

Efficacy Sirulimus and Pathology Report for Angiomyolipoma Patient: A Case Study

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Abstract: Angiomyolipoma (AML) is a benign neo-plasm composed of thick blood vessels, fatty tissue and smooth muscle cells. Herein, we reported a case of revealed diagnosis of AML from kidney and liver features and treated with sirulimus. A 25 year-old woman with a left renal mass incidentally was diagnosed by ultrasound screen for left side of constant abdominal pain. She referred to our cancer center in March 2014. In physical exam of abdomen was normal, without organomegaly. In laboratory exam, the results of urinalysis and other routine blood tests been normal. The decision of urologist at first evaluation with clinical diagnosis of Renal Cell Carcinoma (RCC) was nephrectomy. After nephrectomy in the pathological cross section one encapsulated mass with focal hemorrhage and necrosis were observed. Histopathological examination of the tumor by hematoxylin and eosin staining revealed diagnosis of AML from kidney and liver. We selected her for treatment with mTOR inhibitor of sirulimus (2mg per day). After 3 months, she is in a good condition without any new complaint. During this last three months didn't have any significant side effects of sirulimus. AML is not a malignancy disorder but is very same to renal cell tumor, and usually is diagnosed after surgery. We must explain this matter that different diagnosis of RCC is multiple and all surgery must know about this matter. This case is metastatic case we must know future details about the pathology.

Keywords: AML, RCC, Sirulimus.

INTRODUCTION

AML is a benign neo-plasm composed of thick blood vessels, fatty tissue and smooth muscle cells [1]. Although AML is occasionally reported in different anatomic locations, it is most often found in the kidneys, followed by the liver [2]. It is primarily a benign non-aggressive tumor typically requiring intervention in symptomatic patients or when the tumor diameter exceeds 4 cm as the risk of hemorrhage increases [3]. Here, we reported a case of revealed diagnosis of AML from kidney and liver features and treated with sirulimus.

CASE REPORT

A 25 year-old woman with a left renal mass incidentally was diagnosed by ultrasound screen for left side of constant abdominal pain. She referred to our cancer center in March 2014. In physical exam of abdomen was normal, without organomegaly. In

laboratory exam, the results of urinalysis and other routine blood tests been normal. Abdominopelvic spiral CT scan showed a left renal mass (>50mm) which was enhanced in the early phase and washed out in the late phase (Fig. 1, Fig. 2 and Fig. 3) and also sinks localized hypo-dense mass in the left lobe of liver that the size of tumor in liver was 61.6×48mm (Fig. 4). The decision of urologist at first evaluation with clinical diagnosis of RCC was nephrectomy. After nephrectomy in the pathological cross section one encapsulated mass with focal hemorrhage and necrosis were observed. Histopathological examination of the tumor by hematoxylin and eosin staining revealed diagnosis of AML from kidney and liver. We selected her for treatment with MTOR inhibitor of sirulimus (2mg per day). After 3 months, she is in a good condition without any new complaint. During this last three months didn't have any significant side effects of sirulimus.

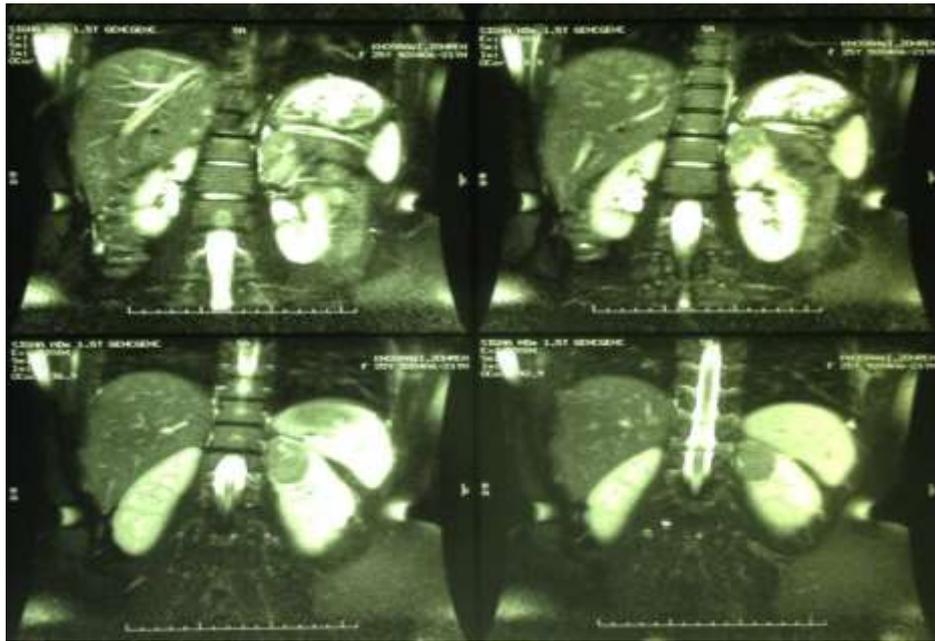


Fig. 1: Hyper signal mass intensity in upper pole of left kidney (>50mm) (before surgery)

