Title	Angiomyolipoma with regional lymph node involvement: a case report and literature review
Author(s)	Shiga, Yoshiyuki; Tsutsumi, Masakazu; Suzuki, Koichiro; Ishikawa, Satoru; Shimogama, Tatsuro
Citation	泌尿器科紀要 (2003), 49(2): 81-86
Issue Date	2003-02
URL	http://hdl.handle.net/2433/114918
Right	
Type	Departmental Bulletin Paper
Textversion	publisher

ANGIOMYOLIPOMA WITH REGIONAL LYMPH NODE INVOLVEMENT: A CASE REPORT AND LITERATURE REVIEW

Yoshiyuki SHIGA
From the Department of Urology, Tsukuba Central Hospital

Masakazu TSUTSUMI, Koichiro SUZUKI and Satoru ISHIKAWA
From the Department of Urology, Hitachi General Hospital

Tatsuro SHIMOGAMA
From the Department of Pathology, Hitachi General Hospital

A 28-year-old man without tuberous sclerosis, who complained of pollakisuria, consulted to our hospital for a left renal mass. Abdominal computed tomography revealed a solid mass without a lipid component, 10 cm in diameter, in the left kidney and with regional lymphadenopathy. Renal arteriography showed a hypervascular mass, demonstrating multiple tumor stains and aneurysms. Left radical nephrectomy and perihilar lymph node dissection were performed for an anticipated diagnosis of malignant tumor in November 2001. The histopathological diagnosis was an angiomyolipoma with lymph node involvement. Immunostaining for myogen markers was positive in the renal mass and lymph node tumors. He was free from disease ten months after surgery.

(Acta Urol. Jpn. **49**: 81–86, 2003)

Key words: Angiomyolipoma, Lymph node involvement

INTRODUCTION

Angiomyolipoma (AML) of the kidney is a hamartoid tumor, its histology consisting of a variable mixture of proliferating blood vessels and mature fat and smooth muscle cells. Recently, AML with regional lymph node involvement has been reported as a benign tumor, and approximately 40 such cases have been recorded. Nodal involvement in association with renal AML is more likely to represent a multicentric growth pattern than true metastases.

CASE REPORT

A 28-year-old man with slight mental retardation and without neurocutaneous symptoms, who complained of pollakisuria, was referred to our hospital because of a left renal mass detected by ultrasonography (US). The mass was not palpable in the left upper abdominal quadrant and laboratory studies were all within normal limits. The US showed a low-echoic, homogenous huge mass in the left upper quadrant. Enhanced computed tomography (CT) revealed a solid tumor of 10 cm in diameter, with no definite fatty tissue, in the upper pole of the left kidney with perihilar and paraaortic lymphadenopathy (Fig. 1). A renal arteriogram showed a hypervascular mass with multiple tumor stains and aneurysms (Fig. 2). Left radical nephrectomy was performed for the anticipated diagnosis of renal cell carcinoma (RCC) or an other malignant tumor such as Wilms' tumor in November 2001.

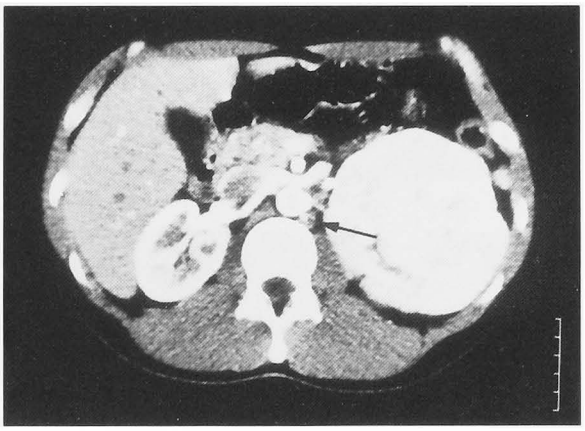


Fig. 1. CT revealed a solid tumor 10 cm in diameter with no definite fatty tissue in the upper pole of the left kidney with perihilar and paraaortic lymphadenopathy.

