CT Finding of Renal Vein Invasion by Aggressive Renal Angiomyolipoma: a case report

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This 46-year-old female was told to have a right renal tumor several years ago in a health examination. Follow-up computed tomography images revealed a fat-containing tumor with invasion of right renal vein and inferior vena cava. Right radical nephrectomy and cavotomy were performed and aggressive renal angiomyolipoma was impressed. We found the characteristic imaging finding of early renal vein invasion of the aggressive renal angiomyolipoma by comparison of the serial follow-up images.

Key words: Angiomyolipoma; Computed Tomography; Inferior Vena Cava; Kidney Neoplasms; Renal Vein

Renal angiomyolipoma (AML) is generally considered to be a benign neoplasm, however aggressive behavior and malignant renal angiomyolipoma do exist [1]. Aggressive renal angiomyolipoma is generally defined if renal AML invades the renal vein [2]. Although some case reports of aggressive renal AML had been published, there is no discussion about the disease process of renal vein invasion and its imaging finding to our knowledge till now. By comparison of the serial follow-up computed tomography images of this case, the characteristic imaging finding of early renal vein invasion by aggressive renal AML was proposed.

CASE REPORT

This 46-year-old female was well before and denied any systemic disease. In an ordinary health examination several years ago, a benign right renal tumor was told and sonography follow-up was recommended. However, progressive enlargement of the tumor mass was noted in the follow-up sonography. Thus further computed tomography was suggested and performed in a medical center. An exophytic mass of heterogeneous fat-density at lower pole of right kidney was noted (Fig. 1). Relatively well-defined margin, prominent internal vessels and fine soft tissue-density strands within the mass were identified. There were also fatty nodules in peri-tumorul renal medulla and tubular fatty masses in renal hilum (Fig. 2). The medullary fatty nodules seemed to be continuous with the hilar tubular fatty masses. Besides, extension of the fatty mass in the renal vein to inferior vena cava was also identified (Fig. 3). Therefore an exophytic angiomyolipoma at lower pole of right kidney with tumor thrombi in right renal vein and inferior vena cava was impressed. Surgery was suggested at that time. But the patient refused because she did not feel any associated discomfort. Follow-up computed tomography two years later showed interval growth of the

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**Figure 1.** Postcontrast axial CT scan of R’t kidney on 2002/02/07. 

**a.** A well-defined fat-density mass at lower pole is shown (arrow). 

**b.** Exophytic growth with well-defined margin is identified. Strong nodular enhancements indicate vascular components (arrow). Strand densifications indicate fine vessels or smooth muscle components.

**Figure 2.** Sequential enhanced axial CT scan of R’t kidney on 2002/02/07 reveals the fatty nodules in peritumoral medulla (arrows) and tubular fatty masses in renal hilum (arrowheads) which are proved to be tumor thrombi in segmental renal veins pathologically.

**Figure 3.** Postcontrast axial CT scan on 2002/02/07 reveals the fat-density thrombi in right renal vein (arrows in 3a) and in inferior vena cava (arrow in 3b).
lower pole tumor mass, fatty nodules in peritumoral medulla and tumor thrombi in renal vein and inferior vena cava (Fig. 4). Further cephalic extension of the tumor thrombi in inferior vena cava to the level of diaphragm was also noted. However, she started to suffer from mild dyspnea and right flank fullness sensation in recent 6 months. Computed tomography revealed progressive growth of the lower pole tumor mass with fatty tubular connection to the bulky fatty mass in the renal hilum (Fig. 5). Progressive enlargement of the fatty tumor thrombi in inferior vena cava was also identified. The patient had no flank pain, gross hematuria, chest pain or exertional dyspnea. Normal urine analysis and normal routine laboratory data were appreciated. Because of mild dyspnea and fear of further cephalic progression of tumor thrombi to right atrium, right radical nephrectomy, paracaval lymph node dissection and cavotomy with removal of tumor thrombi were performed. The surgeon found two yellowish tumors over middle pole and lower pole of right kidney, 4.5 cm × 4 cm × 3 cm and 8 cm × 6 cm × 5 cm respectively. Two columns of tumor thrombi in inferior vena cava were also removed. The sizes were 8 cm × 1 cm and 4 cm × 0.8 cm individually. The pathological report was consistent with an exophytic angiomyolipoma at lower pole of right kidney with renal vein invasion. The middle pole tumor was a conglomeration of tumor thrombi within the lumens of hilar renal veins. The lower pole tumor was confined in renal parenchyma and capsule. No apparent necrosis or hemorrhage was noted. The hilar soft tissue, perirenal adipose tissue and dissected lymph nodes were grossly uninvolved.

