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Brief Report

Orthostatic hypotension and right heart failure as the initial manifestation of intravenous leiomyomatosis

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Abstract A 36-year-old woman, who had a history of myomectomy, presented with lightheadedness after changing position from sitting to standing and effort-related shortness of breath. Echocardiography demonstrated a hyperechoic elongated mobile mass extending from the inferior caval vein to the right atrium. The mass was surgically removed, and histological examination established the diagnosis of intravenous leiomyomatosis. This case caught the attention of our cardiology group to consider the diagnosis when an inferior caval vein or right atrium mass is found in a patient with a history of uterine leiomyomatosis.

Keywords: Intravenous leiomyomatosis; orthostatic hypotension; right heart failure

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Case report

A 36-year-old woman presented to an internist because she reported feeling lightheaded after changing position from sitting to standing. The frequency of these attacks was approximately once per day for 5 days. Physical examination revealed a grade III/VI systolic ejection murmur. On electrocardiography, a new right-bundle-branch block pattern was noted, and she was referred to a cardiologist. A careful history of the patient was obtained and physical examination was performed. She also complained of fatigue, malaise, decreased appetite over the preceding 3 months, and mild decreased exercise tolerance for 20 days. According to her medical history, she underwent a myomectomy in 2008 and was not followed-up with routine imaging.

On physical examination, her heart rate was 92 bpm and her blood pressure was 116/78 mmHg in the supine position. The patient's standing blood pressure was 72/53 mmHg, suggestive of orthostatic hypotension. A raised jugular venous pressure, a grade III/VI systolic murmur at the tricuspid valve auscultation area, and mild lower extremity oedema were noted, and the remainder of the physical examination was unremarkable. The routine laboratory examinations of blood and urine were normal. *Electrocardiography* demonstrated right-bundle-branch block pattern. Echocardiography showed a mobile solid mass in the inferior caval vein, which prolapsed into the right atrium and obstructed the tricuspid valve during diastole. A thoraco-abdominal contrast-enhanced CT scan further showed a mass extending from the right internal iliac vein to the right atrium (Fig 1). Following careful preparation, the patient underwent a one-stage thoraco-abdominal surgical procedure with a total hysterectomy, bilateral salpingo-oophorectomy, and removal of a solid, smooth, and rubbery tumour extending from the pelvis to the right atrium, which measured 580 mm in length and 23 mm in maximum diameter. The mass had well-demarcated borders and was not attached to the vessel walls. Histological examination of the mass showed smooth muscle cells with good differentiation and confirmed the diagnosis of intravenous leiomyomatosis (Fig 2). A control CT scan revealed no evidence of tumour, and the symptoms of lightheadedness, dyspnoea, and oedema diminished when she was discharged 2 weeks later.

Discussion

Intravenous leiomyomatosis is a benign smooth muscle tumour but acts like a malignant tumour and grows within venous channels. All described patients

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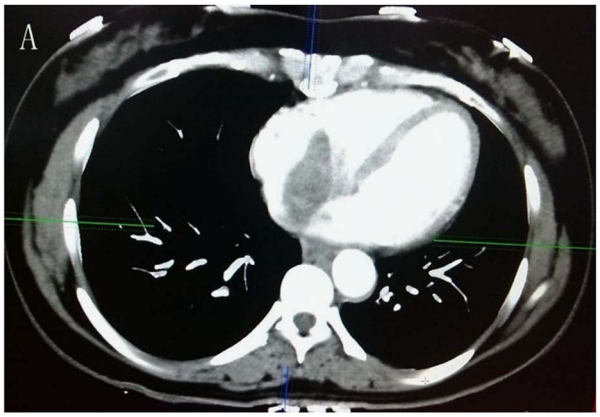

Figure 1.
A contrast-enhanced CT scan showing a mass (leiomyoma) extending to the right atrium.

