



# Bilateral Renal Angiomyolipoma: Think of Bourneville's Tuberous Sclerosis

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## Abstract

Angiomyolipoma is a benign hamartomatous tumor. It constitutes the most frequent renal disease encountered in Bourneville's tuberous sclerosis, the bilateral character of which is highly suggestive. We report the observation of a 48-year-old patient with Bourneville's tuberous sclerosis with bilateral angiomyolipoma.

**Keywords:** Renal angiomyolipoma; Bourneville's tuberous sclerosis; Bilateral

## Introduction

Angiomyolipoma is a rare benign kidney tumor in the general population. It is made up of adipose tissue, smooth muscle and dysmorphic blood vessels. It represents the first cause of kidney damage during Bourneville's Tuberous Sclerosis (BTS) [1]. We report a case of BTS with bilateral renal angiomyolipomas.

## Case Report

A 48-year-old patient, with a family history of a similar case in the brother, presented for chronic low back pain, without hematuria. The urogenital examination found sensitivity without lumbar contact. The dermatological examination showed "fibrous plaque" with multiple tumors in the face, neck and shoulders corresponding to "angiofibromas" (Figure 1). As well as periungual fibromas corresponding to "Koenen's tumors" (Figure 2). Renal ultrasonography revealed rounded, heterogeneous and bilateral hyperechogenic formations. Abdominal CT scan showed a bilateral kidney process with multiple tissue component and greasy islets, suggesting renals angiomyolipomas (Figure 3). The rest of radiological exams including echocardiography and brain CT scan, was normal. On the biological assessment, the blood creatinine level was 15 mg / l, with a clearance at 60 ml / min, the 24 hours proteinuria was normal. The diagnosis of BTS was retained because of the presence of three major criteria



**Figure 1** Clinical photo of our patient showing face "angiofibromas" and "fibrous plaque".



**Figure 2** Clinical photo of our patient showing "Koenen's tumors" in the toes.

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- two criteria being sufficient – face "angiofibromas", "Koenen's tumors" and renal angiomyolipomas. Due to the bilateral nature of kidney lesions and the absence of hematuria, our approach was a surgical abstention with regular clinical, biological and radiological monitoring.



**Figure 3** Abdominal CT scan showing the double tissue and especially greasy component of renal angiomyolipomas in both kidneys.

## Discussion

BTS is an autosomal dominant phacomatosis. It is characterized by the growth of multiple hamartomatous lesions (brain, skin, kidneys, heart, lungs, retina...) [2]. There are two genes involved in STB, TSC1 at chromosome 9q and TSC2 at chromosome 6p [3]. After neurological damage, kidney damage is the second leading cause of death for all age groups [4]. Kidney disease is found in 94% of patients with BTS, and is dominated first by angiomyolipoma (AML) followed by cortical cysts and kidney cancer [5]. In fact, AML represent 75 to 80% of renal affections during BTS [1]. Renal AML is in most cases asymptomatic. The diagnosis is made by ultrasonography and confirmed by the CT scan showing a heterogeneous tissue mass with a greasy component. MRI is the most sensitive examination, but it will only be done if there is doubt about the diagnosis. The risk of spontaneous bleeding becomes significant when its size is greater than 4 cm, responsible for hematuria and spontaneous rupture in the retroperitoneum [6]. Imaging surveillance is required when the tumor is small. For tumors larger than 3.5 cm, we can opt for an arterial embolization by angiography. Nephrectomy will be reserved for cases with massive hemorrhage or lesion larger than 4 cm [6,7].

## Conclusion

Screening for renal disease should be systematic in patients with BTS complex through searching for hematuria and proteinuria, measuring serum creatinine and performing renal ultrasonography. Conservative treatment is recommended whenever possible and nephrectomy is reserved for extreme cases with an increased risk of bleeding.

## Authors Contributions

All the authors contributed to the realization of this work. All authors have read and approved the final version of the manuscript.

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