatment and tumour resection.

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