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Case report

Diagnostic value of multi-slice CT scan for renal angiomyolipoma detection: a case report

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Abstract

Background Angiomyolipoma (AML) is one of benign tumor that commonly affects kidney. They are seen 80-90% as isolated lesions that occur sporadically and seen 25-50% of patients with tuberous sclerosis. Computed tomography may help to diagnose AML, followed by further histopathological examination for definitive diagnosis. Early diagnosis is important to prevent complication such as rupture.
Objective This report describes the role of imaging in detecting renal AML at Bethesda Hospital, Yogyakarta
Case description A 49-year-old male presented with left flank pain persisting for 1 year. Aside from pain, there was no other urological symptoms. Multislice abdominal CT scan with contrast revealed a large hypodense nonhomogeneous mass in the upper pole of the left kidney. The patient underwent total left nephrectomy and the postoperative histopathological examination confirmed as AML.
Conclusion Abdominal CT scan is important to determine the malignancy of the kidney lesion. The presence of macroscopic fatty lesion would distinct AML from other causes.

Keywords: angiomyolipoma, kidney, multidetector computed tomography, flank pain

Introduction

Angiomyolipoma (AML) is a solid benign tumor, composed of blood vessels, smooth muscle cells, and fat. It can occur everywhere, but AML is more commonly found in kidney. In fact, renal AML is the most common benign tumor of the kidney, even though AML itself is not commonly found. Renal AML arises from either the renal pelvis or the sinus, hence it also referred as renal hamartoma.[1,2] AML can occur sporadically as isolated lesions (80-90%) or in association with autosomal dominant disorder tuberous sclerosis (TSC) as a part of phakomatosis (25-50%).[1,3] AMLs are typically identified in adults (mean age of presentation 43 years), with a female predilection (F:M of 2-4:1).[3] Sporadic AML usually occurs unilaterally while TSC usually bilateral.4 When AMLs grow to a size of >10 cm, they are referred to as 'giant' AMLs.[2] Almost all classic AMLs are benign but they do have the risk of rupture with bleeding or secondary destruction of surrounding structures as they grow. Two histological types of AMLs have been described as typical (triphasic) and atypical (monophasic or epithelioid). AML usually asymptomatic and is often found incidentally when the kidneys are imaged for other reasons, or as part of TSC screening. Symptomatic presentation is most frequently with spontaneous retroperitoneal hemorrhage.[3]

Case Description

Patient information

A 49 years old male patient presented with left flank pain for 1 year, an intermittent dull aching with no relieving or aggravating factors. He denied any history of dysuria, hematuria, weight loss, and family history of malignancy.

Clinical findings

On physical examination, there were no cutaneous or other stigmas of TSC. On palpation, a sizeable mass was identified in the left upper quadrant abdominal area, margins ill-defined, non-tender, mobile, and ballotable.

Diagnostic assessment

Laboratory results of complete blood count and renal function were within the normal range, as well as normal urine analysis. Ultrasonography (USG) of the stomach showed hyperechoic mass sized ~9 cm in the left kidney. The patient underwent a multi-slice computed tomography (MSCT) examination with contrast, showed 11 x 9,3 cm hypodense

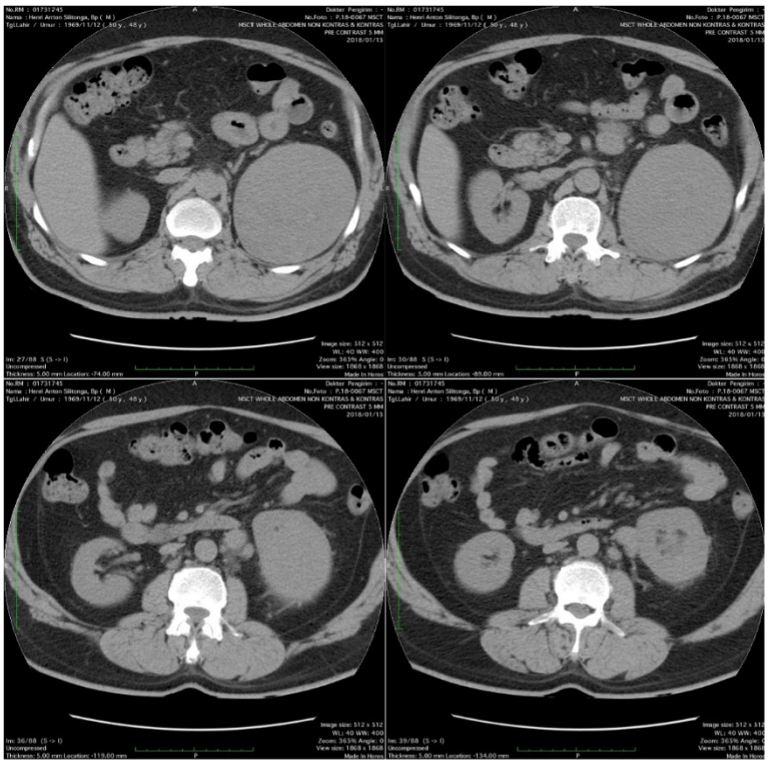

Figure 1. *MSCT images of left renal angiomyolipoma (pre-contrast)*

inhomogenous mass in the upper pole area of left kidney, in continuity with hypertrophy of left kidney with hump sign appearance, there was also enhancement of mass contour and feeding artery can be seen. The right kidney appears normal, and both kidneys had normal secretion phase. The density of liver, bile duct, pancreas, lien, and bladder were normal.