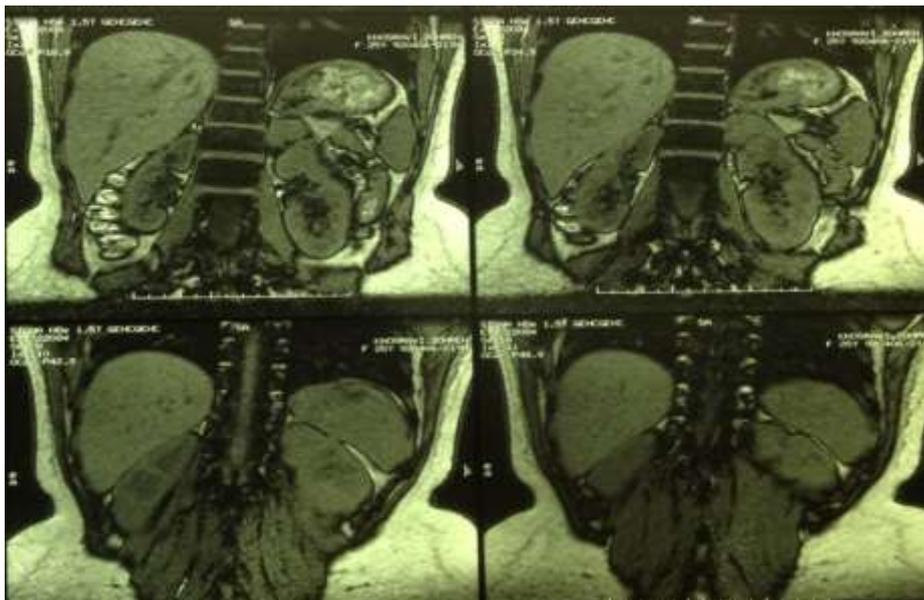


Fig. 2: Hyper signal mass lesion in the upper-pole of left kidney in T 2 MRI (before surgery)

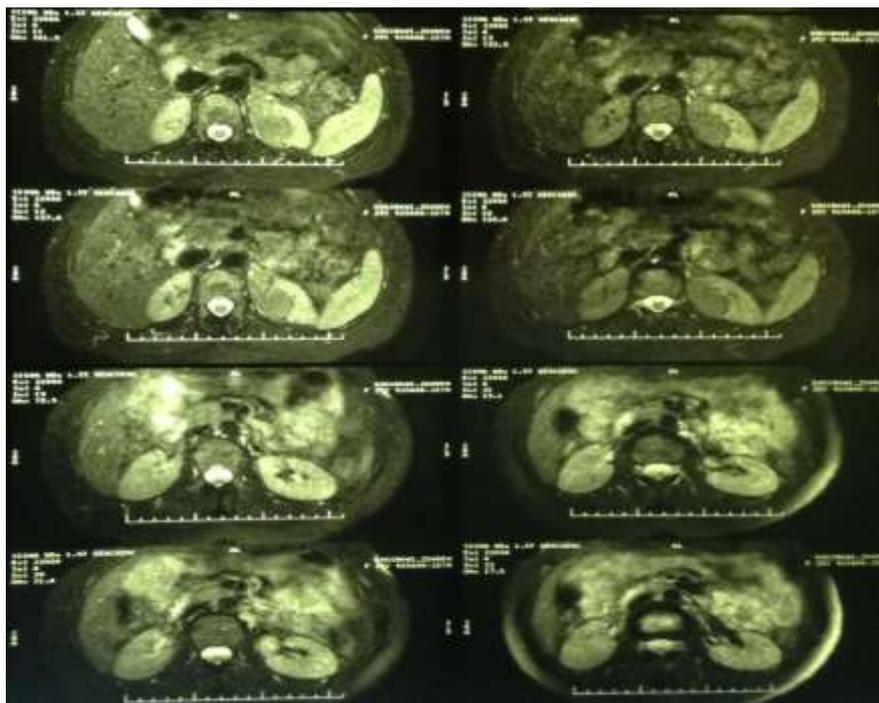


Fig. 3: Hyper signal mass intensity in upper pole of left kidney in T2 MRI (before surgery)