**DISCUSSION**

Renal AML is a benign neoplasm consisting of the admixture of adipose tissues, thick-walled vessels and spindle cell elements in various proportions. Therefore they are categorized into predominantly lipomatous, predominantly leiomyomatous and epithelioid variants in pathology [2]. Clinically, they
Aggressive renal angiomyolipoma

may present as the common sporadic entity or less common tuberous sclerosis-associated entity. Even though renal AML is generally regarded as a benign neoplasm, aggressive behavior and malignant renal AML have been proposed [1]. Aggressive renal AML is defined if concomitant renal vein invasion has occurred. Right side location, large size and central encroachment were proposed to be contributing factors of renal vein invasion [3]. Although no specific histological spectrum is found in this entity, loss of calponin h1 and h-caldesmon proteins (the marker protein in the late phase of smooth muscle cell differentiation) had been proposed to be related to the aggressiveness of some renal aggressive AMLs [2, 3]. Malignant renal AML is defined if concomitant distant metastasis has occurred [1]. Although malignant cytological features (cellular pleomorphism, nuclear atypia, increased mitotic activity or necrosis) are frequently identified in the malignant renal AMLs, currently the only acceptable criterion for malignant AML is distant metastasis [4]. In the cases with lymph node involvement, they could be explained by either multicentricity of the disease or metastasis [1].

In CT imaging, we could confidently diagnose renal AML by using the criteria of well-defined margin, fat-density component, a sharp parenchyma defect, engorged internal vessels and presence of additional AMLs [5]. However other unusual fat-containing tumors of the kidney should be carefully differentiated [6], which consisted of small renal cell carcinoma with osseous metaplasia (presence of small foci of marrow fat and bone), large renal cell carcinoma with lipid-producing necrosis (presence of little amount of fat), renal cell carcinoma with entrapment of renal sinus fat or perirenal fat, oncocytoma with osseous metaplasia (presence of small foci of marrow fat and bone), teratoid Wilms tumor (presentation in childhood with teratoid components such as fat, cartilage and bone) and liposarcoma (usually arising from renal sinus fat or perirenal fat without causing renal parenchymal defect, except in some aggressive types).

This case is a predominantly lipomatous variant with aggressive behavior. In addition to the presentation of the rare aggressive renal AML, we also found the possible characteristic imaging finding of early renal vein invasion. We found that the small fatty nodules in the renal medulla adjacent to the tumor mass progressively enlarged and were continuous with the bulky fatty masses in the renal hilum which were proved to be tumor thrombi within segmental renal veins. Thus we proposed that these small satellite fatty nodules in the peritumoral medulla may indicate invasion of the interlobar veins. We also noted that the tumor thrombi in the inferior vena cava only extended cephaladly. This could be explained that the relatively fast centripetal blood flow in the inferior vena cava will force the tumor thrombi to grow and extend cephaladly.

However, the initial imaging presentation of this case already revealed tumor thrombi in hilar renal veins and inferior vena cava in addition to the satellite fatty nodules. Therefore follow-up studies of renal AML cases with only peritumoral fatty nodules in the medulla are necessary to confirm this characteristic imaging finding.

**CONCLUSION**

Presence of the small satellite fatty nodules at renal medulla adjacent to the angiomylipoma may indicate invasion of the parenchymal renal veins by the rare aggressive variant. This characteristic imaging finding should alert surgeons for early surgical intervention because at that time partial nephrectomy might be performed instead of radical nephrectomy and the mortality and morbidity of cavotomy for tumor thrombi in the inferior vena cava or right atrium could also be avoided.

**REFERENCE**

侵犯性腎臟血管肌肉脂肪瘤之電腦斷層發現：
病例報告

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侵犯性腎臟血管脂肪瘤是一種罕見的腎臟腫瘤，其與良性腎臟血管肌肉脂肪瘤的診斷差別
在於有無侵犯腎臟血管。藉由觀察此病例的長期追蹤影像，我們觀察到腫瘤侵犯血管的早期電
腦斷層影像特徵。如此一來可提供早期診斷與併發症較少的早期手術治療。

關鍵詞：血管肌肉脂肪瘤；電腦斷層攝影；下腔靜脈；腎臟腫瘤；腎靜脈