

## Case Report

# Imaging manifestations of intravascular leiomyoma involving the right atrium: a case report and literature review

Aimei Ouyang, Xinyou Su, Shujing Li, Kun Li, Jian Wang

Medical Imaging Center, Jinan Central Hospital Affiliated to Shandong University, Jinan 250013, Shandong Province, P.R. China

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**Abstract:** The aim of this study is to investigate the imaging features of intravascular leiomyoma (IVL) involving the heart and the imaging techniques in the diagnosis of this disease. The imaging features of contrast-enhanced computed tomography (CT), the clinical data and the pathological data of a case of IVL involving the right atrium were retrospectively analyzed and the literatures were reviewed. A 42-year-old woman was admitted to Jinan Central Hospital with a 7-day history of lower extremity weakness, chest tightness and short breath. Contrast-enhanced CT scanning revealed that there was a mass in the inferior vena cava and right atrium, which was heterogeneously enhanced. There was a gap between the vessel wall and the mass. Spiral CT scanning with multiplanner reformation (MPR) reconstruction revealed the morphology, scope and extension pathways of the tumor clearly. Ultrasonography (US) and magnetic resonance imaging (MRI) also played important roles in the diagnosis and differential diagnosis of IVL. To the patients with a history of uterine fibroids, spiral CT scanning, US and MRI could be used to improve the correct diagnosis rates of IVL before surgery.

**Keywords:** Imaging manifestations, intravascular leiomyoma, the right atrium

## Introduction

Intravascular leiomyoma (IVL) is a rare type of tumor that was first reported by Birch-Hirschfeld in 1896 [1]. Since then, only about 200 cases have been reported in the literature and even fewer are the cases involving the inferior vena cava and the right atrium. There are no specific clinical manifestations in patients with IVL. IVL involving the right atrium can lead to chest tightness, chest pain, palpitations, dizziness, etc. In severe cases, sudden death can occur. Nowadays, little is known about the clinical features of IVL and the early diagnosis is difficult. Thus, it is important to be familiar with the imaging and clinical features of IVL.

## Case report

### *History and examination*

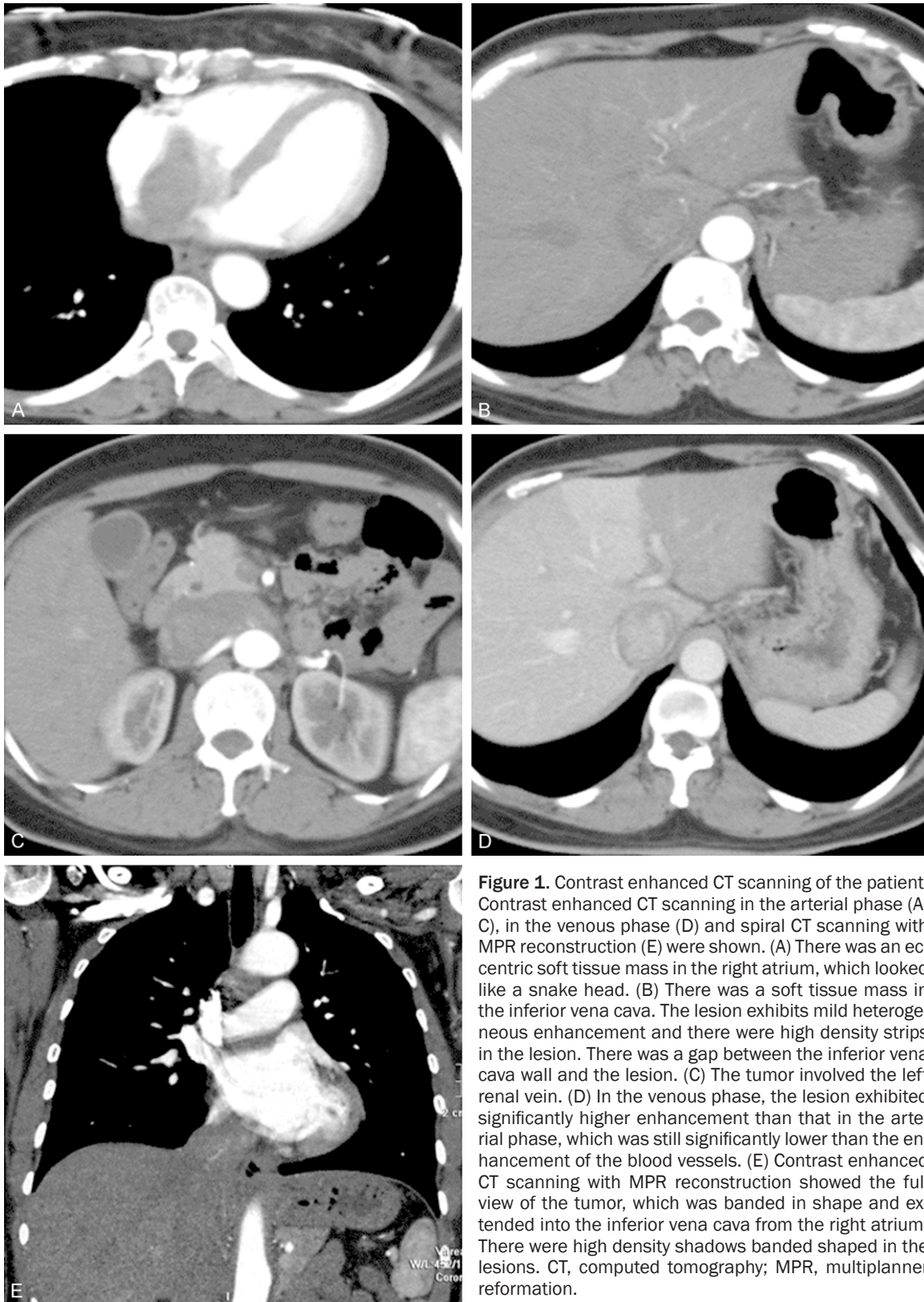
A 42-year-old woman was admitted to Jinan Central Hospital with a 7-day history of lower extremity weakness, chest tightness and short