A cross section of the resected specimen showed a pinkish-white mass, weighing 750 g. Microscopically, the tumor consisted almost entirely of smooth muscle, vascular tissues and minimal adipose elements, consistent with angiomyolipoma (Fig. 3a). The involved lymph nodes had the same characteristics as the renal mass (Fig. 3b). Immunostaining for myogen markers such as smooth muscle actin (SMA) and desmin, as well as HMB-45 (originally described as an anti-melanoma monoclonal antibody) was positive in the tumors and involved lymph nodes (Fig. 4a, b). He made an uneventful recovery and was discharged from our hospital on the seventh post-operative day. Follow-

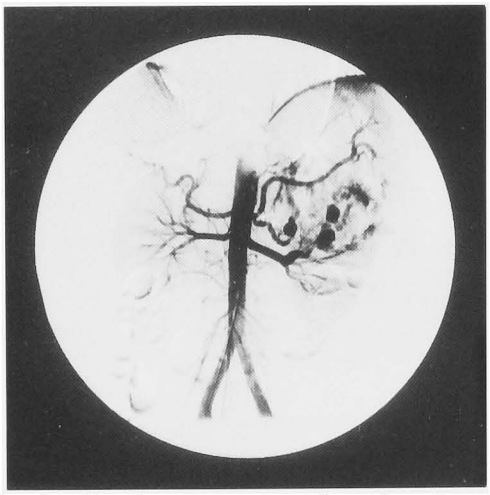
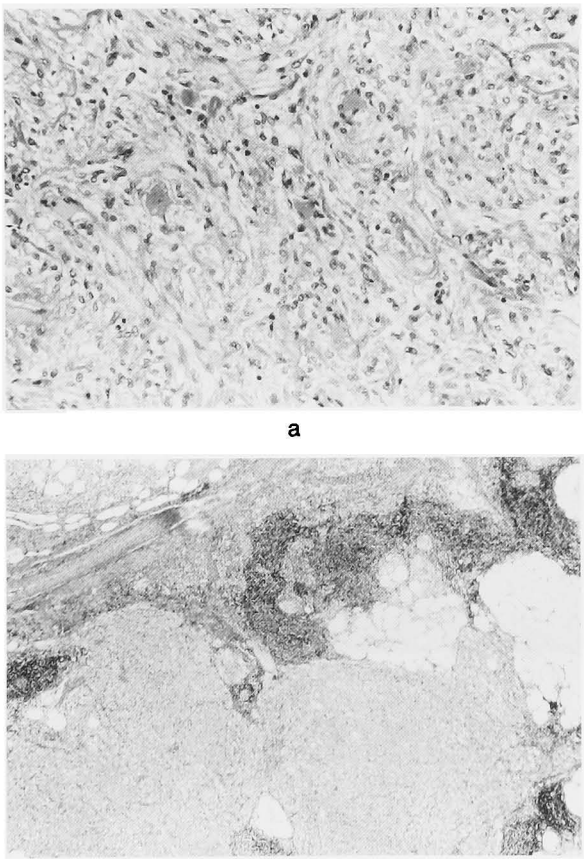


Fig. 2. Renal arteriogram showed a hypervascular mass, demonstrating multiple tumor stains and aneurysms.



