



Giant Bilateral Angiomyolipoma Associated with Tuberous Sclerosis of Bourneville: Exceptional Case and Therapeutic Dilemma

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Author's contribution

The sole author designed, analyzed, interpreted and prepared the manuscript.

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Case Study

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ABSTRACT

Angiomyolipoma (AML) is a mesenchymal tumour composed of variable proportions of adipose tissue and vascular and smooth muscle elements. It can cause potentially life-threatening complications.

This report aims to describe a bilateral angiomyolipoma associated with tuberous sclerosis of Bourneville; the second aim is to discuss the treatment modalities of this disease.

A 51-year-old woman with abdominal mass and characteristic triad: Epilepsy, mental retardation and skin lesions (adenoma) (Fig. 1), with no notion of hematuria or abdominal pain, abdominal ultrasonography and computed tomography showing bilateral renal masses of 28.4×17 cm on the left kidney and 22× 11.7 cm on the right, respectively (Fig. 2-3). Serum creatinine was 13.4g/l.

Body-scan imaging finds cardiac rhabdomyoma (Fig. 4) and brain calcifications (Fig. 5). the patient underwent surveillance with scan imaging every month, and at the last control, she was asymptomatic, serum creatinine was still normal. The management of giant bilateral AML is a complex and multifactorial decision. Patients can knowingly choose an active surveillance program, even with giant AML, but the economic situation and mental status can limit the therapeutic choice.

Keywords: Angiomyolipoma; tuberous-sclerosis; bourneville.

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1. INTRODUCTION

Angiomyolipoma (AML) is a renal mesenchymal tumor composed of varying proportions of adipose tissue and vascular and smooth muscle elements. It can cause significant, potentially life-threatening complications. The first aim of this case report is to describe a bilateral angiomyolipoma associated with tuberous sclerosis of Bourneville (TSB), the second aim is to discuss the treatment modalities of this disease.