breath. The physical examination did not reveal abnormalities in the heart, bilateral lungs and lower limbs. The electrocardiogram recording of the patient was normal. The patient received myomectomy 6 years ago.

### *Imaging*

Contrast enhanced computed tomography (CT) scanning of the chest was performed to rule out pulmonary embolism. It showed a soft tissue density filling defect with clear margin measuring about 3×3 cm at the inlet of right atrium, which looked like a snake head and extended into the inferior vena cava. In the arterial phase, the lesion exhibited mild heterogeneous enhancement (CT value, about 70 HU) and there were high density strips in the lesion (**Figure 1A-C**). In the venous phase, the lesion exhibited significant enhancement (CT value, about 120 HU) and there was an annular gap between the inferior vena cava wall and the lesion (**Figure 1D**). Contrast enhanced CT scanning with multiplanner reformation (MPR) reconstruction revealed eccentric filling defect

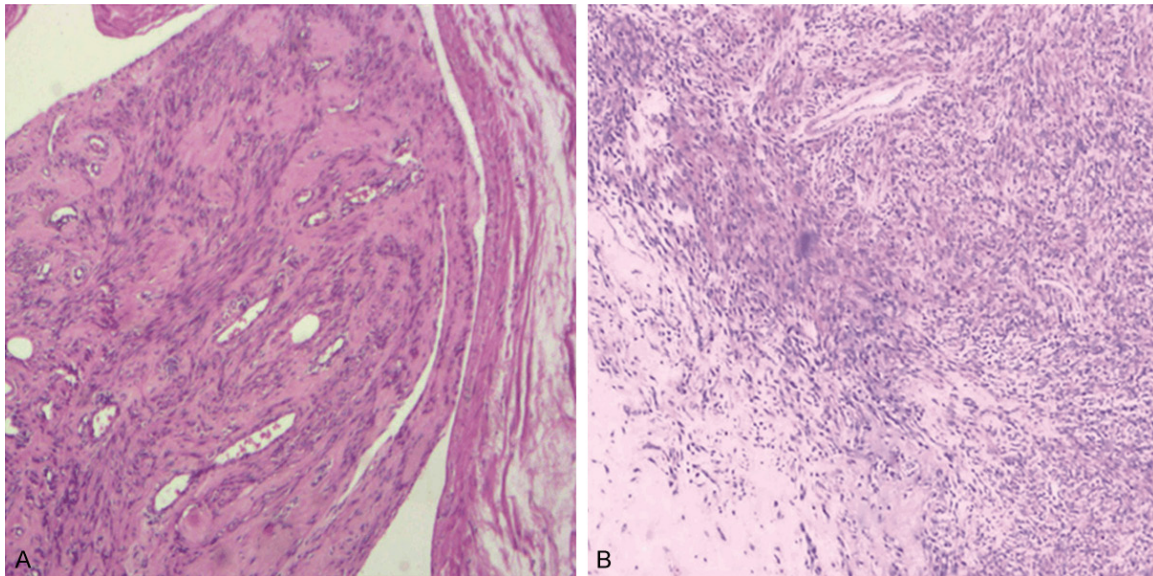
## IVL involving the right atrium



**Figure 1.** Contrast enhanced CT scanning of the patient. Contrast enhanced CT scanning in the arterial phase (A-C), in the venous phase (D) and spiral CT scanning with MPR reconstruction (E) were shown. (A) There was an eccentric soft tissue mass in the right atrium, which looked like a snake head. (B) There was a soft tissue mass in the inferior vena cava. The lesion exhibits mild heterogeneous enhancement and there were high density strips in the lesion. There was a gap between the inferior vena cava wall and the lesion. (C) The tumor involved the left renal vein. (D) In the venous phase, the lesion exhibited significantly higher enhancement than that in the arterial phase, which was still significantly lower than the enhancement of the blood vessels. (E) Contrast enhanced CT scanning with MPR reconstruction showed the full view of the tumor, which was banded in shape and extended into the inferior vena cava from the right atrium. There were high density shadows banded shaped in the lesions. CT, computed tomography; MPR, multiplanar reformation.

extended into the inferior vena cava from the right atrium. And there were high density vascu-

lar shadows which were banded in shape in the lesions (Figure 1E).



**Figure 2.** H&E staining of the tumor sections ( $\times 100$ ). A. Resection of the tumor in the right atrium and inferior vena cava was performed. Histopathologic examination showed the tumor was consistent with an intravascular leiomyoma. The tumor cells were spindle in shape with bundle arrangement and uneven density. The cells were well differentiated and mitotic cells were rare. There were blood vessels in the tumor and endothelial cells at the edge of the tumor. There was a gap between the tumor and the inferior vena cava wall. B. Histopathologic examination of the uterus was performed. There were multiple leiomyomas in the uterus. The tumor was consistent with a smooth muscle tumor. The tumor cells were well differentiated with a swirling arrangement.

