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# Retroperitoneal extrarenal angiomyolipoma: how to differentiate it from retroperitoneal liposarcoma with a case report

*Anya Jafari*[1], *Morteza sanei Taheri*[2], *Farzad Taghizadeh-Hesary*[1,3]

[1] Department of Radiation Oncology, Shahid Beheshti University of Medical Scienced, Tehran, Iran; [2] Department of Radiology, Shahid Beheshti University of medical science, Tehran, Iran; [3] Student Research Committee, School of Medicine, Shahid Beheshti University of Medical Science, Tehran, Iran

**Summary.** Extrarenal retroperitoneal angiomyolipomas (ERAMLs) are extremely rare benign lesions that can imitate some other benign and malignant retroperitoneal masses. In order to prevent the imposing an unnecessary debilitating treatment for these patients, recognizing its clinical characteristics (including imaging features) matters a lot. We present a very rare case with ERAML to shed light, especially on its radiological attributes. We proposed a pair of radiologic findings that may have the potential to differentiate ERAML from its malignant differential diagnosis (i.e. retroperitoneal liposarcoma) that are intratumoral aneurysmal vessels and intratumoral hemorrhage. We presented this especial case report in hope to guide further studies to make a valuable clinical blueprint to differentiate ERAML from retroperitoneal liposarcoma.

**Key words:** angiomyolipoma, liposarcoma, hemorrhage, aneurysm

## Introduction

Angiomyolipomas (AMLs) are benign tumors that are characterized by the presence of three distinct histological components, with varying amounts (i.e. blood vessels, fat cells and smooth muscle cells) (1). They are most commonly found in the kidneys. While extrarenal sites are also reported to harbor AML. One of these uncommon sites is retroperitoneal (RP) space. In light of its extreme rarity, the related features (including epidemiology, pathophysiology, imaging findings, etc.) are still not well defined. In this especial article, by introducing the first Iranian patient with extrarenal retroperitoneal angiomyolipoma (ERAML), we intend to shed light on its specific radiologic features to direct further studies to differentiate it from its malignant counterparts to preclude strenuous surgeries in asymptomatic patients.

## Case presentation

A 29-year-old woman, without significant past medical history, was referred to the Radiation-oncology department of Shohada-e Tajrish General Hospital (Tehran, Iran) with chronic, intermittent, and dull right-sided flank pain. The abdomen was soft on examination with moderate pain during deep palpation of the right lower quadrant (RLQ) of the abdomen. With the initial impression of appendicitis, abdominopelvic ultrasonography was performed with the following report:

*"There is no finding in favor of appendicitis, however a 9 cm ×6 cm × 5 cm heteroechoic mass is present at RLQ with extension to the liver".*

The contrast-enhanced computed tomography confirmed a retroperitoneal mass, with mixed density appearance and areas of fat attenuation and also