Therapeutic intervention and Follow-up

The patient was discharged for an elective surgery. He underwent a left nephrectomy without adrenalectomy because the left adrenal gland was normal. The postoperative histopathological examination confirmed the mass as an angiomyolipoma, showing adipocytes, proliferating spindle cells, and thickened vessels. Postoperative recovery was uneventful, and still in remission after 14 months..

Discussion

Renal angiomyolipoma is a rare benign neoplasm of the kidney arising from mesenchymal elements and is also referred to as renal hamartoma due to its varying composition.2 AML is considered one of several tumors with perivascular epithelioid cellular differentiation (PEComas).[3] AML is composed of angiomatous tissue, smooth muscle, and fat elements.[5] AML is not only restricted to the kidney, but also can occur in various other sites like skin, appendix, colon, liver, lung, and smooth muscle fibers.[1]

We reported a case of 49 years old man with renal AML. The initial symptom was long term left flank pain. Average patient age of renal AML at presentation is 50 years (range 17-74 years old). Renal AML largely favor female patients, however, there is a growing list of renal AML in men nowadays.[6] Most of renal AML cases are found incidentally on imaging but symptomatic presentation does exist. Symptomatic presentation is most frequently related to spontaneous retroperitoneal haemorrhage. Other symptoms and signs include flank pain, a palpable mass, haemturia, anaemia, urinay tract infection, or renal failure.[7] There was no any signs and symptoms which lead to retroperitoneal haemorrhage in this case.

Typical renal AML can be diagnosed accurately based on imaging findings. The cornerstone of diagnosis on all modalities is the demonstration of macroscopic fat, however in the setting of hemorrhage, or when lesions happen to contain little fat, it may be difficult to distinguish an AML from RCC.[3] Ultrasonography (USG) of AML tend to appear as hyperechoic lesions, located in the cortex and with posterior acoustic shadowing. In the setting of tuberous sclerosis, they may be so numerous that the entire kidney is affected, appearing echogenic with the loss of normal corticomedullary differentiation. Contrast-enhanced ultrasound tends to enhance peripherally, decreased central enhancement, compared with the normal cortex.[8] Most AML lesions involve the cortex and computed tomography (CT) demonstrates macroscopic fat (less than -20 HU).[9]

In this case, the USG of patient's stomach showed hyperechoic mass in the left kidney. Further multi-slice computed tomography (MSCT) examination confirm

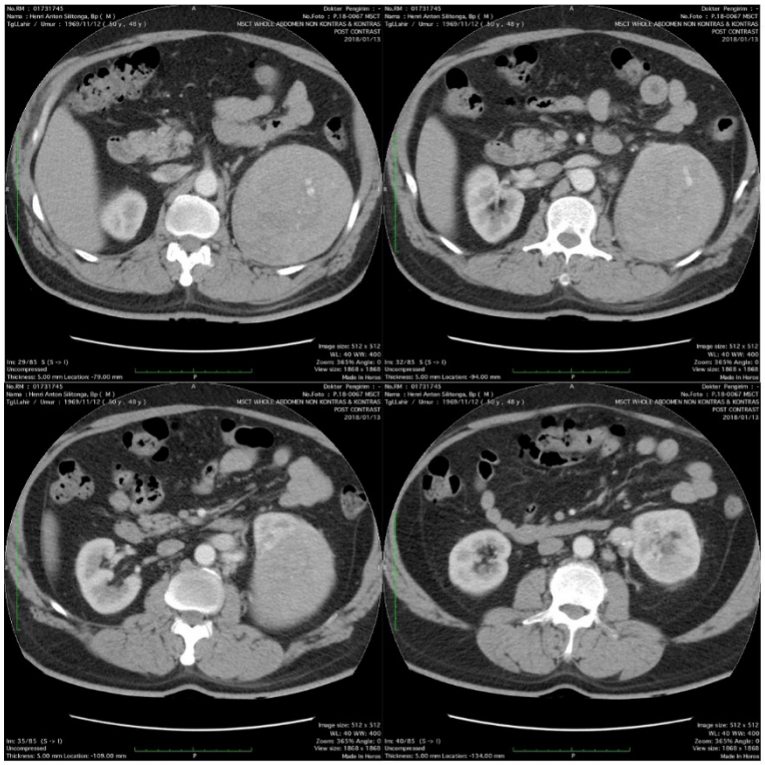

Figure 2. *MSCT images of left renal angiomyolipoma (post intravenous contrast)*

the finding by showing a hypodense inhomogenous mass in the upper pole area of left kidney, in continuity with hypertrophy of left kidney with hump sign appearance. The hyperechoic and hyperdense imaging shows combination of fat, blood vessel, and muscle contents while the shadow shows multiple tissue interfaces between different elements.[10]