### Operation

Resection of the tumor in the right atrium and inferior vena cava was performed. Surgical exploration revealed a solid mass measuring about  $3 \times 3 \times 3$  cm in the right atrium, which extended into the inferior vena cava. The mass was grey, tough and weaving shaped without obvious adhesion to adjacent tissues. The operator held the tumor and pulled it out slowly. The total length of the tumor was about 20 cm and the width of the tumor was uneven with the largest diameter of about 5 cm. The tail of the tumor was beaded shaped. Histopathologic examination showed the tumor was consistent with an intravascular leiomyoma. The tumor cells were spindle in shape and well differentiated. There were endothelial cells at the edge of the tumor (**Figure 2A**). Immunohistochemistry revealed the cells were reactive for SMA (+), actin (+), CD34 (+), ER (++) , PR (++) but negative for S-100 and CD68. The positive rate of Ki-67 was less than 1%.

This patient also experienced irregular vaginal bleeding and was diagnosed as having uterine fibroids by ultrasound (US). One month after the tumor resection, the patient received hysterectomy

plus bilateral appendix resection. The uterus was enlarged to a 3 month pregnancy size with smooth surface and irregular shape. It included a plurality of myoma-like protrusions. The largest protrusion located in the anterior wall of the uterus with the diameter of 6 cm. The right ovary was cystically enlarged to 4 cm in diameter with corpus luteum tissue and clot. The left ovary was normal sized. While there were multiple small fibroids inside the vessels of the left ovary. The bilateral fallopian tubes showed no gross abnormality. The resected uterine was incised. The endometrium and cervical canal were smooth. There were multiple myoma-like protrusions inside the muscular layer and some of them were degenerative. Histopathologic examinations of the uterus and bilateral appendix were performed. 1) There were multiple leiomyomas in the uterus with the largest diameter of about 6 cm. The tumor was consistent with a smooth muscle tumor of low malignant potential. There was local necrosis of the tumor. Intensive hyperplasia of the tumor cells with mild atypia could be observed. The mitotic count was 5~6/10 high-power field (HPF, **Figure 2B**). Immunohistochemistry revealed the cells were reactive for CD10 ( $\pm$ ), p53



(+), SMA (+), desmin (+), but negative for CD34. The positive rate of Ki-67 was about 30%. 2) The histopathological report also showed simple hyperplasia of endometrium plus polyps. 3) There was leiomyoma in the left ovarian ligament. Immunohistochemistry revealed the cells were reactive for SMA (+) as well as desmin (+), but negative for CD10, CD34 and p53. The positive rate of Ki-67 was less than 1%.

### Discussion

Vascular smooth muscle tumor, also known as IVL, is a rare and special tumor. It originates from the uterine leiomyoma with vascular invasion [2]. It can also arise from the smooth muscle tissues of the uterus or the vein walls in the pelvic cavity [3]. The etiology of the disease remains unclear. It has been reported that the occurrence and development of IVL are associated with high estrogen levels [4]. According to the current clinical data from the relevant literatures, IVL occurs in women and most of them are premenopausal with average age of 40 years as well as a history of uterine fibroids [5]. It is a benign tumor. However, it may behave in a malignant manner with invasion of surrounding tissues and potential relapse. The ratio of the patients with IVL involving the inferior vena cava and right atrium is 10%-30%. Furthermore, IVL may also involve the hepatic vein, renal vein [6] and pulmonary arteries [7]. Complete resection is the key factor for successful treatment. In this case, the patient has been followed up for 14 months and no recurrence was observed.

