Case Reports

Journal of INTERNATIONAL MEDICAL RESEARCH

Journal of International Medical Research
49(3) 1–6

DOI: 10.1177/03000605211001710
journals.sagepub.com/home/imr
SAGE

Sporadic multiple renal angiomyolipoma with lymph node involvement: a case report and literature review

Xiaobo Ding[1,*], Meizi Cui[2,*], Tiejun Wang[3], Helei Wang[4], Xinyu Wang[5], Wei Qiu[6,#] and Yanbo Wang[5,#]

Abstract

Angiomyolipoma (AML) is a benign tumor that mainly occurs in the kidneys. Simultaneous involvement of the kidney and local regional lymph nodes is very rare and might be misdiagnosed as a metastasizing malignant cancer. In the present study, a 50-year-old woman was referred to our hospital after a routine health screening ultrasound. Sporadic multiple renal AML with lymph node involvement was suspected based on the clinical manifestations and radiologic features. Partial nephrectomy was performed and a para-inferior vena cava lymph node was removed. The pathologic results confirmed multiple AML with lymph node invasion. We also reviewed the English-language literature regarding renal AML with lymph node involvement. We found that middle-aged women were likely to develop this disease and that loin pain was the main presenting feature. Most patients had no history of tuberous sclerosis complex. Radical nephrectomy was the predominant treatment. No local recurrence or distant metastasis occurred in any patients after radical nephrectomy or partial nephrectomy. In conclusion, renal AML with lymph node involvement is rare but can occur in both patients with tuberous sclerosis complex and those with multiple sporadic AML. Partial nephrectomy should be the first-line treatment, after which further treatment is not necessary.

[1]Department of Radiology, The First Hospital of Jilin University, Changchun, Jilin, P.R. China
[2]Department of Cadre Ward, The First Hospital of Jilin University, Changchun, Jilin, P.R. China
[3]Department of Orthopedic Traumatology, The First Hospital of Jilin University, Changchun, Jilin, P.R. China
[4]Department of Gastrointestinal Surgery, The First Hospital of Jilin University, Changchun, Jilin, P.R. China
[5]Department of Urology, The First Hospital of Jilin University, Changchun, Jilin, P.R. China
[6]Department of Hepatobiliary and Pancreatic Surgery, The First Hospital of Jilin University, Changchun, Jilin, P.R. China

*These authors contributed equally to this work.

#These authors contributed equally to this work.

Corresponding author:
Yanbo Wang, The First Hospital of Jilin University, 1 Xinmin Street, Changchun, Jilin 130021, P.R. China.
Email: wangyanb@jlu.edu.cn

Keywords
Angiomyolipoma, renal angiomyolipoma, lymph node involvement, partial nephrectomy, literature review, case report

Date received: 22 January 2021; accepted: 18 February 2021

Introduction

Angiomyolipoma (AML) is a benign tumor that primarily occurs in the kidneys and is composed of blood vessels, smooth muscle, and adipose tissue.[1] Renal AML, also termed hamartoma, is the second most common benign tumor in the kidney and accounts for 3% of renal tumors.[2] Classic renal AML is easily diagnosed based on computed tomography (CT) or magnetic resonance imaging, and in almost all sporadic cases, renal AML presents as only one lesion.[3] However, few cases of renal AML accompanied by regional lymph node involvement have been described. We herein present a case of multiple renal AML co-occurring with para-inferior vena cava (para-IVC) lymph node involvement and a comprehensive review of the literature. Written informed consent was obtained from the patient for publication of this article and accompanying images. The present study protocol was approved by the local ethics committee of The First Hospital of Jilin University.

Case presentation

A 50-year-old woman presented with multiple right renal masses that had been incidentally detected on a health screening ultrasound. The patient was asymptomatic and had no hematuria or weight loss. Her medical history was unremarkable, and she had no signs of tuberous sclerosis complex (TSC). Physical examination revealed no significant abnormalities. The results of routine laboratory tests, including kidney function and blood tests, were normal with the exception of microscopic hematuria. Unenhanced and enhanced CT scans revealed typical multiple renal AML in the middle pole of the right kidney measuring 4 cm in its greatest dimension. Enlargement of the para-IVC lymph node was also present (Figure 1). Open partial nephrectomy (PN) was performed, and the para-IVC lymph node (2 cm in its greatest dimension) was removed. The pathological diagnosis of the renal specimen was AML (Figure 2(a)), and the lymph node showed AML invasion (Figure 2(b)). The patient was followed up with a CT scan every 6 months, and the tumor had not recurred 12 months postoperatively.

Discussion

Renal AML occurs sporadically or accompanies TSC. Typically, renal AML shows a slow benign growth pattern and no signs of distant metastasis.[2] However, renal AML with regional lymph node involvement might suggest potential metastatic characteristics of AML.