a

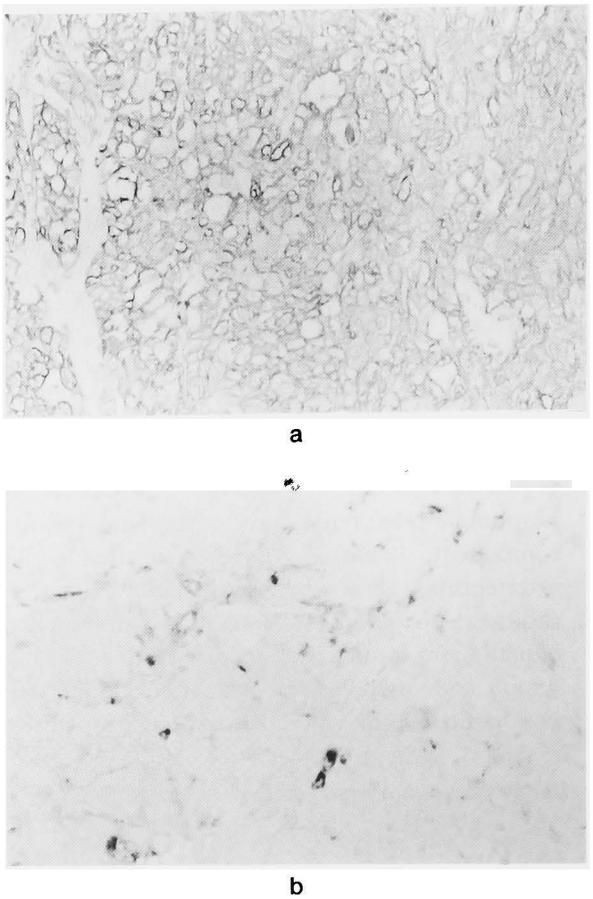
b

Fig. 3. a: The renal mass consisted almost entirely of smooth muscle and vascular tissue and had minimal adipose elements in the small lesion (HE stain ×50). b: The lymph nodes had the same characteristic as AML (HE stain ×10).

up US and CT four months postoperatively demonstrated no evidence of recurring or metastatic disease.

DISCUSSION

Reports of approximately 40 such cases[1–31] of AML



a

b

Fig. 4. Immunostaining of SMA and HMB-45 was positive in the renal mass (a: SMA stain ×75) and involved lymph nodes (b: HMB-45 stain ×75).

involving the lymph nodes have been published in the English and Japanese literature, as shown in Table 1. It is thought that the actual incidence (1–2% of all AML) of nodal involvement is underestimated[11], because of few radical nephrectomies for AML, since preoperative detection has become more accurate. The table demonstrates that tumors associated with tuberous sclerosis (TS) are usually bilateral and multiple and are diagnosed in patients at an average age of 29.8 years, ranging from 9 to 73 years. AMLs in patients without TS are usually single and unilateral and are diagnosed an average of 14 years later than those associated with TS. Additionally, the female-male ratio of the former is almost even, in contrast, the latter group is 3 : 1. In our case, a brain CT demonstrated no calcification of the ventriculus lateralis. Additionally, he has not had any neurocutaneous symptoms. Therefore, he is not considered as TS.

It is important to exclude AML preoperatively for renal mass without definite fatty tissue and with lymphadenopathy. Bosniak and colleagues[10] discussed the importance of thin-slice CT without contrast enhancement as a means of identifying fatty tissue in small renal masses, however, several recent reports have described intratumoral fat in RCC.