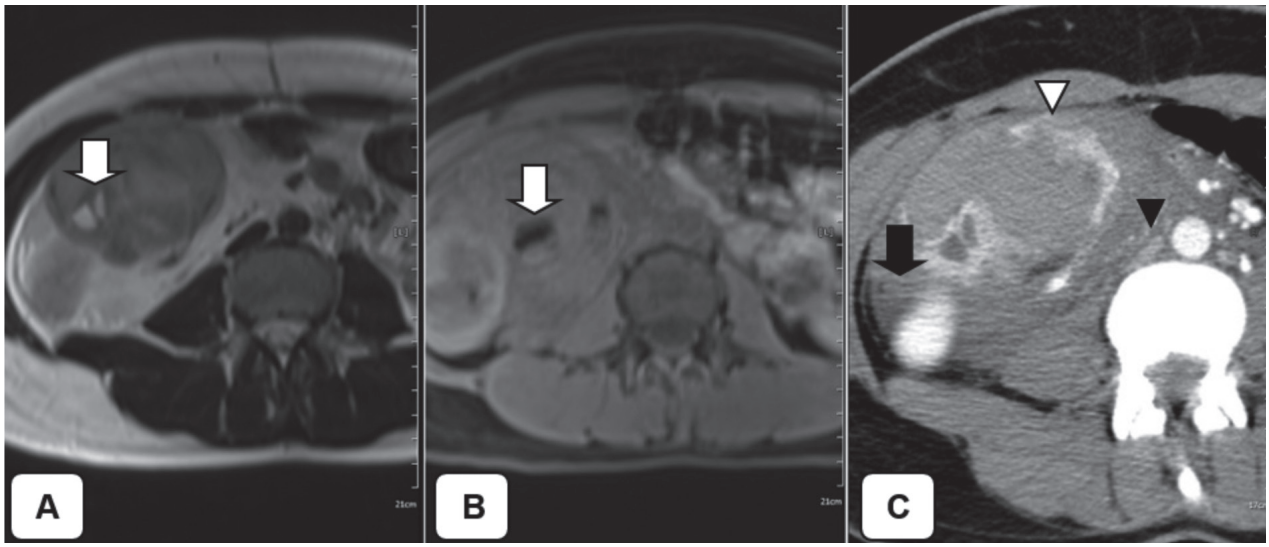


**Figure 1.** Transverse sections of enhanced CT scan and MRI show a well-defined, heterogeneous extra-renal retroperitoneal mass that is located at medio-lateral site of right kidney and contains a central prominent aneurysm. T1-weighted MRI (a) and T2-weighted MRI (b) show vascular aneurysm with apparent signal void feature at T2-weighted MRI (arrow). Contrast-enhanced CT scan (c) shows mixed density mass with fat attenuation and hyperdense area in accordance with hemorrhagic sites (white arrowhead) that has displaced the right kidney and IVC (black arrow and black arrowhead, respectively)

some areas of hemorrhage that has displaced the right kidney (figure 1). These findings also confirmed with MRI HASTE (Half-Fourier-Acquired-Single-shot Turbo Spin Echo) and CT chemical shift sequences (figures 2, 3).

The differential diagnosis were as follow: Retroperitoneal liposarcoma (RL) and ERAML.

Afterward, considering the impression of resectable liposarcoma, the patient underwent complete surgical resection. However, the pathology result was in favor of another suggested diagnosis (i.e. angiomyolipoma). It is noteworthy that the resection specimen contained a fragment of renal tissue that did not have any connection with AML mass. Consequently,

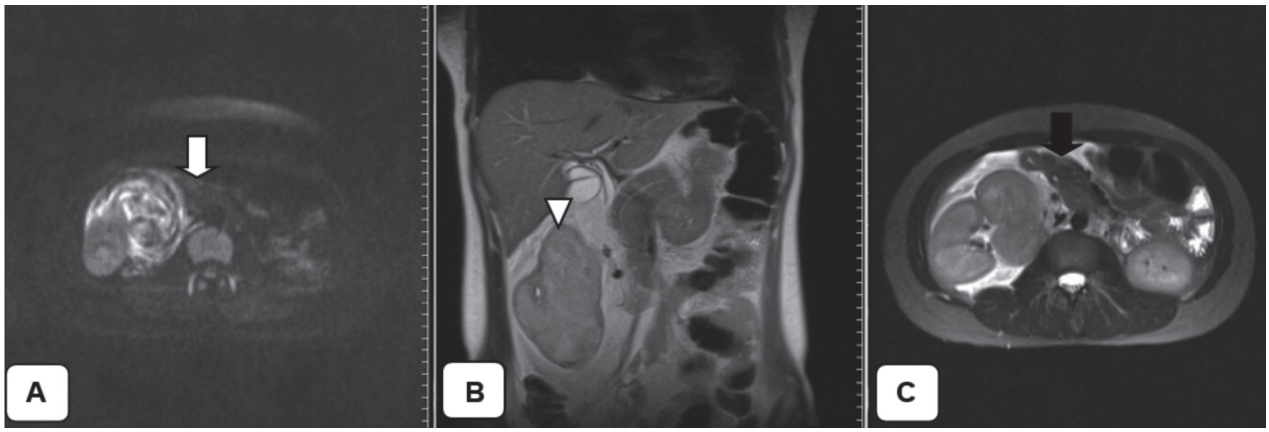


**Figure 2.** Three MRI sequences of the ERAML. a) Axial section of the diffusion-weighted MRI shows diffusion restriction that is in favor of hemorrhage. b) Coronal view of T1-weighted MRI that depicts well-demarcated heterogeneous mass that extends from subhepatic site to pelvic inlet. c) Half-Fourier- Acquired Single-shot Turbo Spin Echo (HASTE) sequence of the ERAML shows a well-defined mass with a hemorrhagic site.

