

## Ruptured Angiomyolipoma with Fatty Tumor Thrombus of the Renal Vein and Inferior Vena Cava

Ching-Hwa Yang, MD; Pei-Hui Chan, MD; Siu-Kei La, MD; Kuan-Ming Chiu<sup>1</sup>, MD

Renal angiomyolipoma is a benign clonal neoplasm that consists of three constituents, namely, blood vessels, adipose tissue and smooth muscle. Although renal angiomyolipoma has a benign nature and grows slowly, extrarenal occurrences have been reported in the hilar lymph nodes, retroperitoneum, liver, colon and direct extension to the renal vein and inferior vena cava as a tumor thrombus. Such a tumor thrombus is not an uncommon preoperative finding in renal cell carcinoma but is a rare presentation in renal angiomyolipoma. We present a case and review the literature, noting that tumor thrombus occur in sizable angiomyolipoma (> 4 cm), right kidneys and, predominantly, in females. The potentially lethal nature of a vena cava thrombus prompted radical surgery. (*Chang Gung Med J* 2006;29(4 Suppl):21-4)

**Key words:** angiomyolipoma (AML), thrombus, inferior vena cava (IVC).

A previously healthy 58-year-old man suffered from sudden onset severe right flank pain and was sent to our Emergency Room. Physical examinations revealed a palpable right flank mass and vital signs that were in shock state (blood pressure: 80/60 mmHg). Transfusion and emergency computed tomography (CT) scanning were arranged. The scan demonstrated a huge right renal mass with high fat content, and an extended, sizable region of hemorrhage within the renal mass and pararenal area was noted (Fig. 1). CT also demonstrated a low density tumor thrombus extending through the right renal vein into the inferior vena cava (IVC) (Fig. 2). Under the impression of a ruptured right renal tumor, emergency laparotomy with right radical nephrectomy was performed. During surgery, a ruptured right renal tumor with dominant adipose tissue was found. The renal vein contained palpable tumor thrombus extending into the IVC. The tumor thrombus was extracted by venacavotomy (Fig. 3). The main renal tumor measured up to 12 cm in diameter and had a

rupture anterolaterally through which hemorrhagic tumor protruded, with a lot of old blood clot. Microscopically, the renal mass showed a mixture of fat, smooth muscle and thick-walled vessels diagno-



**Fig. 1** Ruptured right renal tumor with dominant fat component (long arrow). Large area of high density hemorrhage (short arrow) is seen within the tumor.

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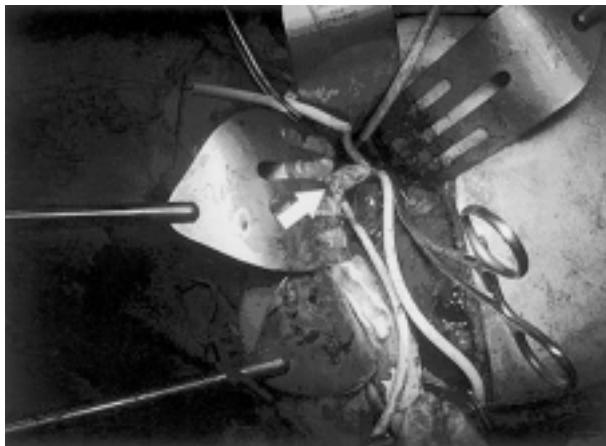
From the Division of Urology, Department of Surgery. <sup>1</sup>Department of Cardiovascular Surgery, Far Eastern Memorial Hospital, Taipei.

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Correspondence to: Dr. Ching-Hwa Yang, Division of Urology, Department of Surgery, Far Eastern Memorial Hospital, 21, Sec. 2, Nan-Ya S. Road, Pan-Chiao, Taipei, Taiwan 220, R.O.C. Tel.: 886-2-2954-6200 ext. 1612, 1634; Fax: 886-2-2-2954-5567; E-mail: uroy@mail.femh.org.tw



**Fig. 2** A low density tumor in the inferior vena cava.



**Fig. 3** Extraction of thrombus by venacavotomy.

tic of angiomyolipoma. The IVC thrombus was unorganized fatty tissue. The patient was discharged seven days postoperatively and remains well to date. He has no mental or physical features of tuberous sclerosis.