Table 1. Renal angiomyolipoma with lymph node involvement in the literature

	Author (report year)	Age/Sex	Side	Specimen source	TS	Prognosis
1	Wilson and Lo (1964)	42/F	Bilat.	Autopsy	(+)	not described
2	Allen and Risk (1965)	52/F	Lt.	Radical nephrectomy	(−)	2 yrs, well
3	Campbell et al. (1974)	28/F	Bilat.	Exploration and biopsy	(+)	not described
4	Snowden (1974)	24/M	Bilat.	Autopsy	(+)	not described
5	Scott et al. (1975)	12/M	Rt.	Radical nephrectomy	(+)	1 yrs, well
6	Busch et al. (1976)	21/M	Rt.	Radical nephrectomy	(−)	not described
7		49/F	Rt.	Radical nephrectomy	(−)	not described
8	Bloom et al. (1981)	19/F	Rt.	Radical niphrectomy	(+)	3 yrs, well
9		11/M	Lt.	Radical nephrectomy	(+)	3 yrs, well
10		24/M	Bilat.	Nephrectomy	(+)	11 yrs, well
11	Imai et al. (1983)	32/F	Bilat.	Autopsy	(+)	5 yrs, dead
12	Amano et al. (1984)	42/F	Lt.	Radical nephrectomy	(+)	34 m, well
13	Ishii et al. (1984)	48/F	Rt.	Radical nephrectomy	(−)	3 yrs, well
14	Dao et al. (1984)	24/M	Lt.	Radical nephrectomy	(−)	1 yrs, well
15	Manabe et al. (1984)	42/F	Lt.	Radical nephrectomy	(−)	3 yrs, well
16	Noguchi et al. (1985)	40/F	Rt.	Radical nephrectomy	(−)	58 m, well
17	Brecher et al. (1986)	63/F	Rt.	Radical nephrectomy	(−)	15 yrs, well
18	Sant et al. (1986)	49/F	Lt.	Radical nephrectomy	(−)	3 yrs, well
19	Nakajima et al. (1986)	22/F	Lt.	Radical nephrectomy	(−)	not described
20	Taki et al. (1987)	35/M	Rt.	Radical nephrectomy	(−)	4 m, well
21	Kuroda et al. (1987)	44/M	Bilat.	Radical nephrectomy	(+)	42 m, well
22	Manabe et al. (1987)	32/F	Lt.	Laparotomy	(−)	not described
23	Taylor et al. (1988)	9/F	Rt.	Heminephrectomy	(+)	20 m, well
24		15/M	Bilat.	Radical nephrectomy	(+)	24 m, well
25		25/F	Rt.	Radical nephrectomy	(+)	8 yrs, well
26	Itakura et al. (1988)	53/F	Lt.	Radical nephrectomy	(−)	not described
27	Nagata et al. (1989)	55/F	Lt.	Radical nephrectomy	(−)	44 m, well
28	Ro et al. (1990)	49/M	Lt.	Radical nephrectomy	(−)	12 m, well
29		53/M	Rt.	Radical nephrectomy	(−)	1 yrs, well
30		58/F	Lt.	Radical nephrectomy	(−)	18 yrs, well
31	Ansari et al. (1991)	46/F	Lt.	Radical nephrectomy	(−)	2 m, well
32	Ackerman et al. (1993)	25/F	Lt.	Radical nephrectomy	(−)	2 yrs, well
33	Tomobe et al. (1993)	72/F	Rt.	Radical nephrectomy	(−)	9 yrs, well
34	Maffezzini et al. (1995)	42/M	Rt.	Lung lobectomy	(+)	not described
35		26/F	Rt.	Radical nephrectomy	(−)	4 yrs, well
36		63/F	Rt.	Radical nephrectomy	(−)	2 yrs, well
37		52/M	Rt.	Radical nephrectomy	(−)	2 yrs, well
38	Csanaky et al. (1995)	38/F	Lt.	Radical nephrectomy	(+)	2 yrs, well
39		39/F	Lt.	Radical nephrectomy	(+)	6 m, well
40		73/F	Lt.	Radical nephrectomy	(+)	6 m, well
41	Frohlich et al. (1999)	31/F	Lt.	Radical nephrectomy	(−)	not described
42	Turker et al. (2000)	40/F	Rt.	Radical nephrectomy	(−)	8 yrs, well
43	Yamamoto et al. (2002)	47/M	Lt.	Autopsy	(−)	3 m, dead
44	Present case (2002)	28/M	Lt.	Radical nephrectomy	(−)	4 m, well

* TS: tuberous sclerosis.

Since no area of low attenuation was observed in the contrast-enhanced CT yielding contiguous 5 mm slices, the Hounsfield numbers were not calculated. By pathological examination, smooth muscle formed the greatest part of the tumor and very little fatty tissue was found.

Regional lymph node involvement in a case of renal AML leads to the question of metastasis versus multicentric origin. Most authors have felt that nodal involvement in AML represents the multicentric nature of the tumor rather than metastatic disease, since these diseases are benign tumors and the lymph nodes on pathological examination, and the lack of evidence of distant spread at more than 10 years follow-up in previous reports[1,12,18] On immunohistochemical study, myogen markers such as desmin, SMA and HMB-45 that are especially reactive in the smooth muscle appearance of AML, were positively stained in the tumors and lymph node tissues in our case. This

positive staining confirmed the benign nature of this tumor. Based on these findings, we diagnosed renal AML with lymph node involvement. Yamamoto et al.[32] reported rapidly progressive malignant epithelioid angiomyolipoma of the kidney, which metastasized at the liver, lymph nodes, lungs and the lumber supine, however, the predominant cell type was epithelioid cells positively stained for HMB-45. Imai et al.[33] reported a case of malignant AML, which metastasized at the virchow lymph node. The pathology of the case revealed severe dysplasia of the smooth muscle cells. At the pathology of our case, epithelioid cells and dysplasia of the smooth muscle cells were not found. Bennington et al.[34] insisted that the incorrect diagnosis of malignancy of the AML is usually based on one or more of following features: 1) The tumors may be multiple or bilateral, or both; 2) the smooth muscle cells exhibit variation in size and shape, nuclear hyperchromatism, mitoses, and bizarre giant cells; and 3) the tumors exhibit what is interpreted as venous invasion. Most authors have maintained this opinion.

