The value of $^{18}$F-FDG PET/CT in the diagnosis of intravenous leiomyomatosis of the uterus extended into the right atrium

**Abstract**

Intravenous leiomyomatosis (IVL) of the uterus is a rare neoplasm which usually occurs after hysterectomy. Due to its rarity and non-specific clinical manifestations, IVL is commonly misdiagnosed as malignant thrombus or thrombosis and treated inappropriately. Herein, we report an unusual case of a 51 years old woman with IVL without hysterectomy or abdominal manifestations. The IVL was detected in the right atrium. This case highlights the usefulness of $^{18}$F-FDG PET/CT in the diagnosis of IVL.

**Introduction**

Intravenous leiomyomatosis (IVL) of the uterus is a histologically benign but it is a quashi-malignant, potentially life threatening tumor with unpredictable biological behavior, due to growing and extension within the intrauterine and extraterine venous system [1]. Since IVL was first described by Birch-Hirschfeld in 1896, less than 400 cases have been reported in the English literature [2]. Due to its rarity and the non-specific clinical manifestations, IVL is frequently misdiagnosed as malignant thrombus or thrombosis. Thus, patients suffering from IVL were often treated inappropriately [3]. The commonly found extension of IVL from intrauterine venules to inferior vena cava, right atrium chambers and even pulmonary artery, obviously increases the risk of sudden death. Fluorine-18-fluorodeoxyglucose positron emission tomography/computed tomography ($^{18}$F-FDG PET/CT) not only offers the glycometabolic information of the lesion in situ, but also facilitates to identify unexpected foci of the tumor.

**Case Report**

A 51 years old woman was admitted in our institution after cardiac syncope, happened twice during the past month. The patient also suffered from hypertension (150/110mmHg) with dizziness, headache, decreased vision, chest distress, breathlessness, paroxysmal nocturnal dyspnea, which could not be alleviated by antihypertensive drugs. Electrocardiogram on admission showed normal sinus rhythm and myocardial ischemia in the left and the inferior wall of the left ventricle (ST-segments decrease of more than 5mm in leads II, III, AVF, V5, and V6). Transthoracic echocardiography showed two free-floating masses in the right atrium chamber (Figure 1A), which had similar acoustic findings as thrombosis [4]. However, the patient had not any history of thromboembolism or deep venous thrombosis, and serum D-dimer was also within normal range. In order to determine the nature of the lesion, especially to exclude the possibility of a malignant thrombus, $^{18}$F-FDG PET/CT was performed, which presented heterogeneously low $^{18}$F-FDG uptake within the lesion (Figure 1B). Interestingly, two irregular masses with moderate $^{18}$F-FDG accumulation (left: 99×63mm, SUVmax=2.16; right: 95×55mm, SUVmax=2.24; Figure 1C-D) in both broad ligaments were accidently found. Aiming to alleviate symptoms and prevent sudden death, thoracotomy was carried out. A gross, lobulated mass was found in the right atrium extending into the inferior vena cava. The lesion was resected and diagnosed as a thrombus via intraoperative rapid pathology.
Considering that symptoms were markedly relieved after surgery, the patient was released. The lesions in the broad ligaments with low $^{18}$F-FDG uptake were considered as benign related to the ovaries and remained untreated. Three months later, the patient returned to our hospital because of abnormal discomfort, distension and leukorrhagia. Pelvic magnetic resonance imaging (MRI; Figure 2) demonstrated that the lesions in both ligaments were significantly enlarged (left: 146×90mm; right: 134×57mm). Hysterectomy plus bi-lateral oophorectomy was performed. The lesion in the uterus and the specimen from the first operation were retrieved for detailed pathological examination and analysis. Pathological examination showed that cells of spindle shape with low mitotic activity, or of leaf shape with obvious proliferation accompanied by myxoid tissues that were homogeneously stained red in their central region were observed after hemoatoxylin and esosin staining (H & E; Figure 3A). Immunohistochemical staining revealed that the lesion was strongly positive for smooth muscle actin (SMA) (Figure 3B), desmin (DES) (Figure 3C), myoglobin (Myo) (Figure 3D), and h-caldesmon. The final diagnosis of IVL was confirmed after multidisciplinary consultation.

**Figure 1.** A, Echocardiography of the patient at the first admission shows a free-floating mass extending to inferior vena cava, which had similar acoustic presentations as thrombosis. B, Axial PET/CT image reveals heterogeneously low $^{18}$F-FDG uptake in this lesion. C-D, Axial and coronary PET/CT images demonstrated two irregular masses with moderate $^{18}$F-FDG accumulation in an area of both broad ligaments (left: 99×63mm, SUVmax=2.16; right: 95×55mm, SUVmax=2.24).

**Figure 2.** Pelvic transversal T1-weighted (A), T2 weighted (B) and contrast-enhanced T1-weighted (C) magnetic resonance imaging performed three months later presented that the lesions in bilateral ligaments were obviously enlarged (left: 146×90mm; right: 134×57mm).

**Figure 3.** Histopathologic examination demonstrated spindle-shape cells with low mitotic activity (A, H & E staining, magnification×10). Immunohistochemical staining was positive for SMA (B, magnification×40), DES (C, magnification×40) and Myo (D, magnification×40).

**Discussion**

Twelve IVL cases confirmed by autopsy or pathological examination were retrospectively analyzed in Zhongshan Hospital from 2009 to 2016. Common clinical manifestations were similar to uterine fibroids, including abdominal pain, fall bilge feeling and irregular bleeding. Unfortunately, PET/CT examination was performed by only one patient. In addition, unlike previously reported IVL cases [5-9], this presented patient had no history of hysterectomy or abdominal manifestations. Fluorine-18-FDG PET/CT did reveal the lesions in both broad ligaments with only mild-to-moderate $^{18}$F-FDG accumulation, which were considered as of benign nature. The possibility of IVL was not considered although the patient had lesions in the right atrium, extended to the vena cava and abnormalities around the uterus. Incomplete resection may result in recurrence or progression of IVL [10-13], just as in our case. However, the long-time prognosis is favorable because of its benign nature. Twenty seven months after the second operation, no obvious radiographic evidence of recurrence was noted in this patient. Diagnosis of IVL and another benign tumor by $^{18}$F-FDG PET/CT alone is still difficult due to lesions' benign nature that lead to low uptake of $^{18}$F-FDG. Fluorine-18-FDG PET/CT facilitates to identifying the unexpected lesions [14].
In conclusion, we report a case of a 51 years old woman with severe symptoms of hypertension and a thrombotic lesion in the right atrium and the vena cava accompanied with abnormalities around the uterus. Pathology showed IVL similar lesions in both broad ligaments.

The authors declare that they have no conflicts of interest.

Bibliography


Norman Rockwell. Before injection, 1958. Oil in canvas, 35x27.5cm. Norman Rockwell Museum, Stockbridge, Massachusetts