Bilateral renal angiomyolipoma presenting as tuberous sclerosis syndrome

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DESCRIPTION

A 30-year-old woman presented with generalised abdominal pain of 3-month duration. She had a history of epilepsy and low intelligence since childhood. On examination, she was found to have multiple painless papules on her face (figure 1) and periungual nodules involving all the toes (figure 2). CT of the abdomen suggested large bilateral angiomyolipoma of the kidney (figure 3). Assuming it to be a case of tuberous sclerosis complex, her neurological, dental and cardiology evaluations were carried out and found to be normal. After explaining the disease and prognosis, she was managed conservatively and advised to be under regular follow-up.

Bourneville or tuberous sclerosis syndrome is an autosomal dominant entity with the incidence

Figure 1 Multiple papules (adenoma sebaceum) over the patient's face.

Figure 2 Periungual fibroma involving all the toes.

Figure 3 CT of the abdomen suggesting large bilateral angiomyolipoma of the kidney.

of 1 in 6000 to 1 in 10 000.¹ In these patients, the typical triad of epilepsy, low intelligence and adenoma sebaceum are found, as in our case. Renal angiomyolipomas are found in 50–80% of tuberous sclerosis patients and are usually bilateral with a high female preponderance suggesting hormonal influence.² There are certain major and minor criteria defined for establishing the diagnosis, in our case, three major criteria—renal angiomyolipoma, adenoma sebaceum and periungual fibroma involving all the toes—were found.¹ Renal

Learning points

- ▶ Tuberous sclerosis can present late.
- ▶ Diagnosis can be established by proper history-taking and examination.
- ▶ Continuous monitoring is a must to decrease mortality.

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angiomyolipoma can increase in size, leading to perirenal haemorrhage.³ Patients may develop renal insufficiency as well. Similarly, other organs can have manifestations such as cerebral tumours and cardiac rhabdomyomas, although in our case none were present. However, patients need regular follow-up to decrease the mortality.

Competing interests None declared.

Patient consent Obtained.

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