Renal Angiomyolipoma with Lymph Node Involvement: A Case Report and Literature Review

Wei-Yu Lin, MD; Cheng-Keng Chuang, MD, PhD; Kwai-Fong Ng[1], MD; Shuen-Kuei Liao[2], PhD

Renal angiomyolipomas (AMLs) are relatively common tumors containing fatty tissue, blood vessels, and muscular cells in various proportions. Typical AMLs can be diagnosed without histological confirmation by a combination of ultrasound and computed tomography imaging in up to 95% of cases. In contrast, simultaneous involvement of the kidney, renal vein, or lymph nodes is uncommon and might be confused with a metastasizing malignant tumor. We present a pathologically proven case of the very uncommon simultaneous involvement of the kidney and the lymph nodes in AML. *(Chang Gung Med J 2003;26:607-10)*

Key words: angiomyolipoma, lymph node, kidney.

Renal angiomyolipomas (AMLs) are relatively common tumors containing fatty tissue, blood vessels, and muscular cells in various proportions. They have a benign clinical course, with a slow growth pattern and lack of distant metastasis. However, a propensity for aggressive local growth has been described, with invasion of the renal calyceal and pelvic spaces, extension of the renal capsule, involvement of the renal vein and inferior vena cava by a fatty thrombus, and the presence of lymph nodes at the renal hilum with angiomyolipoma tissue.

Typical AMLs can be diagnosed without histological confirmation by a combination of ultrasound (US) and computed tomography (CT) imaging in up to 95% of cases. In contrast, simultaneous involvement of the kidney, renal vein, or lymph nodes is less known and might be confused with a metastasizing malignant tumor. We present a pathologically proven case of the very uncommon simultaneous involvement of the kidney and lymph nodes in AML.

CASE REPORT

A 37-year-old woman, with a 2-week history of left flank pain, was referred for evaluation of a left renal mass discovered during US of the abdomen. The physical examination was unremarkable, and laboratory evaluations of hematological as well as serum chemistry parameters were within normal limits. However, CT revealed a heterogenous mass in the left kidney measuring 7×5×5 cm, with suspected invasion of the left renal vein. Enlargement of the hilar lymph nodes was also noted (Figs. 1, 2). A radical left nephrectomy was performed under the impression of renal cell carcinoma with renal vein involvement. Para-aortic lymph nodes were noted during the operation. Grossly, the tumor was confined within the renal capsule.

The major histological components of the tumor and lymph nodes were characterized by abnormally thick-walled vessels, mature fat cells, and smooth muscle bundles (Figs. 3, 4). Consequently, the diagnosis was a multicentric angiomyolipoma.

From the Division of Urology, Department of Surgery; [1]Department of Pathology, Chang Gung Memorial Hospital, Taipei; [2]Graduate Institute of Clinical Medical Science, College of Medicine, Chang Gung University, Taoyuan.
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Address for reprints: Dr. Cheng-Keng Chuang, Division of Urology, Department of Surgery, Chang Gung Memorial Hospital. 5, Fushing Street, Gueishan Shiang, Taoyuan, Taiwan 333, R.O.C. Tel.: 886-3-3281200 ext. 2103; Fax: 886-3-3285818; E-mail: chuang89@cgmh.org.tw

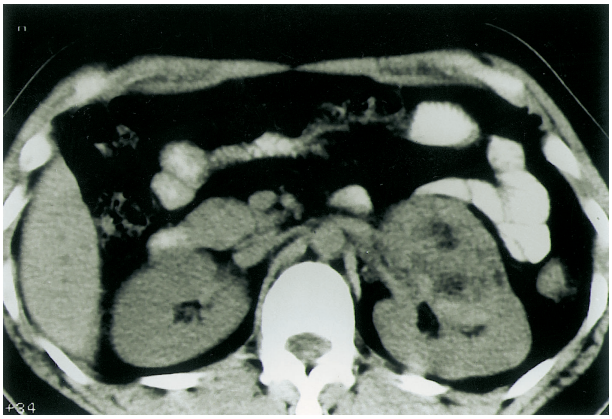

Fig. 1 Non-contrast-enhanced CT.

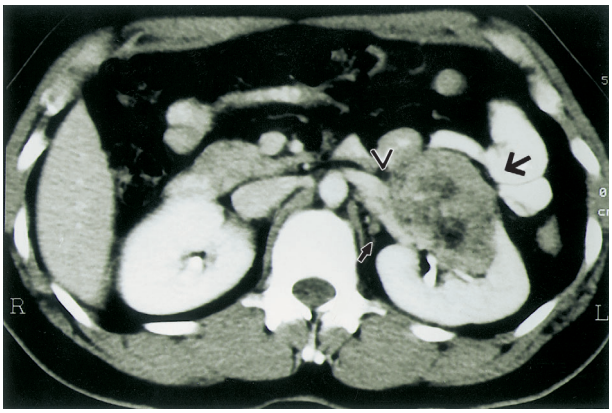

Fig. 2 Contrast-enhanced CT. There is a heterogenous enhanced tumor (7×5×5 cm) (arrow) of the left kidney. There is invasion of the left renal vein by the tumor (arrowhead). The hilar lymph node can be noted (short arrow).

