A 52-year-old woman was admitted to our department with a 1-month history of worsening dyspnea. On examination, she had a grade 2/6 systolic ejection murmur. Computed tomography of the chest revealed a mass in the right atrium and filling defects consistent with pulmonary embolus in the left pulmonary inferior artery (Fig 1A). Multislice computed tomography confirmed a hypodense mass extending into the right atrium through the right renal vein and the inferior vena cava, with a 13 cm × 9 cm heterogeneous renal mass in the lower pole, and a hepatic angiomatous appearance (Fig 1B). Under circulatory arrest, the patient underwent excision of the intravenous and intracardiac lesion and pulmonary embolectomy through a sternotomy and radical right nephrectomy through a laparotomy (Fig 2A; IVC = inferior vena cava). The final pathologic examination of the masses revealed a tumor consisting of mature adipose tissue, smooth muscle, and vessels, and positive immunohistologic stain for HMB-45 consistent with angiomyolipoma (Fig 2B; hematoxylin-eosin, ×10). The patient remains free of dyspnea 2 years after surgery, without evidence of tumor recurrence or metastases.

Surgical excision of the intravenous and intracardiac lesion, and pulmonary embolectomy and radical right nephrectomy through a laparotomy with the patient under circulatory arrest, was a reasonable treatment of choice for this challenging patient. The present case reminds us of the rare possibility of renal angiomyolipoma, which may invade the inferior vena cava or right atrium and cause the pulmonary embolism.