Differential diagnosis of AML are subtypes of sarcoma (fibrosarcoma, leiomyosarcoma, and renal cell carcinoma). It is essential to remember that rarely renal cell carcinomas (RCC) may have macroscopic fat components and as such the presence of fat is strongly indicative of an AML, but not pathognomonic. It is important to realize that ~5% of AMLs are fat poor which do not demonstrate macroscopic fat on CT. The absence of ossification/calcification on imaging is in favor of AML.[3,9] All of the radiograph examination in this case found no signs of calcification in both renal, therefore the diagnose of renal AML is more preferable.

Renal epithelioid angiomyolipoma (EAML) is a recently recognized rare variant that originates from the PEComas and it has aggressive clinical behavior which includes local recurrence and metastasis.[1] AML usually asymptomatic which is often found incidentally when the kidneys are imaged for other reasons or TSC screening. Symptomatic presentation is most frequent with retroperitoneal hemorrhage. Shock due to severe hemorrhage from rupture is described as Wunderlich syndrome.[3] The main complication of AML is rupture hemorrhage which is related to tumor size, increased vascularity, and abnormal thickened blood vessels. Conventional AML has got good prognosis compared to rare EAML which is potentially malignant. 1

AMLs found incidentally usually require no therapy when small, although follow-up is recommended to assess for growth.[11] Larger AMLs, or those that have been symptomatic, can be electively embolized and/or resected with a partial nephrectomy or total nephrectomy.[3] Intervention is considered when an AML size reaches ≥4 cm in size.[12] The lesion that presents with retroperitoneal hemorrhage often requires emergency embolization as a life-saving measure. Another therapy for AMLs is mTOR inhibitors (e.g. everolimus) have been shown to significantly decrease AML size and may help to preserve renal function.[13,14]

Conclusion

In this report, the diagnosis of renal AML was made based on radiograph imaging, especially from USG and MSCT findings. Typical renal AML can be diagnosed accurately based in imaging findings. The demonstration of macroscopic fat within a lesion is the hallmark feature on all modalities. Therefore, it is very important to do some further radiograph imaging to establish the diagnosis, especially to differ renal AML from any renal carcinoma.

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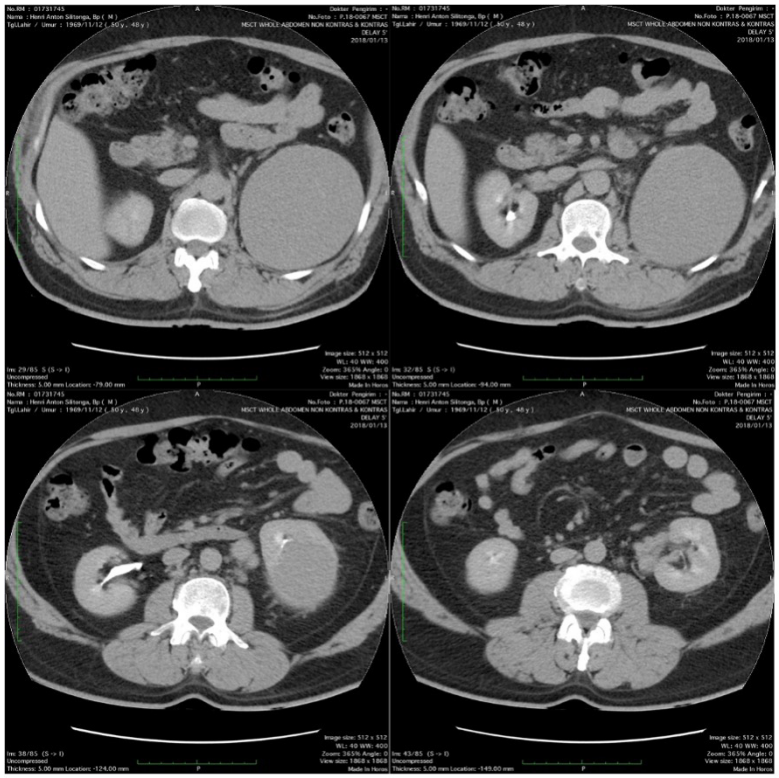

Figure 3. *MSCT images of left renal angiomyolipoma (delayed contrast 5 minutes)*

Conflict of interest

The authors have no conflict of interest.

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