A highly unusual right atrial mass presented in two women

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Intravenous leiomyomatosis is a rare, benign neoplasm of the uterine, affecting adult women. We report two cases in whom intravenous leiomyomatosis extended through the inferior vena cava into the right heart chambers and the pulmonary artery. Both patients underwent staged operation with excision of the cardiac and primary tumour. The differential diagnosis of a right atrial mass in middle-aged women should include intravenous leiomyomatosis.

KEYWORDS
Intravenous leiomyomatosis; Right atrial mass

Differential diagnosis of right atrial masses includes thrombus (pulmonary emboli-in-transit), primary and metastatic cardiac tumours, and vegetations on the tricuspid valve and intracardiac electrodes. We present here two cases in whom an unusual mass in the right heart chambers proved to be intravenous leiomyomatosis.

Case 1

A 52-year-old female with recent transient ischemic attack (TIA) was scheduled for routine echocardiographic examination. This patient had a history of hysterectomy 10 years previously due to leiomyoma. Echocardiography showed a large mobile mass in the right heart chambers originating from the inferior vena cava (IVC) (Figure 1A, see Supplementary data online, Figure S1A–C). The presence of a patent foramen ovale may have been responsible for her previous TIA. A subsequent MRI revealed echogenic findings in the IVC and right heart chambers bulging into the right pulmonary artery resembling a wire; the left ovarian vein was enlarged and filled with thrombus.

The patient was scheduled for surgery. During the first stage, extraction of the cardiac mass was planned, followed by a gynaecologic operation of the ovarian veins. During induction of anesthesia, the patient suddenly collapsed. Transoesophageal echocardiography (TEE) showed enlargement of right heart chambers and a large thrombus-in-transit which migrated into the main pulmonary artery.

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Figure 1 (A) Subcostal echocardiographic view. The mass originating from the inferior vena cava (IVC) enters the right atrium. (B) Surgical finding: A long compact mass removed from the right pulmonary artery and proved be leiomyomatosis.
The main pulmonary artery was explored and the thrombus was removed; the right pulmonary artery was also explored with extraction of a long hard bone-like mass (Figure 1B).

The postoperative pathological staining was consistent with leiomyomatosis. Subsequently, the patient underwent laparotomy and excision of the ovarian vein which also was proved to be involved with leiomyomatosis.

Case 2

A 49-year-old female underwent routine echocardiography, which revealed a large lobular mass, originating from the IVC, bulging into the right atrium (RA), and seemed to be entrapped in the IVC (see Supplementary data online, Figure S2A and B). A subsequent echocardiography showed further enlargement of the RA mass and its protrusion into the right ventricle (see Supplementary data online, Figure S2C). Computed tomography scan showed a filling defect in the IVC that started up in the RA and reached all the way down into the bifurcation of the iliac veins (Figure 2). The patient was referred to surgery. A 25 cm long mass was extracted from the IVC and RA. On a subsequent procedure, extraction of a leiomyomatotic mass from the pelvic veins was performed accompanied by hysterectomy.

Comment

Intravenous leiomyomatosis is a rare benign smooth-muscle tumour arising from uterine leiomyoma or from myometrial vein. The presenting symptom in most cases is congestive heart failure with peripheral oedema due to right atrial and ventricular obstruction,1 syncope,2 pulmonary embolism—occurring in our first case, systolic murmur,3,4 or it can be an incidental finding as in our cases.

We were able to identify 78 reported cases of intravenous leiomyomatosis.4 The differential diagnosis includes thrombus (pulmonary emboli-in-transit), right atrial tumour, more commonly metastatic. Metastases may reach the heart via the lymphatic (pericardial metastases: carcinoma of lung and breast) or by haematogenous route (myocardial metastases: malignant melanoma, lymphoma, leukaemia, soft tissue, and bone sarcoma), or by transvenous extension through the IVC—renal and hepatocellular carcinoma, leiomyoma of the uterus, Wilms’ tumour, and others.5

Extension of intravenous leiomyomatosis into the right pulmonary artery observed in 9.7%4 and is often associated with massive pulmonary embolism.

Patients with intravenous leiomyomatosis often have a history of hysterectomy. The important echocardiographic feature is the penetration of the tumour from the abdominal venous system into the RA, and often into the right ventricle and pulmonary arteries. In summary, a right atrial mass originating from the IVC in middle-aged women in particular with history of hysterectomy, even in the remote past, should raise suspicion for intravenous leiomyomatosis.

Supplementary data

Supplementary data are available at European Journal of Echocardiography online.

References