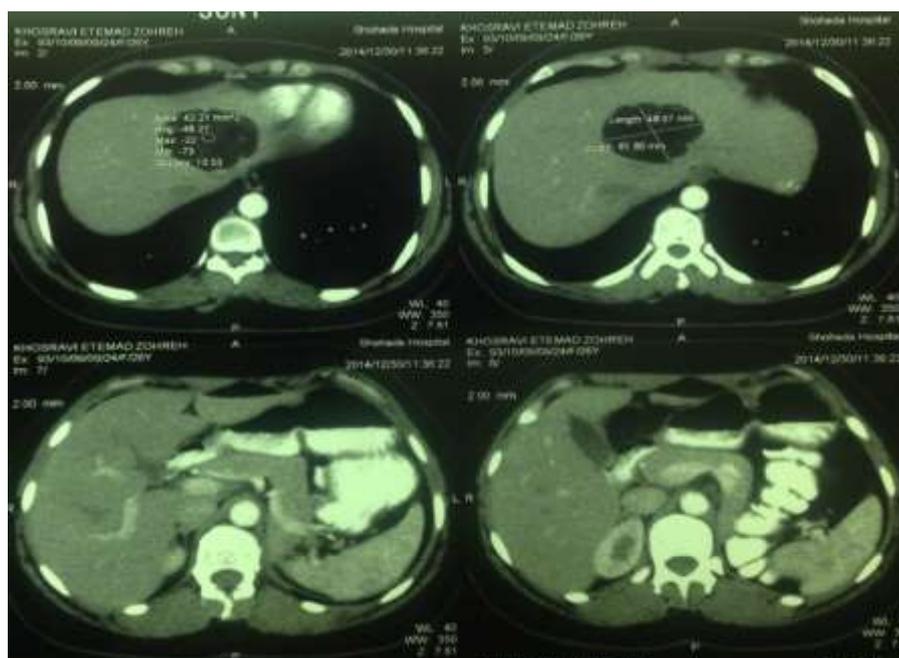


Fig. 4: Abdominal CT scan shows sinks localized hypo-dense mass (61.6×48mm) in the left lobe of liver (before surgery)

DISCUSSION

The term AML is used for a non-metastasizing renal tumor composed of fat, smooth muscle, and blood vessels. AML is usually well demarcated from kidney and may extend into the surrounding retroperitoneum. Wide excision is curative [4]. AMLs of kidneys are one of the common extracranial manifestations of tuberous sclerosis (TSC) [5]. But hepatic epithelioid AML is a rare characterized by predominant or exclusive

population of epithelioid cells coexpressing melanocytic and myogenic markers [2]. Treatment of renal AML can be done by two different options. One option is the classical surgical approach with a total or partial nephrectomy and the other is selective embolization [1]. AMLs are rich in fat and represented on CT scans by a low signal that renders them easy to diagnose [6, 7]. Approximately 5% of them lack fat and cannot be differentiated from RCCs. Thus, for the

AMLs that lack fat are difficult to diagnose by CT. It has been proposed that MRI may improve the accuracy of diagnosis with the help of fat saturation techniques [6, 8, 9]. On histopathologic findings, malignant AML may resemble sarcomatoid RCC. But they can be distinguished on the basis of immunohistochemical evaluation. AML cells are positive for the HMB-45 antibody and negative for cytokeratins, while 94% of sarcomatoid RCC cases test positive for the AE1/AE3 antibody [10, 11]. The diagnostic accuracy of opposed-phase and in-phase GRE MR imaging for the differentiation of minimal fat AML and clear cell RCC has been found to be poor. Low SI on T2-weighted images relative to renal parenchyma and small size has suggested minimal fat AML, while intratumoral necrosis and large size has argued against this diagnosis [12]. A study has shown that renal AML shrank during sirulimus therapy but has tended to re-grow after stopping the therapy. Generally, sirulimus has been reported as an effective and safe therapy for renal AML in the patients with tuberous sclerosis complex or sporadic lymphangioleiomyomatosis [13].

CONCLUSION

AML is not a malignancy disorder but is very same to renal cell tumor, and usually diagnosed after surgery. We must explain this matter that different diagnosis of RCC is multiple and all surgery must know about this matter, but because this case is metastatic case we must know future details about the pathology. It is prefer that this case had one the indication for nephrectomy.

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