We searched English-language articles in the Medline database using the key words "kidney" or "renal," "angiomyolipoma" or "hamartoma" and "lymph node." Twenty-two papers were found. Based on the full text of the articles rather than only the abstracts, 16 papers and 25 cases were found to be relevant[4–19] (Table 1). Busch et al.[4] reported the first two cases of

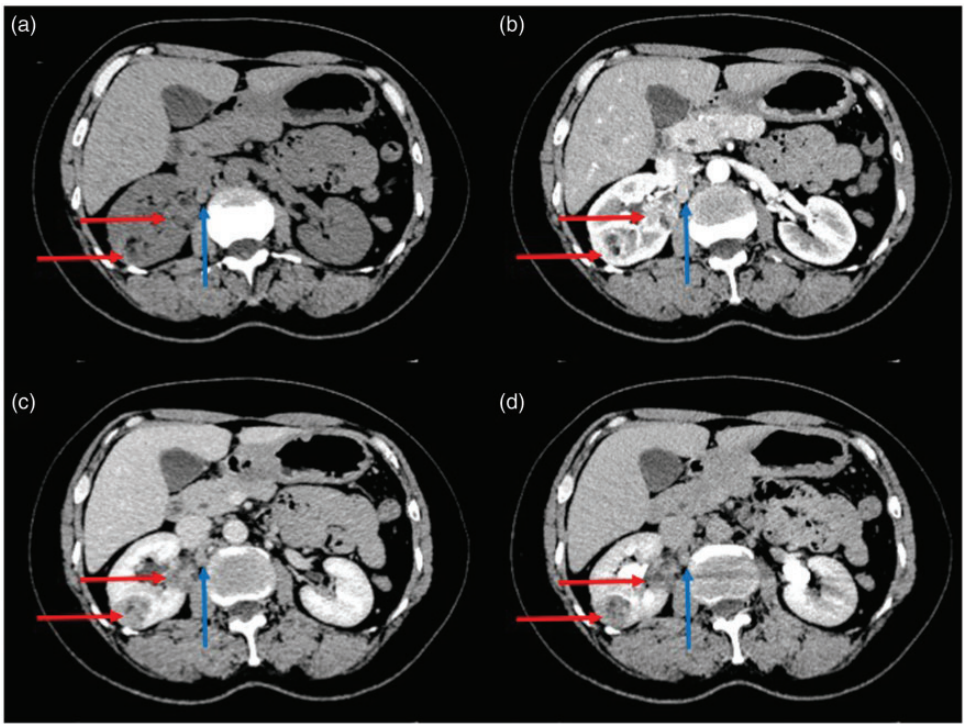

Figure 1. Non-contrast-enhanced computed tomography (a) and contrast-enhanced computed tomography (corticomedullary phase: b, nephrographic phase: c, excretory phase: d). Two heterogeneous enhanced tumors (red arrow) were present in the right kidney. A hilar lymph node was noted (blue arrow).

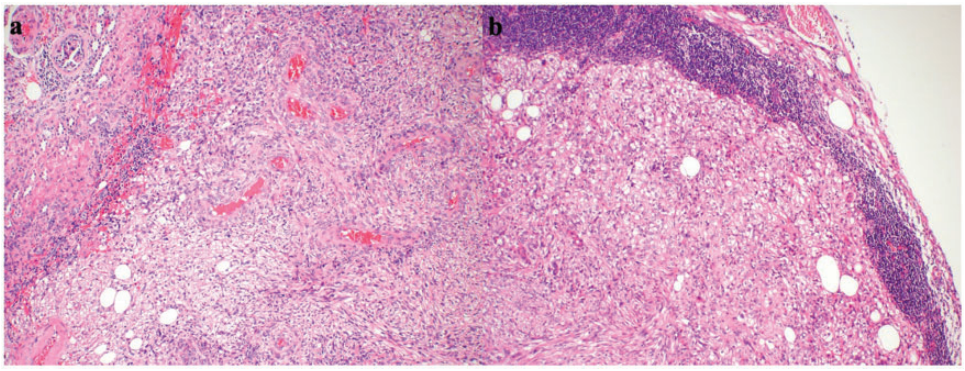

Figure 2. Renal angiomyolipoma. (a) Most of the tumor consisted of smooth muscle, which was interspersed with thick-walled blood vessels and scattered adipose tissues. (b) The remaining components (glomeruli and renal tubules) were visible in the upper left corner (hematoxylin–eosin, ×100).

benign renal AML with regional lymph node involvement in 1976. The mean age of the patients in the 25 reported cases was 40 years and ranged from 9 to 73 years. Women were predominantly affected (female:male ratio of 20:5). Loin pain was the main presenting feature (56%). Most patients had no history of TSC; however,

Table 1. Previously reported cases of renal angiomyolipoma with lymph node involvement.