## DISCUSSION

There are two types of angiomyolipomas (AML).<sup>(1)</sup> One type is seen in about 50 per cent of patients with tuberous sclerosis complex and are small, asymptomatic, usually bilateral and found only at autopsy. The other type usually presents as a large ( $> 4$  cm), unilateral, symptomatic renal mass, frequently associated with intrarenal or retroperitoneal hemorrhage.<sup>(2)</sup> Local progression may also result in nodal involvement<sup>(3,4)</sup> or intravascular tumor

thrombosis.<sup>(5)</sup> In rare cases, distant extrarenal involvement of the liver<sup>(6)</sup> or colon<sup>(7)</sup> has been reported.

The capabilities of intravenous pyelogram, ultrasound, angiography and CT in the diagnosis of AML have been described widely; the typical CT and magnetic resonance imaging (MRI) picture, with areas having density corresponding to fatty tissue, accurately show the presence of AML. However, in atypical AML or epithelioid AML, there is no viable adipose tissue and it might be diagnosed as renal cell carcinoma initially. Immunohistochemical study (HMB45) usually show positive stain in these cells.<sup>(6)</sup> Macroscopic renal vein and vena cava tumor thrombus rarely occur in patients with AML and may be diagnosed preoperatively by the demonstration of abundant fat within the tumor components.<sup>(9)</sup> There have been about 13 cases of AML with IVC involvement according to the literature.<sup>(5,8)</sup> They occurred in sizable ( $> 4$  cm) AML, right kidneys and, predominantly, in females. The progression of AML may be a slow development<sup>(5)</sup> or rapid growth, extending into the renal vein and IVC.<sup>(8)</sup> Tumor thrombus extending into the right atrium has been reported.<sup>(10)</sup> The biological significance of these findings were unclear, it is possible that the tumor could spread across vascular spaces into the vein, particularly if it arose from the wall of a vein and extended locally into the renal vein and IVC. Patients with AML greater than 4 cm in size, who are asymptomatic, may be followed conservatively using CT or ultrasonography annually but should be alerted to the possible life-threatening complications of hemorrhage or rupture. The presence of a vena cava tumor thrombus with potential lethal complications necessitates surgical intervention.

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# 腎臟的肌肉血管脂肪過誤瘤發生破裂合併腎靜脈及下腔靜脈的栓塞

楊清華 陳沛輝 黎兆祺 邱冠明<sup>1</sup>

腎臟的肌肉血管脂肪過誤瘤因包含三種不同胚胎層的成分——肌肉、血管、脂肪而得名。臨牀上，它是一種良性的腎臟腫瘤，生長得較慢，但是也有報告指出它會侵犯周圍的淋巴腺，甚至擴展至肝臟、大腸。若是腫瘤侵犯至腎靜脈和下腔靜脈，而形成栓塞時，常會在手術前被誤為是惡性的腎臟癌合併下腔靜脈的轉移。本文報告一例腎臟的肌肉血管脂肪過誤瘤合併下腔靜脈的栓塞，並回顧文獻，發現此類腫瘤均為較大的腫瘤（> 4 公分），且位於右側，大部分為女性病患。由於腎臟的肌肉血管脂肪過誤瘤若合併腎靜脈及下腔靜脈的栓塞，可能引起致命的後遺症，應即早手術根除。（長庚醫誌 2006;29(4 Suppl):21-4）

**關鍵字：**腎臟肌肉血管脂肪過誤瘤，栓塞，下腔靜脈。

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板橋亞東醫院 外科部 泌尿外科，<sup>1</sup>心臟血管外科

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索取抽印本處：楊清華醫師，板橋亞東紀念醫院 外科部 泌尿外科。台北縣220板橋市南雅南路2段21號10樓。Tel.: (02)29546200 ext. 1612, 1634; Fax: (02)29545567; E-mail: uroy@mail.femh.org.tw