Case Report

Retroperitoneal extrarenal angiomyolipoma at the surgical bed 8 years after a renal angiomyolipoma nephrectomy: A case report and review of literature

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Abstract

Retroperitoneal extrarenal angiomyolipoma (RERAML) are rare and close mimickers of retroperitoneal liposarcoma on both imaging and histopathology. However, imaging findings including heterogeneity, hyperdensity on unenhanced computed tomography, intralesional hemorrhage, absence of calcifications, low signal intensity on T2-weighted magnetic resonance imaging, and dilated intratumoral vessels can lead to the diagnosis of RERAML. Diagnosis of RERAML can avoid unnecessary surgery since conservative medical management with continued surveillance has been proven to be effective for RERAML whereas surgical resection is the treatment for liposarcoma. Imaging and laboratory follow-up for at least 5 years has been recommended in patients who underwent surgical resection of angiomyolipoma (AML). We present a case of RERAML in an asymptomatic patient whose AML recurred in the surgical bed 8 years after an ipsilateral nephrectomy for renal AML.

Keywords: Angiomyolipoma, recurrence, retroperitoneal extrarenal angiomyolipoma

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INTRODUCTION

Extrarenal retroperitoneal space is the third most common primary site for angiomyolipoma (AML). Sixty cases of extrarenal AML (ERAML) have been reported since the first case report in 1982 by Friis and Hjortrup,[1,2] of which only 16 were retroperitoneal extrarenal angiomyolipoma (RERAML). RERAML is difficult to differentiate from the liposarcoma however the history of AML and few characteristic imaging features indicate RERAML which can be managed with mechanistic target of rapamycin (mTOR) kinase inhibitor therapy and surveillance rather than surgery. We report a patient with AML recurrence in the surgical bed 8 years after nephrectomy for a large renal AML. We found no reports in literature with a similar postoperative presentation.

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CASE REPORT

A 35-year-old asymptomatic female had a history of large left renal AML which presented as acute left-sided abdominal pain and massive retroperitoneal hemorrhage on computed tomography (CT) abdomen imaging at another hospital (images unavailable). She had left nephrectomy for the same 8 years ago, details of which were unavailable when

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she presented to our institution. She was then followed up at the same hospital annually for 2 years with CT abdomen showing thin-walled cysts in basal segments of both lungs [Figure 1], suggestive of lymphangioleiomyomatosis (LAM). She was followed up by a pulmonologist for LAM with regular pulmonary function tests, and CT chest 6 years later showed mild interval increase in the number of cysts in both lungs [Figure 2], however without any pulmonary symptoms or complications since her diagnosis. She was worked up for tuberous sclerosis including genetic test tuberous sclerosis complex [TSC] 1 and 2 and magnetic resonance (MR) brain which were negative. She had no significant past medical, social, or family history. Abdomen CT during the first 2 years postnephrectomy showed no residual or recurrent disease in the left renal bed after which she had no further imaging follow-up. She had annual vascular endothelial growth factor-D (VEGF-D) levels which were within normal limits since nephrectomy.

Now, she presented to our hospital 8 years after nephrectomy when her admission VEGF-D level was elevated - 1197 pg/ml (normal <600 pg/ml). CT abdomen showed a new 9.1 cm × 3.5 cm enhancing fat-containing, mixed-density lesion at the left renal bed [Figure 3a and b]. The retroaortic left renal vein extended into the lesion and arborized, with left renal artery seen as short, blind-ending stump. The contralateral right kidney was normal. Imaging diagnosis favored retroperitoneal ERAML, and she was placed on a trial of everolimus, an mTOR4 kinase inhibitor, since she was asymptomatic. Follow-up VEGF-D level at 4 months decreased to 623 pg/ml, and ultrasound (US) showed mild decrease in the size of the retroperitoneal lesion [Figure 4], and hence embolization/surgery was deferred.