Case No.	Authors (year)	Age (years)	Sex	Side	Diameter	Symptoms	Treatment	TSC	Evidence of malignancy	Distant metastasis	Follow-up
1	Busch et al. (1976)[4]	21	Male	Right	11 cm	Pain	RN	No	No	No	Unknown
		49	Female	Right	Unknown	Asymptomatic	RN	No	No	No	Unknown
2	Chawla et al. (1983)[5]	28	Female	Right	12 cm	Pain	PN	Yes	Unknown	Unknown	Unknown
3	Manabe et al. (1984)[6]	42	Female	Left	20 cm	Pain	RN	No	No	No	3 years
4	Sant et al. (1986)[7]	61	Female	Left	17 cm	Pain	RN	Yes	No	No	9 years
		49	Female	Left	4 cm	Fatigue	RN	No	No	No	3 years
5	Brecher et al. (1986)[8]	63	Female	Right	6.5 cm	Pain	RN	No	No	No	15 years
6	Manabe et al. (1987)[9]	37	Female	Left	9 cm	Pain	RN	No	No	No	Unknown
7	Taylor et al. (1989)[10]	9	Female	Right	10 cm, 3 cm	Pain	PN	Yes	No	No	20 months
		15	Male	Bilateral	Unknown	Unknown	RN	Yes	No	No	2 years
		25	Female	Right	Unknown	Unknown	RN	Yes	No	No	8 years
8	Ansari et al. (1991)[11]	46	Female	Left	12 cm	Pain	RN	No	No	No	2 months
9	Ackerman et al. (1994)[12]	25	Female	Unknown	Unknown	Asymptomatic	Unknown	Unknown	Unknown	Unknown	Unknown
10	Maffezzini et al. (1995)[13]	42	Male	Right	Unknown	Unknown	RN	Yes	No	No	5 years
		26	Female	Right	15 cm	Pain	RN	No	No	No	4 years
		63	Female	Right	15 cm	Unknown	RN	No	No	No	2 years
		52	Male	Right	2 cm	Unknown	PN	No	No	No	2 years
11	Csanaky et al. (1995)[14]	38	Female	Left	10 cm	Pain	RN	No	No	No	2 years
		39	Female	Left	6 cm	Malaise	PN	Yes	No	No	6 months
		73	Female	Left	4 cm, 2 cm, 2 cm	Asymptomatic	RN	Yes	No	No	6 months
12	Türker Köksal et al. (2000)[15]	40	Female	Right	18 cm	Pain	RN	No	No	No	8 years
13	Göğüş et al. (2001)[16]	17	Female	Left	8 cm	Pain	RN	No	No	No	6 months
14	Lin et al. (2003)[17]	37	Female	Left	7 cm	Pain	RN	No	Unknown	Unknown	Unknown
15	Cui et al. (2011)[23]	47	Male	Left	28 cm	Pain	RN	No	Yes	No	2 years
16	Kon-Nanjo et al. (2017)[19]	62	Female	Left	15 cm	Asymptomatic	RN	No	No	No	10 years

TSC, tuberous sclerosis complex; PN, partial nephrectomy; RN, radical nephrectomy.

eight (32%) cases were associated with TSC. All 25 cases were benign AML rather than malignant epithelioid AML. No distant metastasis occurred in any patients. Radical nephrectomy (RN) was the main treatment, and PN was performed in only four (16%) patients.

Clinically, most cases of AML are sporadic, and 10% to 20% of cases present in association with TSC. Among the 25 published cases of AML with node involvement, 8 (32%) had a history of TSC. Most cases of sporadic AML are asymptomatic, whereas TSC-associated AML is often symptomatic. However, among the 17 published cases of AML with node involvement without TSC, only 2 (12%) were asymptomatic. Moreover, all 17 cases of AML with node involvement without TSC were solitary. Our report describes the first case of sporadic multiple renal AML with lymph node involvement.

Based on the benign nature of AML and the absence of local recurrence and distant metastasis in the present case, the involvement of a local regional lymph node was thought to be simply a multicentric growth instead of metastasis. Tan et al.[20] analyzed the genomic instability of renal AML with regional lymph node involvement in 12 patients and found a potential lineage relationship between renal AML and involved local lymph nodes.

The biological assessment of renal AML is controversial. However, considering that epithelioid renal AML may present with malignant behavior, all patients described in previously published reports were followed up from 2 months to 15 years postoperatively, and no local recurrence or distant metastasis occurred although no further treatment was performed. However, only 16% of the patients in these published studies underwent PN; the others underwent RN. No patients developed local recurrence or distant metastasis, indicating that RN should be avoided when PN is feasible by any approach. Some atypical renal AMLs may be misdiagnosed as renal cancers; thus, biopsy of renal tumors is needed, particularly when small tumor-like lesions are incidentally detected. This may help to avoid unnecessary nephrectomy.[21,22] Additionally, we may be able to hypothesize that PN is the standard treatment for small renal masses without preoperative renal mass biopsy, even with lymph node metastasis. Considering the benign behavior of renal AML with lymph node involvement, an unenhanced CT scan can be performed every 6 to 12 months.

