

Clinical Quiz 1

Answer:

Solid fat-poor lesions in the kidneys of a patient with TSC are most often angiomyolipomas, but they can occasionally be oncocytomas or renal cell carcinoma (RCC). Oncocytomas occur more frequently in TSC than in the general population, where they are uncommon benign adenomatous hamartomas accounting for only 3–5 % of renal parenchymal tumors. RCC occurs in 3–5 % of patients with TSC at an average age of 28–50 years, with very few reports in childhood. However, not all reported RCC cases have been studied in detail (e.g., histology) with possible misclassifications. MRI is the best imaging method for detecting fat-poor lesions. However, there are currently no imaging methods that clearly distinguish a fat-poor angiomyolipoma from an RCC. Growth rate has been suggested to help the differentiation between these lesions. The current recommendations suggest a diagnostic needle biopsy for fat-poor lesions growing faster than 0.5 cm per year to distinguish RCC from angiomyolipoma.

If interested, you can read:

De Waele L, Lagae L, Mekahli D. Tuberous sclerosis complex: the past and the future. *Pediatr Nephrol*. 2015 Oct;30(10):1771-80.