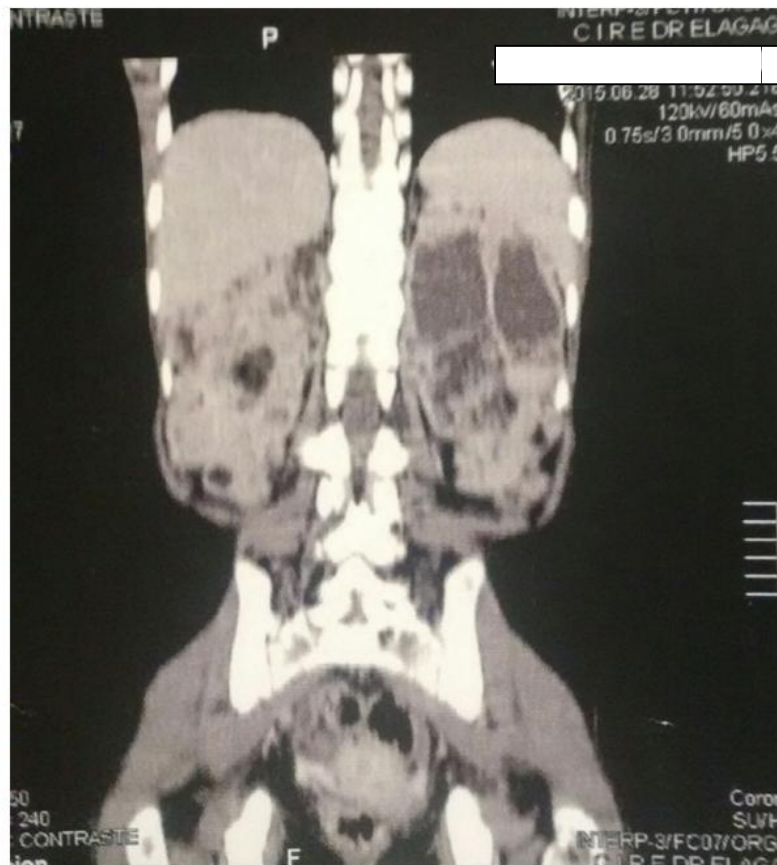
2. CASE PRESENTATION

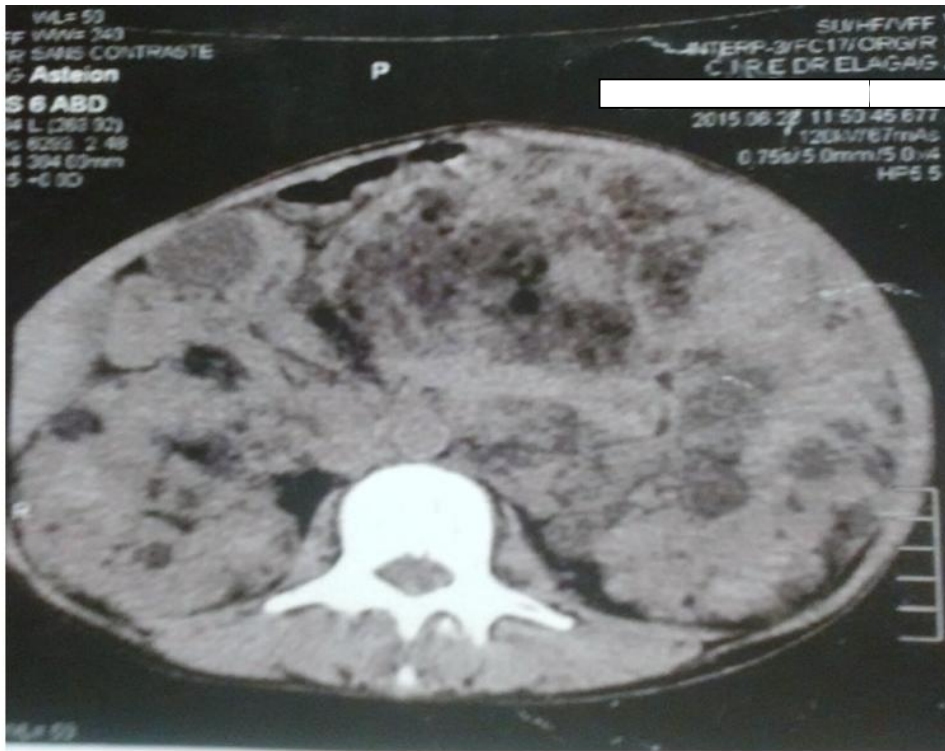
A 51-year-old woman with an abdominal mass and a characteristic triad: epilepsy, mental retardation and skin lesions (adenoma) (Fig. 1), we found no evidence of hematuria or abdominal pain, she underwent ultrasonography and then computed tomography, which showed the presence of bilateral renal masses of 28.4×17 cm on the left kidney and 22× 11.7 cm on the right kidney (Fig. 2-3). Serum creatinine was: 13.4g/l.

The patient also underwent a whole body examination, which revealed cardiac rhabdomyomas (Fig. 4) and calcifications in the brain (Fig. 5). She underwent a surveillance regimen with monthly imaging, and at the last follow-up she was asymptomatic and serum creatinine was still normal. Treatment of giant bilateral AML is a complex and multifactorial decision. Patients may make a conscious decision to follow an active surveillance program, even in the case of giant AML, but economic situation and mental status may limit therapeutic choices.