Recently, DNA flow-cytometry has been used in the analysis of AML to identify a malignancy[5,19] In general, the diploid DNA content of tissue from the kidney and the regional lymph node tumors supports the benign nature of these tumors. Ro et al.[5] first reported DNA flow-cytometric analysis in three cases of renal AML with lymph node involvement. In their report, the primary kidney tumor and tumors in the lymph nodes of all patients contained diploid DNA. Although the DNA flow-cytometry was not performed in our case, it may be usefel study for the difference from malignancy. Genetic studies may resolve the question of metastasis versus multicentric nature in the future.

AML composed predominantly of smooth muscle may be impossible to distinguish from renal cell carcinoma (RCC) preoperatively. Because of its atypical appearance and the presence of lymphadenopathy such as in our patient, RCC, Wilms' tumor, leiomyosarcoma or other malignancies rather than AML, are more likely to be diagnosed preoperatively.

In conclusion, it can be said that nodal involvement in association with renal AML is more likely to represent a multicentric growth pattern than true metastases. Accordingly, we feel that the prognosis is not grave and further treatment such as radiation or chemotherapy would not be necessary. However, we are going to closely follow-up this patient over the long term.

REFERENCES

1) Bloom DA, Scardino PT and Waisman J: The significance of lymph node involvement in renal angiomyolipoma. J Urol **128**: 1292–1295, 1983
2) Buch FM, Bark CJ and Clyde HR: Benign renal angiomyolipoma with regional lymph node involvement. J Urol **116**: 715–717, 1976
3) Scott MB, Halpern M and Cosgrove MD: Renal angiomyolipoma, two varieties. Urology **6**: 768, 1975
4) Allen TD and Risk W: Renal angiomyolipoma. J Urol **94**: 203, 1965
5) Ro JY, Ayala AG, El-Naggar A, et al.: Angiomyolipoma of the kidney with lymph node involvement. Arch Pathol Lab Med **114**: 65–67, 1990
6) Hulbert JC and Graf R: Involvement of the spleen by renal angiomyolipoma: metastasis or multicentricity. J Urol **130**: 328–329, 1983
7) Sant GR, Ucci AA Jr and Meares EM Jr: Multicentric angiomyolipoma: renal and lymph node involvement. J Urol **28**: 111–113, 1986
8) Wilson GC and Lo D: Tuberous sclerosis: a case with pulmonary and lumph node involvement. Med J Aust **51**: 795–796, 1964
9) Ackerman TE, Levi CS, Lindsay DJ, et al.: Angiomyolipoma with lymph node involvement. Can Assoc Radiol J **45**: 52–55, 1994
10) Bosniak MA, Megibow AJ, Hulnick DH, et al.: CT diagnosis of renal angiomyolipoma: the importance of detecting small mounts of fat. AJR Am J Roentgenal **151**: 497–501, 1988
11) Ansari SJ, Stephenson RA and Mackay B: Angiomyolipoma of the kidney with lymph node involvement. Ultrastruc Pathol **15**: 531–538, 1994
12) Brecher ME, Gill WB and Strauss II FH: Angiomyolipoma with regional lymph node involvement and a long-term follow-up study. Hum Pathol **17**: 962–963, 1986
13) Manabe T, Tasaka Y, Amano M, et el.: Regional lymph node involvement in benign renal angiomyolipoma. Acta Pathol Jpn **34**: 889–893, 1984
14) Campbell EW, Brantley R, Harrold M, et al.: Angiomyolipoma presenting as fever unknown origin. Am J Med **57**: 843, 1974
15) Snowdon JA: Cerebral aneurysm, renal cysts and hamartomas in a case of tuberous sclerosis. Br J Urol **46**: 583, 1974
16) Dao AH, Pinto AC, Kirchner FK, et al.: Massive nodal involvement in a case of renal angiomyolipoma. Arch Pathol Lab Med **108**: 612–613, 1984
17) Manabe T, Moriya T and Kimoto M: Benign renal angiomyolipoma with regional lymph node involvement: report of a case showing enlargement five years after nephrectomy. Acta Pathol Jpn **37**: 1853–1858, 1987
18) Turker KI, Tunc M, Kilicaslan I, et al.: Lymph node involvement by renal angiomyolipoma. Int J Urol **7**: 386–389, 2000
19) Csanaky G, Szereday Z, Mafyarlaki T, et al.: Renal

angiomyolipoma: report of three cases with regional lymph node involvement and/or with renal cell carcinoma. Tumori **81**: 469–474, 1995

