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Renal Epithelioid Angiomyolipoma: Diagnostic and Therapeutic Difficulty!

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Case Report

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Abstract

Renal epithelioid angiomyolipomas (AMLeR) are rare tumors and account for 8% of angiomyolipomas (AML) operated ; We illustrate an observation of the diangotic and therapeutic difficulties of these types of tumors which remains rare, and whose management is not yet well defined both radiologically and urologically.

Keywords: Epithelioid Angiomyolipoma; Partial Nephrectomy; Malignant Forms

Introduction

Renal epithelioid angiomyolipomas (AMLeR) are rare tumors and account for 8% of angiomyolipomas (AML) operated [1]. This entity poses a diagnostic and therapeutic problem for the urologist as well as for the anatompthological, as well as for radiologists [2].

Case Report

A.R., 35 years old, had been complaining for 2 months of medium-intensity left low back pain associated with a single episode of hematuria. On clinical examination, is normal, the performance of an ultrasound found a hyperechoic mass of the left kidney, the performance of a CT scan which objectified a mass of the lower lip of the left kidney, well limited, heterodense, enhanced after injection of contrast agent. Measure 60 mm of major axis arrive at the contact of the psoas (Figures 1 & 2). The patient underwent a left partial nephrectomy by the subcostal. On macroscopic examination, solid tumor mass measuring 7 x 6 cm of whitish appearance. Microscopic examination revealed a tumor proliferation made up of three components : one vascular, one component of epithelioid cell represents more than 70% of the tumor, the third is adipocytic. Absence of atypical mitosis. This analysis concluded that there was a benign epithelioid angiomyolipoma of the left kidney.

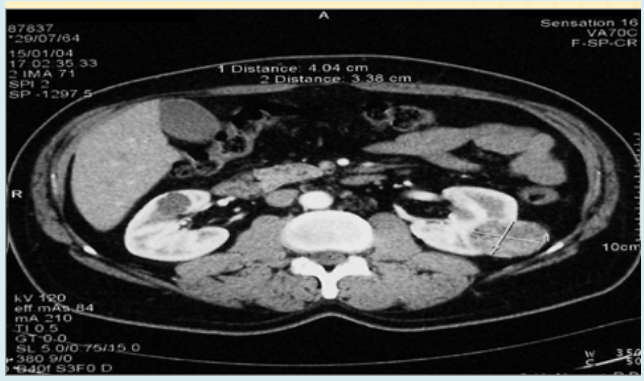

Figure 1: Renal CT scan performed during the initial assessment. Left renal mass 39x32mm with double component.

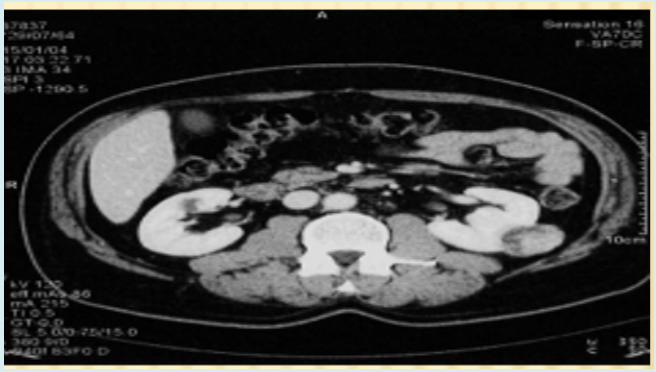

Figure 2: Renal computed tomography performed 2 years after the first. Left kidney lesion, tumor-type, without fat contingent, heterogeneous, enhanced after injection.

Discussion

(Angiomyolipoma (AML) is a benign tumor of the kidney composed in varying proportions of abnormal vessels, smooth muscle cells, and adipose tissue. This tumor accounts for approximately 3% of solid tumors in the kidney [3]. In 80% of cases, AML is unique and in 20% of cases, the lesions are multiple and bilateral, then associated with phacomatoses, in particular tuberous sclerosis of Bourneville. In the majority of cases, AML is asymptomatic and incidentally discovered during an ultrasound or abdominal CT scan [3]. It is visualized in ultrasound a non-specific hyperechoic renal mass syndrome. The thin-section CT scan found a negative hypodensity, varying between -10 and -30 Hounsfield Units (UH), corresponding to the fatty component [4]. The presence of a fatty component within a renal tumor mass, in computed tomography is almost pathognomonic of AML. However, 5% of AMLs do not have fat visible on imaging. In addition, cases of renal cell carcinoma with areas of radiological fat density have been reported, in cases of hemorrhage, necrosis or invasion of sinus or perirenal fat [5]. Other fatty tumors or tumors containing a fatty contingent can cause false positives, such as lipomas, liposarcomas or nephroblastomas in children. MRI has a higher sensitivity than computed tomography. This examination found a fat hypersignal in T1 and a hyposignal in T2 [5]. Fat saturation or phase and phase opposition sequences can be used to refine the diagnosis.

Mild forms of AML with an epithelioid component have an excellent prognosis. Malignant forms, on the other hand, are dreadful with recurrences or even a metastatic evolution possible even after radical surgery and require prolonged monitoring. They are exceptional: only 12 cases are found in the literature. They most often have in common a computed tomography of renal cell carcinoma and a rapid degrading course. In most cases, it is difficult to say whether it is a «primary» malignant AML due to major cytonuclear atypia of the epithelioid cells or malignant degeneration of an AML [6,7].

Conclusion

AML is classically a benign tumor of slow progression, for which a simple radiological monitoring is sufficient when it is asymptomatic, less than 4 cm in size and with a typical CT scan. In AML with an epithelioid component, the CT scans may change and mimic a malignant tumor leading to a sometimes radical excision procedure. The limited means of rectifying the diagnosis preoperatively and the malignant potential of this condition may encourage such an attitude.

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