On subsequent 6-month follow-ups for the next 2 years, everolimus levels remained within the therapeutic range, there were no changes on US, and the patient continued to be symptom free.

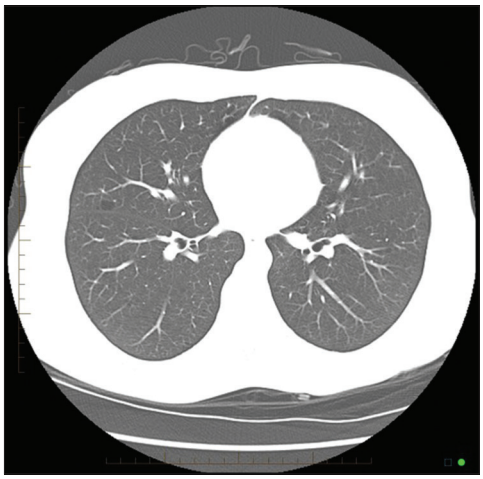

Figure 1: Axial computed tomography chest lung window in 2006 showed few thin-walled cysts in both lungs suggestive of lymphangioleiomyomatosis

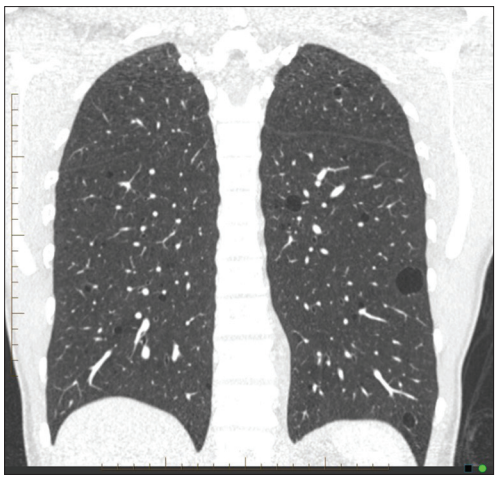

Figure 2: Computed tomography chest lung window coronal reformatted image in 2014 showed interval increase in the number of bilateral thin-walled cysts; the patient was asymptomatic

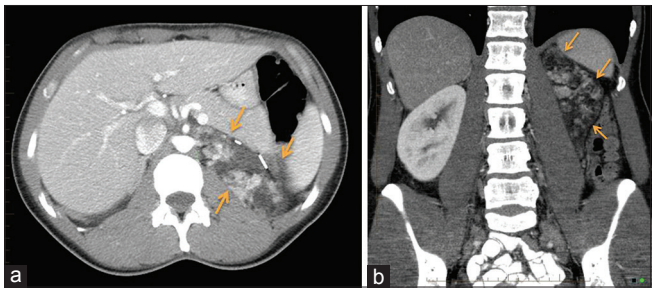

Figure 3: Computed tomography abdomen in 2014 in axial section (a) and coronal reformation (b) showed new heterogeneous, predominantly fat-containing, enhancing lesion measuring 9.1 cm × 3.5 cm at the left renal surgical bed (arrows), with features suggesting angiomyolipoma

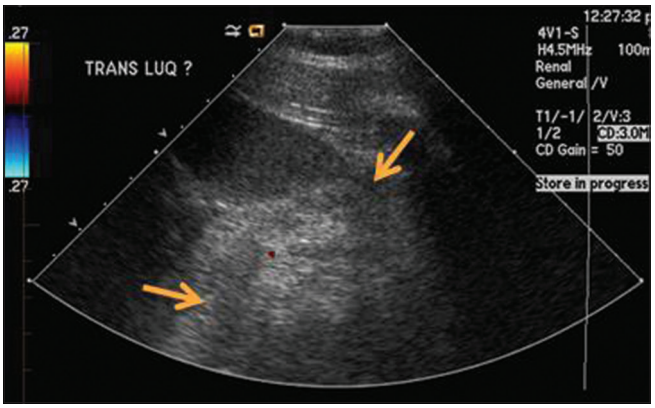

Figure 4: Follow-up renal ultrasound performed 1 year after starting the patient on everolimus showed mild decrease in the size of the left renal bed mass

