

## Tuberous Sclerosis Complex

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A 30-year-old woman presented to our outpatient clinic with a painless left abdominal mass, which had gradually increased in size over the past two years. She gave history of seizures at one year of age. Her general and neurological examinations were normal. She had a large left flank mass, which was bimanually palpable and ballotable.

Contrast enhanced computed tomography revealed a large well-circumscribed heterogeneous retroperitoneal mass measuring 25 × 20 cm arising from the left kidney with predominant fat attenuation. Multiple smaller lesions were seen in the opposite kidney. Features were suggestive of diffuse angiomyolipomatosis of the kidneys. Hypodense lesions were also seen involving the lungs- lymphangiomyomatosis and the liver- hepatic angiomyolipoma, a rare finding.

A diagnosis of tuberous sclerosis complex (TSC) was made and presence of other stigmata of the syndrome was ruled out. She underwent a nephron sparing excision of the left renal mass and the diagnosis of angiomyolipoma was confirmed on histopathology examination. She made an uneventful recovery. Regular follow-up and screening of family members were advised.

Tuberous sclerosis complex is a multisystem, autosomal dominant disorder resulting from mutations of *TSC1* or *TSC2* genes.<sup>(1)</sup> The diagnostic criteria of TSC consist of a set of major and minor features.<sup>(2)</sup> Management of TSC must include appropriate diagnosis and long-term follow-up enabling early recognition of potential life-threatening complications. Finally, genetic counseling should be offered to patients to aid with family planning.

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### REFERENCES

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