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Case report

Giant angiomyolipoma in a tuberous sclerosis patient and review of the literature



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KEYWORDS

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Mammalian target of
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Abstract

Introduction: About 20% of renal angiomyolipomas (RAML) are associated with tuberous sclerosis complex (TS). About 34–80% of patients with TS present with RAML. RAMLs associated with TS are at higher risk of potentially life-threatening hemorrhage and hypovolemic shock. Only a few case reports of giant RAML, defined as larger than 10 cm in diameter, and its management, have been reported.

Observation: We present a 21 year old woman with abdominal distension over the last 2 years. A contrast-enhanced CT scan revealed a giant RAML on the left side. Based on the presence of at least 3 major features of the clinical diagnostic criteria of tuberous sclerosis complex, the diagnosis was made. An open nephrectomy was performed. Therapeutic options described in literature are conservative management, medical treatment with mTOR inhibitors, arterial embolization, radioablation and partial or total nephrectomy.

Conclusion: In giant TS-associated RAML total nephrectomy, rather than conservative treatment, is the treatment of choice in order to reduce the risk of potentially life-threatening bleeding.

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Introduction

Renal angiomyolipomas (RAML) are rare, benign tumors consisting of vascular elements, smooth muscle and adipose tissue. About

20% of them occur in patients with the tuberous sclerosis complex (TSC) [1–3]. In the presence of bilateral lesions, one should consider a diagnosis of this neurocutaneous condition that also causes benign tumors in the brain, heart, eyes, lung and skin [1]. Smaller RAMLs are often incidentally found on radiological examination, whereas RAMLs larger than 4 cm are at higher risk of bleeding [1,3]. RAML associated with TS are more likely to lead to potentially life-threatening retroperitoneal hemorrhage due to their size, and multifocal and bilateral nature [1–3]. Embolization, partial or total nephrectomy as treatment of larger RAMLs have

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Figure 1 A. Facial angiofibromas. B. Hypomelanotic macule on the left chest >5 mm.

been described [3]. Only a few case reports about giant RAML, which is defined as larger than 10 cm in diameter, and its management, have been published [4]. We present a case of giant RAML in a young woman who presented with left abdominal pain and in whom the diagnosis of tuberous sclerosis was made.

Case presentation

A 21 year old woman was referred to the urology department of Groote Schuur Hospital in Cape Town (South Africa) in November 2017 because of weight loss and progressive abdominal distension over the last two years. No relevant medical or family history was reported. An ultrasound of the kidneys which was done at the referring hospital reported a big solid mass on the left side of the abdomen.

Skin inspection showed multiple (>10) facial angiofibromas and 4 hypopigmented macules on the chest, lower back and right buttock (Fig. 1). Multiple skin lesions with a ‘confetti’ appearance were seen on both arms and legs. No other dermatologic or dental features of TS were seen. Inspection of the abdomen showed abdominal distension on the left side. A large non-tender mass was palpated in the left upper quadrant extending beyond the midline.

Laboratory results showed anemia (Hb of 9,4 g/dl) and microscopic hematuria. A contrast enhanced CT-scan of the abdomen revealed a heterogeneous left renal mass with areas of soft tissue and fat components (Hounsfield units of $-68,97$), measuring $123,0 \times 159,4$ mm (Fig. 2). The right kidney showed multiple cortical hypodensities with the largest measuring 11 mm. Both lesions were reported as suspicious for angiomyolipoma (AML).

Blood transfusion with 2 units of packed cells was given. A left radical nephrectomy was performed through a chevron incision under general anesthesia. A mass with a diameter of $290 \text{ mm} \times 215 \text{ mm} \times 120 \text{ mm}$ and a weight of 3,58 kg was removed (Fig. 3). The procedure was complicated intraoperatively by pancreatic tail injury requiring distal pancreatectomy. She developed a pancreatic leak which required a stay in the Intensive Care Unit. The leak resolved spontaneously. The patient was discharged 3 weeks after surgery.

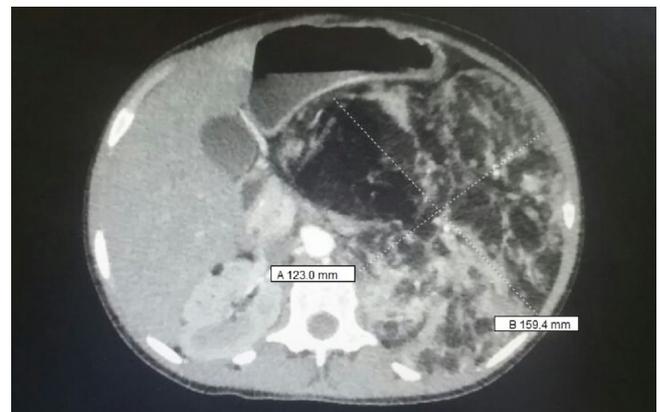


Figure 2 Axial image from a contrast enhanced computed tomography (CT) shows a primarily fat containing mass arising from the left kidney suggestive of a giant angiomyolipoma (Hounsfield units of $-68,97$).



Figure 3 Excised tumor of 3,58 kg, showing a mixture of mature adipose tissue and thick walled blood vessels.

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