DISCUSSION

AML is a benign neoplasm of monoclonal origin, sometimes referred as renal hamartoma, choristoma, or perivascular epithelioid cell tumors. It has triphasic morphology on histology that includes mature adipose tissue, thick-walled blood vessels, and perivascular spindle cells.[3-6] They show immunoreactivity to smooth muscle actin, HMB-45, and melanocytic marker Melan-A.[7] AMLs can occur in inherited forms in association with tuberous sclerosis (10%–20%); however, the sporadic form is the most common (80%–90%).[1,7-11]

ERAML are rare and usually present as incidentalomas on imaging.[3,12] These rare tumors were reported in liver, retroperitoneum, adrenal glands, colon, urinary bladder, hilar lymph nodes, lungs, ribs, oral and nasal cavity, abdominal wall, fallopian tube, uterus, and skin.[1,3,5,9,12-14] Literature review reports only 60 cases of ERAML since its first description in 1982 by Friis and Hjortrup in the above-mentioned locations, of which only 16 cases have been reported in the retroperitoneum, which is the 2nd most common ERAML primary location.[2,3,13] RERAML are usually >10 cm and asymptomatic in view of their retroperitoneal location.[1,12,13,15] However, they can less commonly present with nonspecific symptoms such as vague abdominal pain, weight gain, fullness in epigastric region/abdomen, hematuria, and constipation, and rarely with enlarged abdomen or ureteric obstruction or retroperitoneal hemorrhage.[4,9,13]

Imaging has a crucial part in the diagnosis of RERAML since they are mostly asymptomatic and difficult especially in obese patients and helps in determining the extent of the tumor as well as guides surgical planning.[13] It is very helpful in the surveillance of the postnephrectomy AML patients. US determines the size and extent of the mass, mass effect on adjacent organs, presence of metastatic lesion, and guides biopsy.[4] The classical sonographic findings are well-defined hyperechogenic mass with acoustic shadowing.[1] Cross-sectional imaging assessment with CT, CT angiography, and/or MR imaging (MRI) includes size, internal characterization of the mass, margins, extent, involvement of regional vessels, and also guides biopsies.[3,4] Thin-section non-enhanced CT (NECT) is preferred for identifying the intralesional fat content.[6] On contrast-enhanced CT, these present as noncalcified macroscopic fat-containing hyperdense mass which can help in differentiating them from renal cell carcinoma (RCC).[1,6-8,10] The challenge to CT imaging is radiation concerns since the commonly affected population with this condition are young females. MRI features include heterogeneous signal intensity on T1-weighted images due to fat content and foci of hemorrhage, low signal intensity on T2-weighted images due to its smooth muscle component, signal dropout on gradient-echo or spin-echo images with fat suppression, India Ink artifact at fat-water interface on chemical shift imaging, profound diffusion restriction on diffusion-weighted imaging, and/or rapid arterial enhancement with contrast administration.[6,8,10] MRI uses the fat suppression techniques such as inversion recovery and chemical saturation to identify the intratumoral fat component and differentiate it from intratumoral hemorrhage.[8] Although not pathognomonic, RERAML demonstrates aneurysmal dilatation of intratumoral vessels, linear vascularity, bridging veins, and/or hematomas on angiography.[3,4]

