Commentary

The authors presented a case of a 24-year-old woman with tuberous sclerosis and a computed tomography (CT) scan of the abdomen showing 6.5 × 5.0 × 4.4 cm mass lesion in kidney with significant para-aortic lymphadenopathy with no evidence of fat in the mass. With a provisional diagnosis of renal cell carcinoma (RCC), she underwent radical left nephrectomy. Histological examination showed multicentric angiomyolipoma (AML) involving kidney and para-aortic lymph nodes.

AML is found in 0.3% of all autopsies and in 0.13% of the population screened by ultrasonography. Approximately 20 to 30% of AMLs are found in patients with tuberous sclerosis syndrome, an autosomal dominant disorder characterized by mental retardation, epilepsy, and adenoma sebaceum. Approximately 50% of patients with tuberous sclerosis develop AMLs, and in this group of patients, AMLs is more likely to be bilateral and multicentric, and a tendency toward accelerated growth rates and symptomatic presentation has been reported.

CT has been the most useful modality for diagnoses of AMLs. A small amount of fat in a renal lesion on CT, as confirmed by less than 10 Hounsfield Units, is thought to virtually exclude the diagnosis of RCC and it is considered diagnostic of AML. However, the preoperative radiological diagnosis of AMLs with minimal fat component poses a diagnostic challenge. A surprisingly high number of resected AMLs was not suspected radiographically (33 to 65% in a series of 209 patients) indicating the importance of precise radiographic characterization to minimize nephrectomy for fat poor AML, which should remain a research priority.[1]

Several imaging techniques in both CT scan and magnetic resonance imaging (MRI) have been extensively studied to differentiate fat-poor AML from RCC. In a study with 81 patients, Kim *et al*. concluded that homogeneous enhancement (observed in 79% of AMLs×5% of RCCs) and prolonged enhancement pattern (observed in 58% of AMLs×10% of RCCs) on biphasic helical CT were valuable predictors for differentiating AML with minimal fat from RCC at multivariate analysis.[2] Other authors reported similar opinion.[3] Unenhanced thin-section CT (5 mm or thinner) should optimize fat detection.[4] Other techniques, such as pixel histogram analysis of unenhanced CT scan images, showed contest results. However, Milner *et al*. related that not all fat-poor AMLs have high attenuation on unenhanced CT, which makes this an unreliable finding for characterizing these lesions.[5]

Some authors reported that MRI is an effective means of detecting both the macroscopic and microscopic adipose components of AMLs, and is especially successful in the identification of minimal fat AMLs. Comparing the fat-suppressed and non-fat-suppressed images on MRI[6] and utilizing the opposed-phase chemical shift technique[7] may be helpful in difficult cases.

In synthesis, a high degree of attenuation on unenhanced CT images, enhancement on contrast-enhanced CT images, hypointensity on T2-weighted MRI images, enhancement during the early phase on dynamic MRI images, and abundant pulsatile blood vessels on color Doppler examination should be used as indicators of AML with minimal fat to differentiate such lesions from RCC.

Yearly radiological follow-up of indeterminate renal masses is recommended for individuals with tuberous sclerosis and any mass that shows a progressive increase in size should be treated as suspicious for malignancy, indicating either biopsy or surgical excision.

As discussed by authors, fat-poor AMLs defy diagnosis and raise the suspicion of RCC. This suspicion if further emboldened by the presence of enlarged regional lymph nodes may result in radical nephrectomy. More than 40 cases of renal AML with lymph nodal involvement have been reported in the literature and the natural history is the same presented by authors: The patient underwent radical nephrectomy with a provisional diagnosis of RCC and histopathological examination showed multicentric AML.[8] The consensus from other studies suggests that this phenomenon represents a multifocal version of the tumor rather than a metastatic disease, a belief that arose from the benign appearance of tumor and lymph nodes on pathological examination and the lack of evidence of distant spread on follow-up.

A nephron-sparing approach, by either selective embolization or partial nephrectomy, is clearly preferred in patients with small AMLs requiring intervention because of symptoms, in patients with tuberous sclerosis or multicentric AML, and in patients for whom preservation of renal function is at issue. It is critical to note that due to the bilateral nature of the renal lesions in tuberous sclerosis, and in order to preserve functional renal mass, nephrectomy should not be undertaken without a very careful risk-benefit analysis.[6]

Despite all the new imaging techniques available, the precise preoperative radiological diagnosis of fat-poor AMLs remains challenging and sometimes impossible, especially in cases of fat-poor AML associated with lymph node involvement.

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