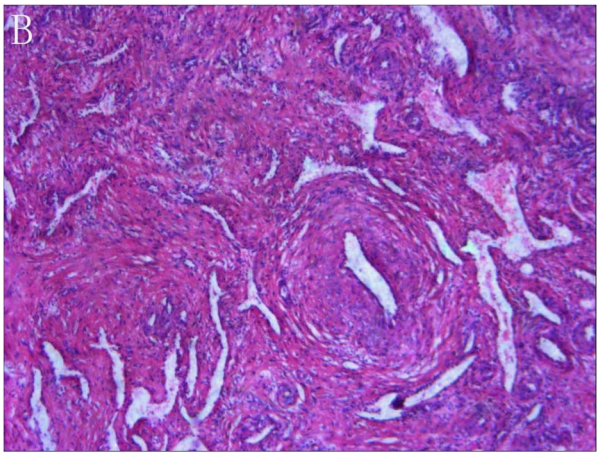

Figure 2.
A histological examination of a haematoxylin- and eosin-stained tissue section (100X). Microscopically, the tumour is composed of interlacing bundles of bland smooth muscle cells.

are female, pre-menopausal, and most of them are parous. The median age is 44 years, with patients ranging from 26 to 70 years old. Although the aetiology of intravenous leiomyomatosis is unknown, the presence of oestrogen and progesterone receptors in the cell nucleus argues that the tumour is indeed of uterine origin.[1]

In most cases, intravenous leiomyomatosis remains localised within the uterine vessels, but extension into the central vessels and right heart can occur, as seen in our patient. Intravenous leiomyomatosis with heart extension may remain asymptomatic during the early stage. As the mass increases in size, it can cause a range of symptoms of heart disease, including exertional dyspnoea, shortness of breath, orthostatic hypotension, syncope, right heart failure, or even sudden cardiac death. Our patient presented with orthostatic but not supine hypotension, which was probably a consequence of drastically reduced venous return in the standing position. The possible mechanisms include a reduced venous return reserve by the tumour mass or a mechanical movement of the tumour mass downward, resulting in obturation of the caval vein lumen or tricuspidalis dysfunction.

The differential diagnosis of intravenous leiomyomatosis with heart extension included atrial myxoma, vegetation, enlarged Eustachian valve, or a tumour thrombus commonly metastasised from the lung or breast. The recent literature has recommended that an intravenous leiomyomatosis diagnosis should be considered in all women presenting with a mass visualised by echocardiography or CT scan in both the right atrium and the inferior caval vein, right heart failure-related symptoms associated with a history of pelvic surgery, hysterectomy, or uterine leiomyomatosis.[2] Echocardiography typically shows a mobile heterogeneous mass in the right atrium with or without protrusion into the right ventricle.[3] A definitive intravenous leiomyomatosis diagnosis is confirmed by histopathological analysis.

In order to reduce the risk of sudden death, surgically treating intravenous leiomyomatosis with heart extension should be performed as soon as possible, and it has a favourable prognosis. There are two surgical procedures available:[4] one requires complete extraction of the intra-cardiac and intra-caval mass through a one-stage surgical approach. Although the one-stage surgery could result in a much longer operative time, it is the preferred operation for patients with a clear pre-operative diagnosis, good health conditions, and low operative risks. If the tumour is too extensive, or adheres to the cardiac and vascular structures, then a separate operation involving an abdominal stage and a thoracic stage may be mandatory. In addition, two-stage surgeries are typically planned for high-risk patients, including those with cardiopulmonary co-morbidities and risk factors for major bleeding. More than half of patients with intravenous leiomyomatosis and extension had a history of hysterectomy and it has been suggested that the tumour is oestrogen dependent. As a result, either surgical approach should also include total hysterectomy and bilateral salpingo-oophorectomy. Anti-oestrogenic drugs have been used pre- and post-operatively to reduce the tumour burden and control residual tumours.[5] A long-term follow-up of patients after resection of intravenous leiomyomatosis is needed because recurrences up to 17 years after primary resection have been reported.

In conclusion, intravenous leiomyomatosis should be suspected in young women who have cardiac symptoms in whom a right-sided intra-cardiac mass is present.

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Conflicts of Interest

None.

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