Although most of these RERAML have been benign, it is difficult to exclude malignancy which is the closest differential diagnosis due to similarities in imaging appearance, and the common differentials include lipoma, liposarcoma, papillary RCC, and adrenal myelolipoma.[2,3,8,10,12,13,14] No serum biochemistry or urinalysis investigation is specific for RERAML.[4] The most common differential, liposarcoma, is difficult to differentiate from RERAML even on positron emission tomography/CT and histopathological examination.[15] CT and MRI help in the diagnostic dilemma by the following: liposarcomas arise from outside the Gerota's fascia whereas RERAML arise from perinephric fat; features favoring RERAML include history of AML, microscopic fat, heterogeneity on imaging, hemorrhage, absence of calcifications, NECT hyperdensity, T2 low signal intensity, dilated intratumoral vessels.[6,9,16] Although not mandatory for making a diagnosis, HMB-45 positivity of RERAML and positive FISH test for MDM2 amplification in liposarcomas can help in differentiation.[3,5,12,13] RCC can be differentiated from RERAML by the presence of calcifications, enhancing intratumoral nodules, and invasion into renal vein or inferior vena cava being more common in the former than the latter.[7,8,16]

Most common and dreaded complication of RERAML is retroperitoneal hemorrhage.[9,12,13] Rarely malignant degeneration and metastasis occur with recurrence considered very rare in ERAML with only two cases in literature describing distant metastasis to mediastinum, liver, and bone.[3,13] Usually, AML patients do not have recurrence after renal sparing nephrectomy or embolization even at 5-year follow-up period as described in literature.[3] However, our patient had RERAML presenting as AML recurrence at the surgical bed following total nephrectomy for renal AML. Close follow-up with CT imaging during the 1st year following surgery with continued follow-up to

5 years has been recommended.[3,4] It is recommended that management of AML should be dependent on the size of lesion, symptomatology, and estimated compliance with follow-up, though historically size >4 cm was considered as universal standard cutoff for invasive treatment given its risk for rupture and life-threatening retroperitoneal hemorrhage.[3,7,10,11,14]

Treatment options include minimally invasive techniques such as radiofrequency ablation, cryoablation, microwave ablation, selective angioembolization (SAE), and surgery; however, the latter two have proven to be the most effective for symptomatic AML.[4,7,9-12] SAE is usually performed and reserved for patients with active bleeding, large retroperitoneal hemorrhage, plan for subsequent staged surgical resection, or large tumors with the advantage of short recovery period and preserved renal function.[3,4,7] Recently, hormonal or targeted therapies including sirolimus have been used for downgrading tumors by decreasing the size of the tumor and as an adjuvant treatment to patients undergoing SAE; however, controversies exist due to the increase in the size of tumor once the treatment is discontinued.[7,11,13] RERAML management is similar to AML, and current recommendations by the International TSC Consensus group for the management of AML are embolization and corticosteroids as first-line therapy in acute hemorrhage. However, for asymptomatic patients with growing AML >3 cm, growing and/or seen in association with tuberous sclerosis, mTOR inhibitor is recommended as first-line therapy followed by SAE when unresponsive to mTOR therapy.[4] When metastatic, aggressive surgery with resection and vascular reconstructions can improve outcome.[13] However, RERAML can be biopsied and/or safely followed up with imaging if the imaging findings are highly suggestive of RERAML given the benign nature. Our patient had asymptomatic postnephrectomy surgical site mass with imaging features highly suggestive of RERAML. Therefore, surgery was deferred and she was started mTOR inhibitor trial with good response to therapy.

CONCLUSION

RERAML is a rare entity specially to present as recurrence following nephrectomy, however, should be considered in the differential when it occurs. It is difficult to differentiate RERAML from retroperitoneal liposarcoma by imaging and histology. Imaging features such as heterogeneity, hemorrhage, absence of calcifications, hyperdensity on NECT, low signal intensity on T2-weighted MR images, and dilated intratumoral vessels can help in making a confident diagnosis of RERAML on imaging. Imaging helps to decide medical management over invasive surgery in asymptomatic patients, with surgical resection reserved for larger lesions with impending risk of bleeding or suspicious features for malignancy. If surgically resected, it is important to follow up these patients with imaging and laboratory tests for at least 5 years from surgical resection to ensure stability and no recurrence.

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Conflicts of interest

There are no conflicts of interest.

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