# Giant renal angiomyolipomas in a patient with tuberous sclerosis

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A 71-year-old woman presented with left flank pain, fever and abdominal distension. She had been known to have an unknown neurological disease. Neurological examination and skin examination were unremarkable. Abdominal examination revealed soft abdomen with mild generalized tenderness. Laboratory investigation revealed eGFR of 43 ml/min. Contrast-enhanced computed tomography (CT) of abdomen showed giant angiomyolipomas (AMLs) in both kidneys [Figure 1a and b]. Small amount of enhancing renal parenchyma could be identified on both sides. There was excretion of contrast by the kidneys. A small AML was also seen in the right lobe of liver. CT images of included lung bases, demonstrated cystic lung disease with focal consolidation in left lower lobe, probably pneumonia. There were ill-defined sclerotic lesions in multiple dorsal vertebrae. Cranial CT revealed bilateral small calcified subependymal nodules [Figure 2], confirming the diagnosis of tuberous sclerosis (TS). The symptoms were attributed to left lower pneumonia and treated with antibiotics. The patient refused any immediate treatment for AMLs.

Tuberous sclerosis, also known as Bourneville disease, is an autosomal dominant neurocutaneous syndrome, with variable expressivity, characterized by hamartomatous lesions in multiple organs. Its prevalence reported ranges from 1 in 6,000–12,000. The classical Vogt's triad of adenoma sebaceum, seizures and mental retardation is seen in less than half of patients, which makes imaging studies important for diagnosis.

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Renal cysts and AML are the commonest abdominal findings in TS, the latter seen in up to 75% of the cases.[1] Multiple hepatic AML are often found in patients with TS and particularly in patients with bilateral diffuse renal AML.[2] Rarely, renal cell cancer can be seen in younger patient with TS.[3] AML can range in size from 3 mm to those larger than 10–15 cm. AML associated with TS are usually larger than sporadic cases. The most fearful complication of an AML is spontaneous rupture and

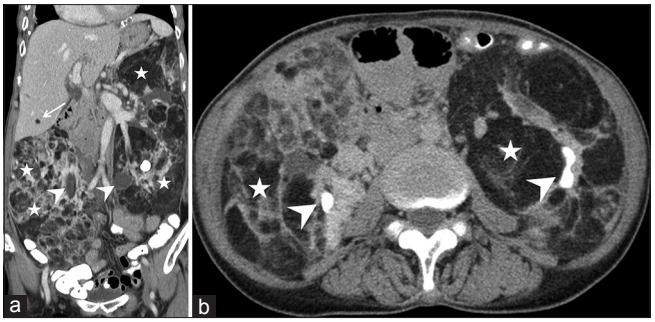


**Figure 1: (a) Coronal reconstruction of contrast-enhanced computed tomography (CT) abdomen shows infiltrative fat containing giant masses (white asterisks) in retroperitoneum, replacing almost entire both kidneys. Renal pelvis is seen on either side (white arrowheads). There is a fat containing lesion in right lobe of liver (white arrow). (b) Axial CT image of delayed contrast study shows bilateral fatty angiomyolipomas (white asterisks) with excretory contrast in renal pelvis (white arrowheads)**

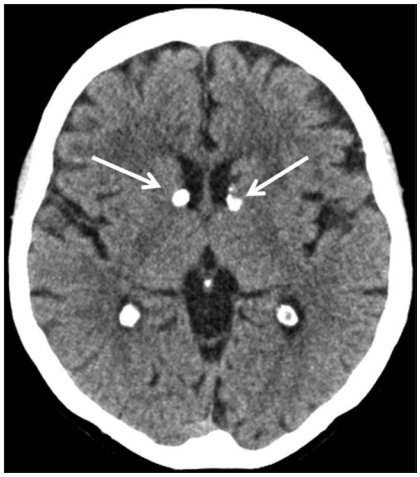

**Figure 2: Unenhanced computed tomography head shows bilateral calcified subpendymal nodules (white arrows), classically located along the striothalamic groove of the lateral ventricles**

perinephric hemorrhage. Spontaneous hemorrhage is related to the size of the lesion. Smaller lesion <4 cm are unlikely to bleed and hence are not treated. The lesions between 4 and 8 cm should be closely followed up and treated, if the patient becomes symptomatic. Larger lesions more than 8 cm are treated by transcatheter embolization or conservative surgery.[4] Calcified subependymal nodules are seen in approximately 95% of patients with TS and are virtually diagnostic of TS. Based on the major and minor clinical-radiological criteria[5], our patient falls in definite TS category.

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