Total nephrectomy with nephron-sparing surgery for a giant bilateral renal angiomyolipoma: A case report

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Abstract. The current report presents the case of a 50-year-old female who presented with a giant bilateral renal angiomyolipoma (AML). After assessing the patient's symptoms, the tumor size and renal function, the patient underwent a total nephrectomy for the right AML, which measured 28x20x14 cm and nephron-sparing surgery was performed without preoperative selective angiographic embolization for a further 3 AMLs in the left kidney, the largest of which had a diameter of 12 cm. The introperative bleeding volume was at an acceptable level and the renal function was stable. No local recurrence was observed and no dialysis was required during follow-up. The strategy of the treatment in this report should be considered when treating similar tumors.

Introduction

Renal angiomyolipoma (AML) is a benign tumor containing elements of fat cells, smooth muscle and vascular tissue. There are two variants of this tumor; one is associated with tuberous sclerosis complex (TSC) and the other is sporadic. TSC is a multisystem, autosomal disease with various presentations, such as mental retardation, epilepsy, dermatological manifestation, renal AML and pulmonary lymphangiomyomatosis (1,2). Approximately 20% of renal AML cases are associated with TSC, in which 70-90% are bilateral (3,4).

Early studies summarized the clinical management of renal AML and established that renal-preserving treatments, including selective angiographic embolization (SAE) and nephron-sparing surgery (NSS), are preferred (5,6). SAE is considered to be an effective and durable treatment for preventing bleeding in large renal AML, particularly for patients with an aneurysm. Recent studies demonstrated that 91-93% of renal AML cases are successfully embolized, however, the cost of surveillance and the morbidity of re-embolization were not negligible (7,8). Although, NSS has been widely performed in the past two decades with good results, the majority of results are for sporadic renal AML cases, and the tumors are typically are unilateral and small (9,10).

In the current report, a case of giant bilateral renal AML without TSC is presented. The patient underwent a total nephrectomy for a right AML measuring 28x20x14 cm and NSS was performed for 3 AMLs in the left kidney, the largest of which had a diameter of 12 cm. The patient provided written informed consent to participate in the study and the study was approved by the Ethics Review Committee of the Third Xiangya Hosiptal of Central South University (Changsha, China).

Case report

In April 2013, a 50-year-old female presented to the Department of Urology at the Third Xiangya Hospital of Central South University with a flank mass and abdominal fullness, which were confirmed by palpation. The patient did not present with any additional symptoms that met the diagnostic criteria for TSC. The serum creatinine level was 0.93 mg/dl (normal range, 0.51-1.19 mg/dl), and the hemoglobin level was 131 g/l (normal range, 115-150 g/l). The results of coagulation function and biochemistry were within the normal limits. Abdominal computed tomography (CT) revealed a large AML measuring 28x20x14 cm (Fig. 1) arising from the right kidney and deviating the rest of the abdominal contents across the midline, and a 12x9x6 cm AML arising from the upper pole of the left kidney. Several small AMLs were also observed in the left kidney (Fig. 2). The glomerular filtration rate (GFR) was evaluated using a technetium 99 m-diethylenetriminepentaacetic acid (DTPA) nuclear renal scan, revealing that the right renal function was severely impaired, with a right kidney GFR of 12.57 ml/min. A right total nephrectomy was performed, and the postoperative serum creatinine and hemoglobin was 0.97 mg/dl, 120 g/l, respectively. Histopathological analysis revealed that the renal mass was composed of fat, vascular structures and smooth muscle, indicating a diagnosis of AML. Upon immunohistochemical analysis, the tumor cells stained positive for smooth muscle actin, human melanoma black 45 and vimentin, and negative for cluster of differentiation 34, desmin and S-100. Furthermore, the Ki-67 proliferation rate was <3%. Based on the histopathological and immunohistochemical findings, a diagnosis of a renal AML was determined.