20) Maffezzini M, Vlassopoulos G, Simonato A, et al.: Renal angiomyolipoma with extra renal involvement—in vascular, lymph node and perirenal tissue. reports of four cases. Scand J Urol Nephrol **29**: 327–329, 1995
21) Taylor RS, Joseph DB, Kohaut EC, et al.: Renal angiomyolipoma associated with lymph node involvement and renal cell carcinoma in patients with tuberous sclerosis. J Urol **141**: 930–932, 1989
22) Kuroda A, Machida T, Masuda F, et al.: Angiomyolipoma of the kidney with regional lymph node involvement. Acta Urol Jpn **34**: 478–481, 1988
23) Taki Y, Hiura M, Ikai K, et al.: Two cases of angiomyolipoma. Acta Urol Jpn **33**: 562–567, 1987
24) Chawla K, Silber L and Alexander LL: Renal angiomyolipoma with retroperitoneal adenopathy. J Natl Med Assoc **75**: 431–434, 1983
25) Ishii D, Matsuno T and Koyanagi T: A case of coincident renal cell carcinoma and angiomyolipoma in the same kidney. Rinsho Hinyokika **38**: 535–538, 1984
26) Noguchi S, Shuin T, Fujii H, et al.: Angiomyolipoma derived from kidney and hilar lymph-node: a case report. Rinsho Hinyoukika **39**: 491–494, 1985
27) Nakajima H, Tsuji A and Muka K: Angiomyolipoma of the kidney with regional lymph node involvement. Rinsho Hinyokika **40**: 231–233, 1986
28) Nagata K, Yanagi K and Joko K: Renal angiomyolipoma with regional lymph node involvement: a case report. Nishinihon Hinyokika **51**: 577–580, 1989
29) Tomobe M, Sasaki A, Ishikawa S, et al.: A case of renal angiomyolipoma with regional lymph node involvement. Rincho Hinyokika **47**: 397–399, 1993
30) Amano M, Okunobo T, Kawahara H, et al.: Renal angiomyolipoma: report of two cases. including spontaneous rupture. Acta Urol Jpn **30**: 1813–1825, 1984
31) Itakura H, Munakata A, Hayashida M, et al.: Regional lymph node involvement in renal angiomyolipoma. Rinsho Hinyokika **42**: 39–41, 1988
32) Yamamoto T, Ito K, Suzuki K, et al.: Rapidly progressive malignant epithelioid angiomyolipoma of the kidney. J Urol **168**: 190–191, 2002
33) Imai Y, Nakamoto Y, Miki K, et al.: An autopsy case of tuberous sclerosis accompanied by renal failure and systemic angio myolipoma. Nihon Naikagakkai Zasshi **72**: 98–103, 1983
34) Bennington JL and Beckwith JB: Angiomyolipoma. in: Atlas of tumor pathology. Tumor of the kidney, renal pelvis and ureter. Armed forces institute of pathology. Fascicle **12**, 2nd series, Washington, DC: 204–212, 1975

(Received on June 24, 2002
Accepted on September 24, 2002)

和文抄録

腎門部リンパ節にも併発病理所見をみた巨大腎血管筋脂肪腫の1例

つくばセントラル病院泌尿器科
志賀淑之

日立総合病院泌尿器科
堤　雅一，鈴木康一郎，石川　悟

日立総合病院病理
下釜達朗

28歳，男性，主訴は頻尿で軽度精神発達遅延をもち，左腎腫瘤を指摘され当科紹介初診となった．CTでは左腎に10 cm大の充実性腫瘍とリンパ節腫大を認めた．血管造影では濃染像や動脈瘤を多数認めるhypervascular tumorであった．悪性腫瘍の疑いにて2001年11月，根治的左腎摘除術およびリンパ節郭清術を施行した．病理はangiomyolipoma with lymph node involvementであった．左腎腫瘍ならびにリンパ節のミオーゲンマーカー免疫染色が陽性で診断を支持していた．術後4カ月経過した現在，再発を認めていない．

(泌尿紀要 **49**：81-86，2003)