Fig. 1. Characteristic skin lesions of tuberous sclerosis de bourneville





Figs. 2 and 3. Abdominal scan showing giant bilateral angiomyolipoma

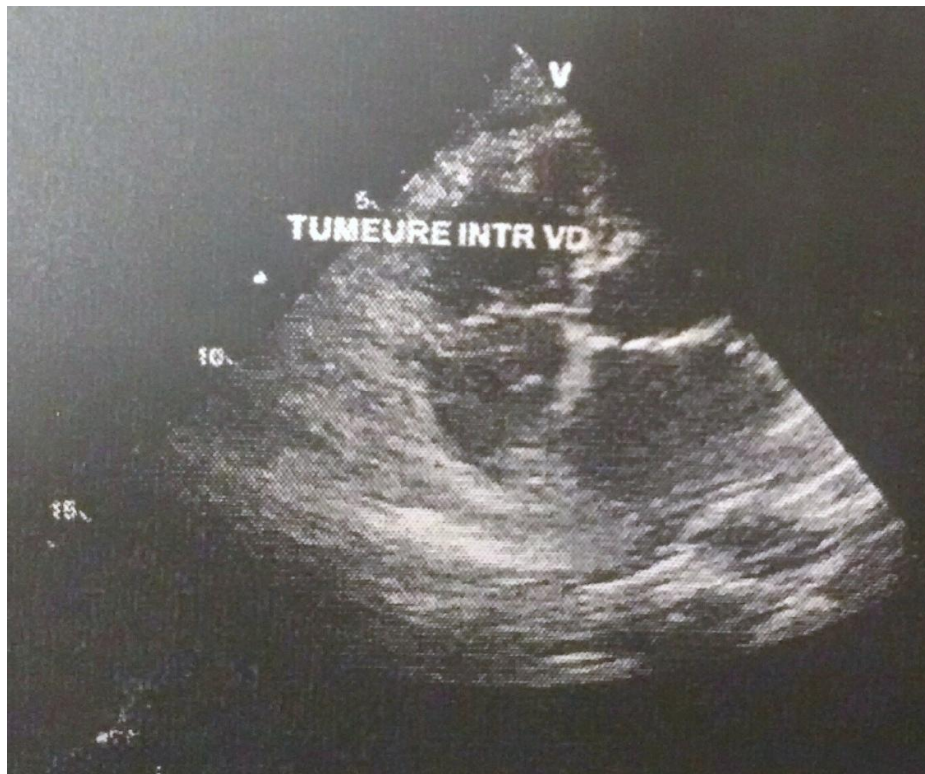


Fig. 4. Cardiac rhabdomyome

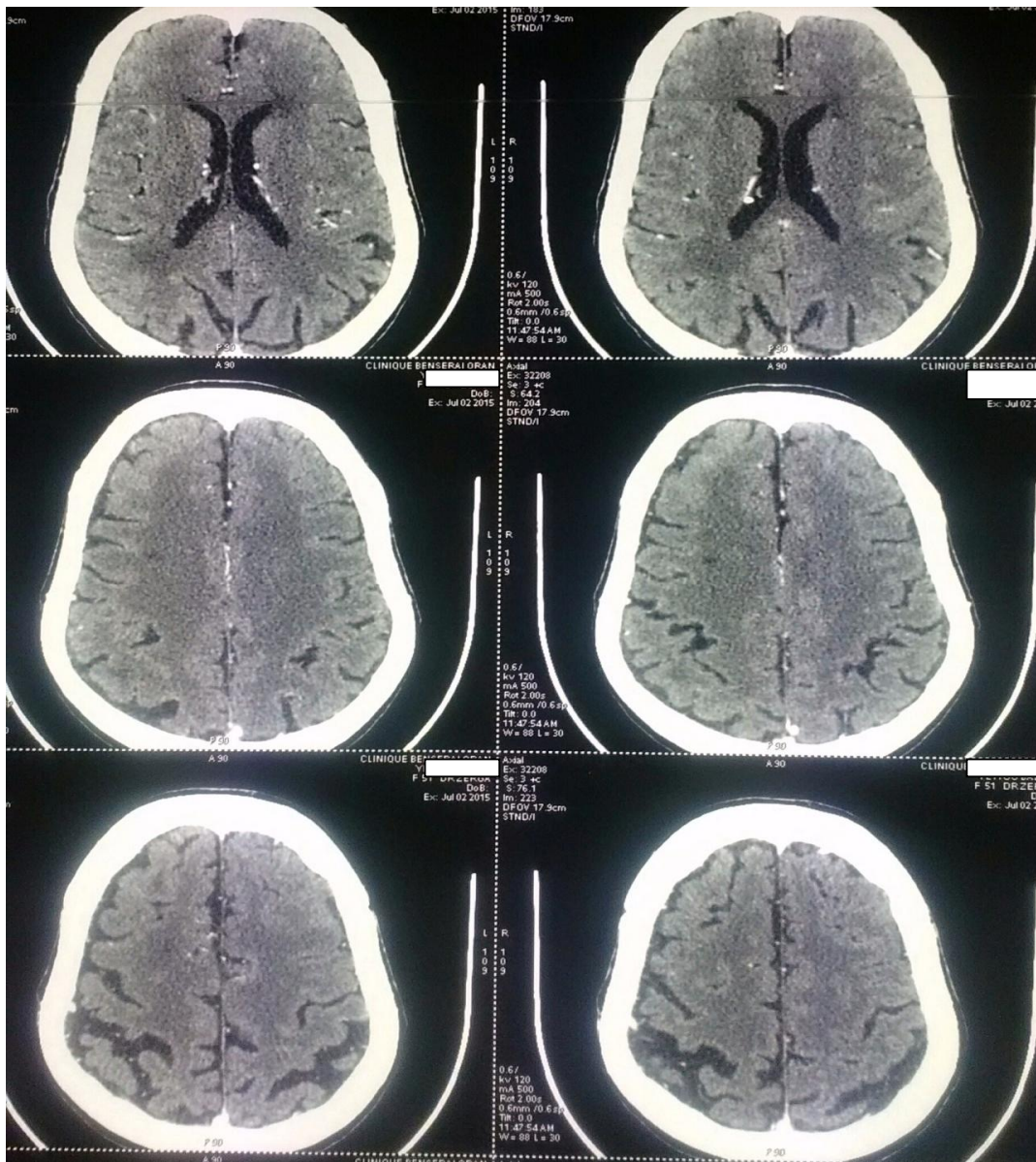


Fig. 5. Calcified cortical and subependymal tubers

3. DISCUSSION

Angiomyolipomas represent 1% to 3% of solid renal tumors, with females predominance [1].

Bilateral and multiple localizations are observed in 50% to 80% of cases. To our knowledge, the volume has never been 28 cm in the published literature, and the maximum diameter has ranged from 2 to 15 cm [2].

TSB is a dominant autosomal affection with variable pénétrance.

The association of bilateral bilateral and TSB is exceptional, we can say that in such a situation a dangerous risk of bleeding is possible, but the size alone is not sufficient to assess the risk of bleeding.

The importance of vascular and fatty contingencies and the presence of intra-tumoral microanomalies greater than 5 mm could also be risk factors.

Unfortunately, detection of micro-aneurysms requires arteriography embolisation generate in

6% of cases a "Post-embolization syndrome" with low back pain and fever that can be controlled with non steroidal anti-inflammatory treatment or short-term corticosteroid treatment- the reduction in diameter depends on the proportion of vascular and fat. Embolization can be repeated during surveillance [3].

Cryoablation, radiofrequency and angiogenesis inhibitors may be used. m TOR-inhibitors may also play a role.

Because of their potential adverse effects, mTOR inhibitors should be prescribed only by specialized teams and, when possible, in the context of clinical trials.

m TOR inhibitors should not be used as first-line therapy in the treatment of renal AMLs. Some authors recommend prophylactic treatment, particularly in large tumor .

AMLs that accumulate risk factors for bleeding (size 80 mm, predominant vascular contingencies, microaneurisms), if possible by embolization.

Patients in whom follow-up or access to emergency care may be inadequate [4].

4. CONCLUSION

Giant bilateral angiomyolipomas associated with tuberous sclerosis of Bourneville remain the exception.

Diagnosis is straightforward, assessment is mandatory to evaluate risk. Therapeutic options: Surgery, endovascular, endovascular, adapted to the situation.

The management of bilateral giant angiomyolipoma is complex, and the therapeutic decision depends on several tumor and patient factors.

DISCLAIMER

Some part of this manuscript was previously presented in the following conference.

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CONSENT

As per international standard or university standard, patient's consent has been collected and preserved by the authors.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Author has declared that no competing interests exist.

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