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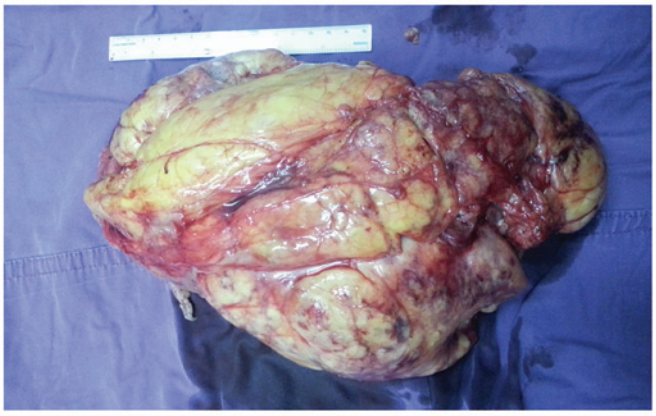

Figure 1. Clinical appearance of the giant resected right renal angiomyolipoma.

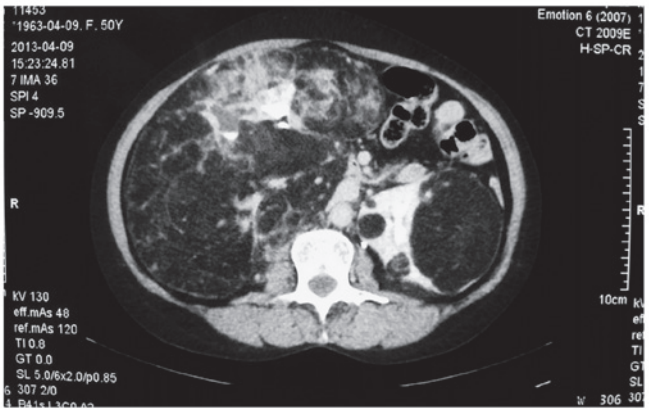

Figure 2. Computed tomography scan demonstrating giant bilateral renal angiomyolipomas.

NSS for the AML present in the left kidney was performed 3 months after the right total nephrectomy. A CT scan was conducted prior to NSS to evaluate the tumor size in the left kidney and for the follow-up investigation, and demonstrated several AMLs (Fig. 3). NSS was performed using a flank incision in the eleventh intercostal space. Following careful dissection, renal vessel occlusion was identified. A total of 3 AMLs were resected: The largest tumor (diameter, 12 cm)was resected and the other two tumors were enucleated. The total ischemia time was 24 min. Histopathological analysis revealed that the 3 masses were composed of fat, vascular structures and smooth muscle, consistent with a diagnosis of AML. Upon immunohistochemical analysis, the tumor cells stained positive for smooth muscle actin and vimentin, and negative for human melanoma black 45, desmin and S-100. In addition, the Ki-67 proliferation rate was ~5%. The preoperative and postoperative serum creatinine and hemoglobin levels were 0.93 mg/dl and 129 g/l, and 1.27 mg/dl and 102 g/l, respectively. The estimated intraoperative bleeding volume was 500 ml and 2 units of blood transfusion was performed.

Regular routine blood tests, biochemistry, ultrasonographic and/or CT scan were evaluated for follow-up investigation. No local recurrence was observed and no dialysis was required.

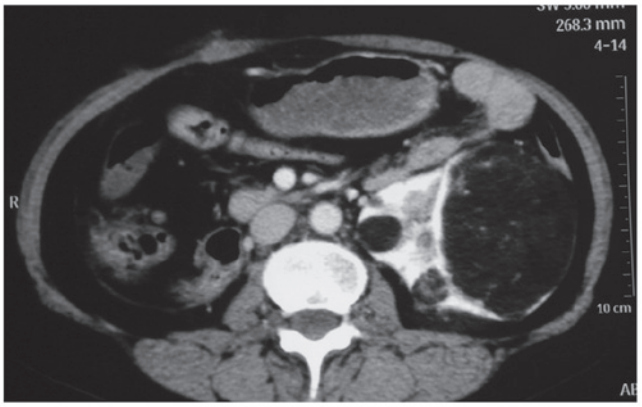

Figure 3. Computed tomography image of the left renal angiomyolipoma when the patient was hospitalized for the second time (axial view).

