AJR FOCUS ON:

# Renal Angiomyolipoma With Nontraumatic Pulmonary Fat Embolus

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A 31-year-old previously healthy man developed acute onset of chest palpitations, tachycardia, and chest pain. There was no recent history of trauma. At presentation, he was hypoxic, with oxygen saturation of approximately 90% on room air. CT angiography was performed, revealing a large pulmonary embolus containing macroscopic fat in the right pulmonary artery (Fig. 1A). A subsequent contrast-enhanced CT of the abdomen and pelvis showed a 5 × 6 × 4 cm lobulated hypoattenuating mass with thin internal septations in the upper pole and pelvis of the left kidney, invading the left renal vein, and extending into the inferior vena cava (IVC) (Fig. 1B). According to measured attenuation, the mass was composed predominantly of macroscopic fat. There were no other masses or enlarged lymph nodes. The preoperative differential diagnosis included angiomyolipoma and liposarcoma. Histology from specimens from concurrent nephrectomy and embolectomy confirmed the diagnosis of a benign angiomyolipoma (Fig. 1C).

Angiomyolipoma is the most common benign renal mesenchymal neoplasm arising from the perivascular epithelioid cells (also known as PEComas) and contains a variable proportion of blood vessels, smooth muscle, and adipose tissue [1]. Angiomyolipoma can degenerate into a malignant form known as epithelioid angiomyolipoma; this rare subtype does not contain macroscopic fat and radiographically resembles renal cell carcinoma [2]. Growth into the renal vein is an unusual, but known, complication of benign angiomyolipoma and does not imply malignant potential or degeneration. A review by Islam et al. [3] in 2004 of 26 patients with renal angiomyolipoma invading the IVC suggested that angiomyolipomas are usually found incidentally in middle-aged patients, either as sporadic, often symptomatic, solitary tumors or in association with tuberous sclerosis, in which they are more likely to be multiple, small, and asymptomatic. Tumors with IVC invasion are generally larger than noninvasive angiomyolipomas and are more likely to be symptomatic.

**Keywords:** angiomyolipoma, fat embolus, pulmonary embolus

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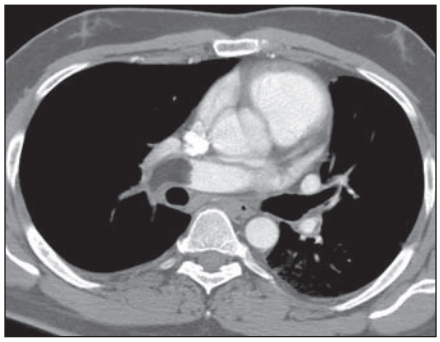
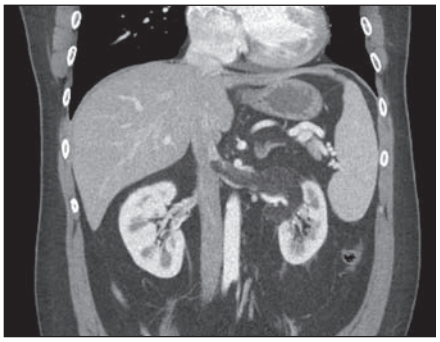


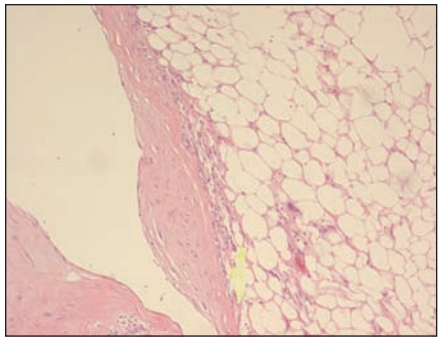
A



B

**Fig. 1**—31-year-old man with acute onset of chest palpitations, tachycardia, and chest pain.
**A,** Axial contrast-enhanced pulmonary CT angiogram reveals embolus in right main pulmonary artery with CT attenuation value of –80 HU, consistent with macroscopic fat.
**B,** Coronal reformation image from contrast-enhanced CT of abdomen shows fatty mass in left kidney, expanding left renal vein, and extending into inferior vena cava.

**(Fig. 1 continues on next page)**



C

**Fig. 1 (continued)**—31-year-old man with acute onset of chest palpitations, tachycardia, and chest pain. **C,** Pathology showed benign angiomyolipoma in both renal mass and embolus. Photomicrograph from surgical explant shows adipose and muscular components, consistent with angiomyolipoma in left kidney and renal vein. (H and E)

Although benign angiomyolipoma, angiomyolipoma with malignant epithelioid degeneration, and liposarcoma invading the renal vein and IVC have been reported, we know of no cases in the recent English-language literature of these entities presenting as a fat embolus in the pulmonary vasculature. Fat embolus syndrome is a known sequela of traumatic long-bone fracture or orthopedic instrumentation [4]. In such cases, however, the diagnosis is primarily clinical because macroscopic fat is rarely apparent at CT. In the case presented here, the tumor remained largely intact after embolization, and there was no clinical evidence of fat embolism syndrome. The patient did not display symptoms of respiratory distress during hospitalization and was discharged on the 10th hospital day after an uneventful postoperative course.

## References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