Conclusion

Renal AML with lymph node involvement is rare and can occur in both patients with TSC and those with multiple sporadic AML. PN should be the first-line treatment, after which further treatment is not necessary.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

Funding

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

ORCID iD

Yanbo Wang https://orcid.org/0000-0003-4052-6421

References

1. Kingswood JC, Bissler JJ, Budde K, et al. Review of the tuberous sclerosis renal guidelines from the 2012 consensus conference: current data and future study. *Nephron* 2016; 134: 51–58.
2. Van Oostenbrugge TJ, Fütterer JJ and Mulders PFA. Diagnostic imaging for solid renal tumors: a pictorial review. *Kidney Cancer* 2018; 2: 79–93.

3. Razik A, Das CJ and Sharma S. Angiomyolipoma of the kidneys: current perspectives and challenges in diagnostic imaging and image-guided therapy. *Curr Probl Diagn Radiol* 2019; 48: 251–261.
4. Busch FM, Bark CJ and Clydine HR. Benign renal angiomyolipoma with regional lymph node involvement. *J Urol* 1976; 116: 715–717.
5. Chawla K, Silber L and Alexander LL. Renal angiomyolipoma with retroperitoneal adenopathy. *J Natl Med Assoc* 1983; 75: 431–434.
6. Manabe T, Tasaka Y, Amano M et al. Regional lymph node involvement in benign renal angiomyolipoma. *Acta Pathol Jpn* 1984; 34: 889–893.
7. Sant GR, Ucci AA Jr and Meares EM Jr. Multicentric angiomyolipoma: renal and lymph node involvement. *Urology* 1986; 28: 111–113.
8. Brecher ME, Gill WB and Straus FH 2nd. Angiomyolipoma with regional lymph node involvement and long-term follow-up study. *Hum Pathol* 1986; 17: 962–963.
9. 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* 1987; 37: 1853–1858.
10. 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* 1989; 141: 930–932.
11. Ansari SJ, Stephenson RA and Mackay B. Angiomyolipoma of the kidney with lymph node involvement. *Ultrastruct Pathol* 1991; 15: 531–538.
12. Ackerman TE, Levi CS, Lindsay DJ, et al. Angiomyolipoma with lymph node involvement. *Can Assoc Radiol J* 1994; 45: 52–55.
13. Maffezzini M, Vlassopoulos G, Simonato A, et al. Renal angiomyolipoma with extrarenal involvement–in vascular, lymph-node and perirenal tissue. Reports of four cases. *Scand J Urol Nephrol* 1995; 29: 327–329.
14. Csanaky G, Szereday Z, Magyarlaki T, et al. Renal angiomyolipoma: report of three cases with regional lymph node involvement and/or with renal cell carcinoma. *Tumori* 1995; 81: 469–474.
15. Türker Köksal I, Tunç M, Kiliçaslan I, et al. Lymph nodal involvement by renal angiomyolipoma. *Int J Urol* 2000; 7: 386–389.
16. Göğüş C, Safak M, Erekul S, et al. Angiomyolipoma of the kidney with lymph node involvement in a 17-year old female mimicking renal cell carcinoma: a case report. *Int Urol Nephrol* 2001; 33: 617–618.
17. Lin WY, Chuang CK, Ng KF, et al. Renal angiomyolipoma with lymph node involvement: a case report and literature review. *Chang Gung Med J* 2003; 26: 607–610.
18. Liu G, Yao D, Zhang S, et al. Minimal fat renal angiomyolipoma with lymph node involvement: a case report and literature review. *Can Urol Assoc J* 2015; 9: E568–E571.
19. Kon-Nanjo K, Baba Y, Yoshio Y, et al. Renal angiomyolipoma with para-aortic lymph node involvement. *Pathol Int* 2017; 67: 273–275.
20. Tan P, Xu H, Jiang Y, et al. Microsatellite analysis for differentiating the origin of renal angiomyolipoma and involved regional lymph node. *Sci Rep* 2017; 7: 362.
21. Silverman SG, Gan YU, Mortele KJ, et al. Renal masses in the adult patient: the role of percutaneous biopsy. *Radiology* 2006; 1: 6–22.
22. Volpe A, Kachura JR, Geddie WR, et al. Techniques, safety and accuracy of sampling of renal tumors by fine needle aspiration and core biopsy. *J Urol* 2007; 2: 379–386.
23. Cui L, Hu XY, Gong SC, et al. A massive renal epithelioid angiomyolipoma with multiple metastatic lymph nodes. *Clin Imaging* 2011; 35: 320–323.