eComment. Leiomyomatosis: intracardiac extension and pulmonary embolization

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I read with great interest the article by Li *et al*. [1]. As rightly outlined in this remarkable review of the literature, extracting data solely from case reports is associated with an inherited bias. Only three intraoperative deaths were recorded, thus yielding a mortality rate of 1.5%. The one-stage approach in which a thoraco-laparotomy is performed under cardiopulmonary bypass with or without hypothermic circulatory arrest is challenging, is time-consuming, and carries a high incidence of perioperative complications. This one-stage operation is the procedure of choice for patients with cavo-atrial extension of renal cell carcinoma.

Renal cell carcinoma, like uterine leiomyomatosis, characteristically grows into the inferior vena cava and extends to the right heart chambers. In spite of the notable extension of the tumour thrombus into the right atrium, surgical resection can result in prolonged survival albeit at an increased risk of postoperative mortality. The operative mortality of this procedure remains substantial, ranging from 2.7%-13% [2]. Accordingly, complete surgical resection of intracardiac leiomyomatosis carries a significant risk of postoperative mortality. However, surgical procedure remains the mainstay of therapy and can be curative.

To date, a handful of reports, most of these isolated case reports and small clinical series, have described the management of intracardiac leiomyomatosis. However, it seems that Li *et al.* have overlooked two relevant papers. Thukkani *et al.* [3] report the case of a 36-year old woman with a history of recurrent pelvic leiomyomatosis and metastatic leiomyomatosis to the right heart chambers. The patient underwent a resection of the tumour with tricuspid valve replacement. A life-threatening bleed from the pelvic tumour was successfully managed at the end of the procedure. Zhang *et al.* [4] published their experience with intravenous leiomyomatosis with intracardiac extension in 5 women. Multiple approaches were described with excellent outcomes; the authors concluded that successful management requires timely recognition along with planed surgical intervention.

Although there is a consensus about surgical resection of intracardiac leiomyomatosis because of the risk of sudden death, there are no guidelines for the treatment of pulmonary leiomyomatosis. Some advocated a surgical excision of the pulmonary nodule as a first-line strategy with or without hormone therapy. Others have shown that a wait-and-see strategy is an acceptable option in high-risk patients or in case of unrespectable tumour [5].

Conflict of interest: none declared.

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eComment. Misdiagnosis of intravenous leiomyomatosis with cardiac extension

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We read with great interest the article by Li *et al*. [1]. They performed a comprehensive literature search including 194 cases of intracardiac leiomyomatosis. We also would like to add some comments on misdiagnosis of this complicated pathology.

Intravenous leiomyomatosis (IVL) is a rare neoplastic disease, which is characterized by intravenous growth of histologically benign, smooth muscle tumours arising from either a uterine myoma or from the wall of a uterine vessel [2, 3]. The tumour usually enters through the lumen of the iliac vein and extends upward to varying locations, including the inferior vena cava, adrenal and renal veins, right heart (intracardiac leiomyomatosis) and pulmonary artery. It is an insidious process and may lead to heart failure, pulmonary embolization, or even sudden cardiac death.

Intracardiac extension may be easily misdiagnosed as either a primary cardiac tumour or a venous thrombus-in-transit. Misdiagnosis subsequently may lead to improper treatment [2, 4, 5]. Differential diagnosis of right-sided cardiac masses includes thrombi-in-transit, right atrial myxomas, vegetations on the tricuspid valve, primary caval leiomyosarcoma, intracardiac electrodes, and tumour thrombi from malignancies such as hepatocellular carcinoma, renal cell carcinoma, Wilms' tumour (in children), and others [3, 4]. IVL may be diagnosed by echocardiography, abdominal ultrasound, magnetic resonance imaging, enhanced chest and abdominal computed tomography, and venography. The diagnosis is confirmed intraoperatively or by postoperative histopathological examination [3, 5].

A thrombus-in-transit appears as an elongated mobile mass of venous cast giving a "popcorn" appearance within the right-sided cardiac chamber, whereas there are multiple points of attachment in the inferior vena cava and right-sided cardiac chambers in patients with IVL [8]. A correct diagnosis depends on a high index of suspicion. This pathology should be considered in young women with a right-sided cardiac mass and a pelvic mass or those who have previously undergone hysterectomy for uterus leiomyoma [2, 3, 5].

Finally, we think that IVL is an underdiagnosed pathology and a huge index of clinical doubt with the modern diagnostic imaging tools will lead to an increased number of reported cases in the future.

Conflict of interest: none declared.

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