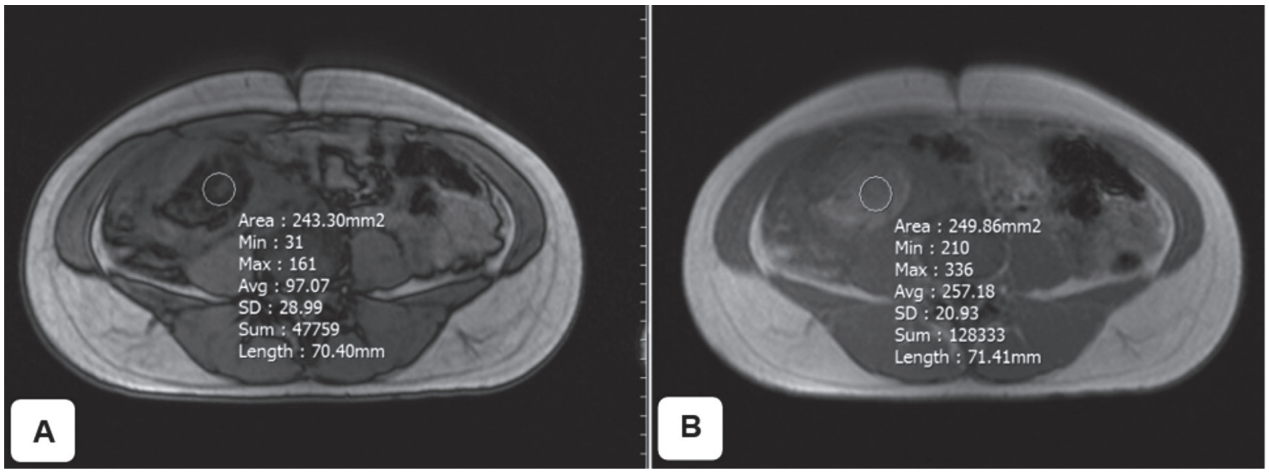


**Figure 3.** Chemical shift sequence. The figures show significant drop on out-of-phase image of mass. The signal intensity ratio is about 0.38 compatible with fat content of the ERAML. a) out-of-phase, b) in-phase.

the ERAML was proposed as the final diagnosis. The mass located adjacent to the right-sided kidney measured 10 cm × 6 cm × 6 cm that excised with negative surgical margins. Pathology review was done and according to positive immunohistochemistry (IHC) for HMB-45, the initial diagnosis was confirmed.

Considering the benign nature of AML and achievement of acceptable negative surgical margins, follow-up was planned for her status (with history taking, physical examination and the abdominopelvic US every six months for initial two years and after that annually until ten years).

## Discussion

AMLs are benign lesions that are composed of varying amounts of three distinct mature tissues (i.e. adipose tissue, dysmorphic blood vessels with spindle and epithelioid cells). They commonly arise within the kidneys. Renal AMLs have two distinct variants: classic and epithelioid (1). The epithelioid variant is categorized from the classic type by the presence of epithelioid cells (2). ERAMLs, also known as retroperitoneal angiomyolipomas (RAMLs), are uncommon retroperitoneal lesions that can imitate other retroperitoneal masses. Considering the liver as the most common extrarenal site of AMLs, RP space constitutes the second place in this perspective (3). Its incidence rate is extremely low, that based on a recent review article, only 30 cases were reported until January 2016. According to this study, it occurs predominantly in females, with a median age of 39 years. Common presentations of ERAMLs are pain (esp. on abdomen, groin, back), weight gain/loss, fullness of epigastrium, constipation, and hematuria. However, sometimes it appears incidentally, following imaging. There are some lesions that can mimic ERAML (e.g. liposarcoma, leiomyosarcoma, rhabdomyosarcoma, renal cell carcinoma). Among them, liposarcoma is more challenging to differentiate, in terms of location and histopathology (4). The distinction between ERAML and retroperitoneal liposarcoma (RL) is an important issue. Since ERAML is a benign lesion, debilitating surgical resection and radiotherapy often are not essential. On the other hand, regardless of radiologic findings, the diagnosis of ERAML may be mistaken. For example, according to the Ellingson et al study, based solely on biopsy, one out of nine AML cases was reported as liposarcoma (5).

Imaging modalities can play an essential role in differentiating ERAMLs from RLs. According to the literature review, the radiologic findings of RAMLs are characterized from RLs (Table 1) (6). Some other possible AMLs peculiarities, that can help to differentiate it from RLs, are suggested in the literature. For

**Table 1.** Summary of MDCT features of differentiating renal AML from RL

| MDCT features | Renal AML (n=31) (%) | RL (n=11) (%) | P value |
|---|---|---|---|
| Renal parenchymal defect | 31 (100) | 1 (9) | 0.000 |
| Renal artery vascular supply | 31 (100) | 0 (0) | 0.000 |
| Tumoral vessels extending through renal parenchyma | 31 (100) | 0 (0) | 0.000 |
| Dilated intratumoral vessels | 29 (94) | 1 (9) | 0.000 |
| Hemorrhage | 11 (35) | 0 (0) | 0.041 |
| Non-fat-attenuating enhancing intratumoral nodules | 0 (0) | 9 (82) | 0.000 |
| Intratumoral calcification | 0 (0) | 4 (36) | 0.003 |
| Renal sinus enlargement | 18 (58) | 1 (9) | 0.006 |
| Anterior displacement of the kidney | 10 (32) | 9 (82) | 0.011 |
| Associated AML | 12 (39) | 0 (0) | 0.018 |

AML: angiomyolipoma, RL: retroperitoneal liposarcoma, MDCT: multi-detector CT
*Courtesy of Wang et al. "Differentiating renal AML from retroperitoneal liposarcoma", September 2015.*

instance, the presence of the region of interest (ROI) with attenuation (in CT scan) less than -10 HU can depict the presence of fat cells in AML (7). Nevertheless, RL also contains fat cells. Therefore, this method seems not to be so confident.