The onset of IVL is insidious. The clinical manifestations vary greatly with the differences in the scope and extent of the disease. Usually, the symptoms of inferior vena cava obstruction are not obvious in the early stage. The patients may have varying clinical degrees of venous disorders, including leg soreness, hepatomegaly, ascites, etc. If the heart is involved, the patients will suffer from cardiac dysfunction including palpitations, shortness of breath, syncope, etc [8, 9].

Nowadays, IVL is mainly diagnosed by US, CT and magnetic resonance imaging (MRI). The imaging features of IVL were listed as the follows. 1) The US shows that the uterus enlarges due to multiple fibroids. There is continuous hypoechoic area along the iliac vein, inferior vena cava to the right intracardiac cavity since

IVL can grow along the vein. The affected veins are enlarged with banded hypoechoic area inside. Because ultrasonic examination cannot show the full view of the lesions, it may lead to misdiagnosis. In this case, the patient was misdiagnosed as having inferior vena cava thrombus by US. 2) The CT examination is the most important means of IVL diagnosis. Plain CT scanning shows the right atrium is enlarged and the affected veins are expanded and thickening. Contrast enhanced CT scanning shows free filling defect in the right heart cavity, which looks like a snake head in some cases and can extend both upward into the pulmonary arteries as well as its branches and downward into the inferior vena cava as well as the uterine veins. The lesions are free and banded in shape with a clear border to the surrounding vascular walls. They exhibit mild heterogeneous enhancement and there are vascular shadows which are banded in shape in some lesions. Furthermore, CT scanning can show free filling defect in the hepatic vein, renal vein and iliac vein. Spiral CT scanning with MPR, maximum intensity projection (MIP) or volume rendering (VR) reconstruction reveals the morphology, scope, relationship to the vascular walls and extension pathways of the tumors. The lesions of IVL usually show the shape of radish root-like hypertrophy. 3) MRI can show the scope of the tumor clearly. The MRI signal is uniform throughout the tumor with the characteristics of equal or low-intensity signal on T1 weighted imaging (T1WI) and high-intensity signal on T2 weighted imaging (T2WI). The signal increases slightly or significantly after contrast enhancement.

IVL should be differentially diagnosed from the following diseases. 1) IVL involving the inferior vena cava should be differentiated from venous thrombosis, cancerous emboli and primary leiomyoma of the inferior vena cava. The venous thrombosis is close to the wall of the vessel and there is no gap between them. There is no enhancement at contrast enhanced CT. Usually, there were no uterine lesions in the patients with venous thrombosis. Cancerous emboli have different degree of enhancement in contrast scan. The patients often show a clear history of malignancy and the lesions of these patients are limited. In most cases, primary leiomyoma of the inferior vena cava occurs in older women and the lesions locate in the middle and lower segment of inferior vena cava.

The inferior vena cava is expanded and there is no clear border between the lesions and the surrounding vascular walls. The lesions exhibit heterogeneous enhancement at contrast enhanced CT scanning. 2) IVL involving the right heart cavity should be differentiated from primary cardiac tumors such as myxoma and angiosarcoma. There is clear boundary between the myxoma and the right atrial wall. In most cases, myxoma attaches to the interatrial septum with a short pedicle. In some cases, myxoma could also attach to the right atrial wall. Different from IVL involving the right heart cavity, myxoma does not connect to the mass in the inferior vena cava. It reciprocates with systole and diastole using the pedicle as a fixed point. Angiosarcoma is irregular in shape and grows quickly with heterogenous density and liquefaction necrosis inside the lesion.

In summary, IVL is a rare type of tumor with unique imaging characteristics. It occurs in women and most of them have a history of uterine fibroids. CT scanning or MRI reveals continuous and free filling defect in the heart and blood vessels. The lesions show focus enhancement on enhanced CT scanning or MRI. The presence of these clinical and imaging characteristics should raise suspicion of IVL.

## Disclosure of conflict of interest

None.

**Address correspondence to:** Aimei Ouyang, Medical Imaging Center, Jinan Central Hospital Affiliated to Shandong University, No. 105, Jiefang Road, Jinan 250013, Shandong Province, P.R. China. Tel: +86-0531-85695258; Fax: +86-0531-86951300; E-mail: 13370582510@163.com

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