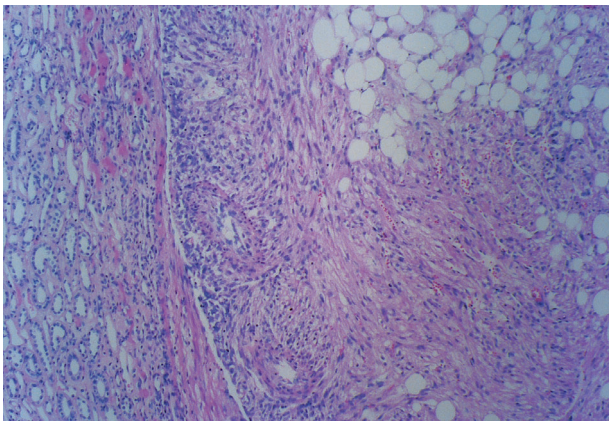

Fig. 3 Renal angiomyolipoma with 3 classic elements. (H&E, reduced from ×100)

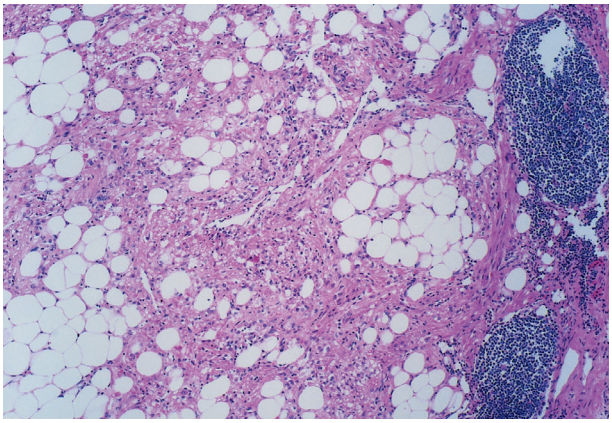

Fig. 4 Lymph node with involvement by an angiomyolipoma.

DISCUSSION

Most cases of AML with its typical appearance of echogenicity are diagnosed incidentally by the widespread use of ultrasound. In ultrasonography, the echogenicity of the AML depends on the proportion of fat in the renal mass. A typical renal AML is suggested by renal masses that are markedly hyperechoic relative to the renal parenchyma. In hyperechoic renal masses, the presence of acoustic shadowing, a hypoechoic rim, and intratumoral cysts are important findings that help distinguish AML from renal cell carcinoma (RCC).[1-4] Meanwhile, demonstration of fat within the tumor by CT (-20 to -80 Hounsfield units) imaging is thought to confirm the diagnosis.[5]

Although most fat-containing renal tumors are not RCC, the appearance of fat in a renal mass should not rule out this diagnosis when calcification is identified within the mass or when other features of the mass strongly suggest malignancy.[6-8]

Diagnosis becomes uncertain when the amount of fat in the tumor is too low to be clearly imaged. The use of 5-mm-thick, non-enhanced CT sections increases the chance of establishing the presence of fat within the tumor.[9]

Lymph node involvement in AML represents a multifocal version of the tumor rather than a metastatic disease, a belief that arose from the benign appearance of the tumor and lymph nodes on pathological examination and the lack of evidence of distant spread on follow-up.[10] No recurrence or meta-

synchronous growth after complete resection has been described.

Even if the typical features of an angiomyolipoma are present, para-aortic lymph adenopathy may increase suspicions of a more-aggressive tumor, possibly leading to unnecessarily aggressive surgery.

Despite improvements in current imaging methods, multicentric AML of the kidney remains a difficult preoperative diagnosis in some cases, not least due to its very low prevalence. Any metastasizing malignant disease should be considered. Because a preoperative diagnosis cannot be made with certainty, renal parenchyma-sparing surgery should not be a therapeutic option unless the pathology proves it to be benign.

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腎臟血管肌肉脂肪瘤併淋巴結侵犯：病例報告及文獻回顧

林威宇 莊正鏗 吳桂芳[1] 廖順奎[2]

腎臟血管肌肉脂肪瘤是由脂肪組織、血管及肌肉細胞依不同比例所構成。典型的血管肌肉脂肪瘤不需要組織細胞學的確認即可藉由超音波及電腦斷層診斷出百分之九十五的病例。相反的，同時侵犯腎臟及淋巴結的血管肌肉脂肪瘤較不被瞭解，並且常和惡性轉移腫瘤混淆。我們報告一位病例——病理學上確認同時侵犯腎臟及淋巴結的血管肌肉脂肪瘤。(長庚醫誌 2003;26:607-10)

關鍵字：血管肌肉脂肪瘤，淋巴結，腎臟。

長庚紀念醫院 台北院區 外科部 泌尿一科，[1]病理科；[2]長庚大學 臨床研究所
受文日期：民國91年9月16日；接受刊載：民國92年1月10日。
索取抽印本處：莊正鏗醫師，長庚紀念醫院 外科部 泌尿一科。桃園縣333龜山鄉復興街5號。Tel.: (03)3281200轉2103; Fax: (03)3285818; E-mail: chuang89@cgmh.org.tw