Considering extreme rarity of ERAML, a valuable study has not yet been done regarding its radiological characteristics. Therefore, introducing potentially available key imaging features of ERAML can resolve the existing ambiguity between ERAMLs and RLs. Since it appears that ERAML originates from the extrarenal tissue, some of its radiological features possibly are different from its renal counterpart. For example, we expect "Renal artery vascular supply" not to exist in ERAML. While it is conceivably logical to extrapolate some intrinsic features of AML to ERAML (e.g. hypervascularity with the presence of aneurysmal vessels inside the tumor mass). Another finding that may be in favor of ERAML is intra/extra tumoral hemorrhage (due to the presence of intratumoral dysplastic blood vessels). However, in generalizing this issue, it has to be cautious; because in a case report intratumoral hemorrhage was reported in a patient with the diagnosis of RP liposarcoma (8). Noteworthy, among all of these characteristics there were only two features that were present in our patient (based on retrospective assessment). They are including intratumoral aneurysmal vessels and intratumoral hemorrhage (Figure 1, 2).

These findings have the potential to be a valuable clue for further studies to define the comprehensive radiological characteristics of ERAML, in hope that it would be helpful in clinically distinguish it from its malignant differential diagnosis (i.e. RL) to spare the affected patient from a radical surgery.

**Contributors:** Farzad Taghizadeh Hesary performed main parts of literature review and manuscript draft. Anya Jafari provided the data and edited the manuscript. Morteza Sanei Taheri provided the imaging data and edited the manuscript. All authors read and approved the final manuscript.

## References

1. Lane BR, Aydin H, Danforth TL, Zhou M, Remer EM, Novick AC, Campbell SC. Clinical correlates of renal angiomyolipoma subtypes in 209 patients: classic, fat poor, tuberous sclerosis associated and epithelioid. J Urol 2008; 30; 180(3): 836-43.
2. Nese N, Martignoni G, Fletcher CD, Gupta R, Pan CC, Kim H, Ro JY, Hwang IS, Sato K, Bonetti F, Pea M. Pure epithelioid PEComas (so-called epithelioid angiomyolipoma) of the kidney: a clinicopathologic study of 41 cases: detailed assessment of morphology and risk stratification. Am J Surg Pathol 2011; 1; 35(2): 161-76.
3. Minja EJ, Pellerin M, Saviano N, Chamberlain RS. Retroperitoneal extrarenal angiomyolipomas: an evidencebased approach to a rare clinical entity. Case Reports in Nephrology 2012.

4. Venyo AK. A review of the literature on extrarenal retroperitoneal angiomyolipoma. Int J Surg Oncol 2016; 17; 2016.
5. Ellingson JJ, Coakley FV, Joe BN, Qayyum A, Westphalen AC, Yeh BM. Computed tomographic distinction of perirenal liposarcoma from exophytic angiomyolipoma: a feature analysis study. J Comput Assist Tomogr 2008; 32(4): 548.
6. Wang Q, Juan YH, Li Y, Xie JJ, Liu H, Huang H, Liu Z, Zheng J, Saboo US, Saboo SS, Liang C. Multidetector computed tomography features in differentiating exophytic renal angiomyolipoma from retroperitoneal liposarcoma: A Strobe-Compliant Observational Study. Medicine (Baltimore) 2015; 94(37).
7. Angulo JC, López JI, Carnicero JA, Flores N. Extrarenal retroperitoneal angiomyolipoma. Urol Int 1994; 52(1): 58-60.
8. Wong AM, Chen YL, Chen SC, Tan CF, Tsai ML, Ng KK, Cheung YC. Retroperitoneal liposarcoma complicated with bleeding: CT Features. Chinese journal of radiology. Taipei 2002; 1; 27(6): 315-8.

Correspondence:
Farzad Taghizadeh-Hesary
Department of Radiation Oncology,
Shahid Beheshti University of Medical Sciences, Tehran, Iran
Tel. +989126086713, +9826651517
Fax +99826651517
E-mail: Farzadth89@gmail.com; f_taghizadeh@sbmu.ac.ir