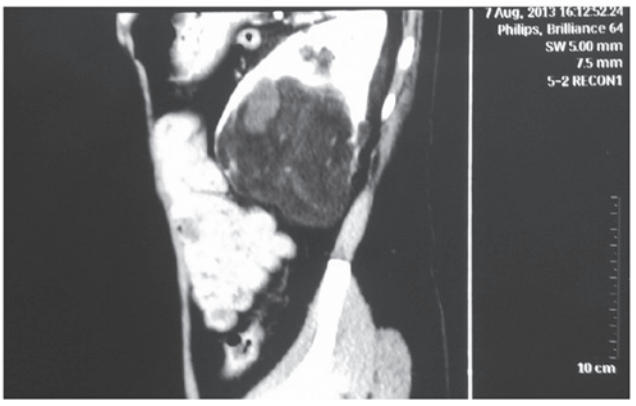

Figure 4. Computed tomography image of the left renal angiomyolipoma when the patient was hospitalized for the second time (sagittal view).

Discussion

The incidence of renal AMLs is rare and a proportion of AMLs are asymptomatic. Indications for intervention include dimensions >4 cm, associated symptoms, including spontaneous hemorrhage, pain and hematuria and the suspicion of malignancy (11). Asymptomatic AML and lesions that result in minor symptoms may be managed conservatively. However, the 4 cm threshold is not a definite criteria. Cavicchioli *et al* (12) reported a 50-year-old man with bilateral renal AML that was 27.5x19.5x21 cm on the left kidney and 28.5x19.6x27.5 cm on the right. The patient was asymptomatic and underwent a strict surveillance with magnetic resonance imaging every 4-5 months. At the last follow-up, the patient was asymptomatic and serum creatinine was normal.

AML has been reported to have the potential to increase in size by up to 4 cm per year (13,14). Given this nature, the majority of giant renal AML are symptomatic and treatments for renal AML are aimed at preserving renal function. SAE and NSS are indicated as effective renal function-preserving approaches. Ramon *et al* (7) analyzed the outcomes of 41 patients who received SAE for renal AML with a mean follow-up period of 4.8 years. SAE was performed through an artery using angiographic catheters. Digital subtraction angiographic examinations were used for diagnosis, followed by an aortogram for evaluation of renal arterial feeders to the tumor. Then, embolization was performed with different materials,

such as absolute alcohol and metal coils. Successful SAE was achieved in 40/41 patients (91%) and avoidance of surgery was achieved for 79/82 (96%) of the kidneys. No retroperitoneal hemorrhage was noted and 80/82 (98%) of the kidneys were preserved during follow-up. Boorjian *et al* (15) analyzed the outcomes of 58 patients who recieved NSS for renal AML, with a median follow-up of 8-years. The analysis demonstrated that NSS for renal AML results in preservation of renal function with acceptable rates of complication and low local recurrence rates.

For large renal AML, preoperative SAE is recommended to reduce excessive blood loss during surgery. Singla *et al* (16) reported a case of giant renal AML measuring 26x22 cm that was treated with preoperative embolization followed by partial nephrectomy without complications. Similarly, Luo *et al* (17) presented a 44-year-old man with bilateral giant AMLs with a diameter of 38.2 cm in the left kidney and 9.5 cm in the right kidney. The patient was treated successfully with preoperative SAE and NSS without hilar clamping. However, postembolization syndrome, the reduction of tumor volume and the optimal time window for NSS following SAE should be taken into consideration. Postembolization syndrome is a side effect of embolization procedures and consists of one or more of the following symptoms: Flank pain, fever, nausea and vomiting attributable to inflammatory mediators (18,19).

In the current report, the patient's symptoms, the tumor size and the damaged renal function were the main reasons for the decision to undertake a right total nephrectomy. As the patient had a solitary kidney when she was hospitalized for the second time, preoperative SAE was not performed in order to reduce the additional burden on the remaining renal function. Without preoperative SAE, the intraoperative bleeding volume was acceptable and the renal function was stable.

In conclusion, the management of giant bilateral renal AML is challenging and complex. A strict assessment, including the patient's symptoms, the tumor size and renal function, should be taken in choosing an effective and safe treatment approach. The present case demonstrates that NSS without preoperative SAE was effectively used to treat a solitary kidney. The strategy of treatment undertaken in this report may be considered when treating similar tumors.

Acknowledgements

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