

# Bilateral nephron-sparing surgery for giant bilateral renal angiomyolipomas associated with tuberous sclerosis complex

*Dev bilateral renal anjiyomyolipom ve tuberosklerozis kompleksli hastada bilateral nefron-koruyucu cerrahi*

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A significant number of patients with tuberous sclerosis complex (TSC) have renal angiomyolipoma and these patients tend to have bilateral lesions grow more rapidly. Bleeding is the most fearful complication in the patients. We described a young woman with TSC and multiple bilateral giant renal angiomyolipomatosis. We performed bilateral nephron-sparing surgery (NSS) and did not see any significant intraoperative or postoperative complication. Renal function was normal in the first year of follow-up.

**Key words:** *bilateral renal angiomyolipomas, tuberous sclerosis complex, nephron-sparing surgery*

Tuberosklerozis kompleksli hastaların önemli bir kısmında renal anjiyomyolipom vardır. Bu hastalardaki lezyonlar bilateral olma ve daha hızlı büyüme eğilimindedir. Renal kanama ise bu hastalığın en çok korkulan komplikasyonudur. Biz multipl, bilateral dev renal anjiyomyolipomlara sahip tuberosklerozisli bir genç bayanda bilateral nefron-koruyucu cerrahi yaptık. Hastada önemli bir intraoperatif veya postoperatif komplikasyon gözlemedik. Bir yıllık takip süresince hastanın böbrek fonksiyonları normal olarak kaldı.

**Anahtar sözcükler:** *bilateral renal anjiyomyolipoma, tuberosklerozis kompleksi, nefron-koruyucu cerrahi*

**R**enal angiomyolipoma is a rare benign neoplasm (1-3% of all renal tumors) composed of varying amounts of mature adipose tissue, smooth muscle and thick-walled blood vessels. A 20% of all renal AMLs are associated with tuberous sclerosis. In patients with TSC, AML is usually multiple, bilateral and symptomatic natures. Patient's symptoms and size of the lesion are the determinants for the choice of the treatment. In general, symptomatic or greater than 8 cm masses require intervention.

Due to its benign nature, renal AMLs represents an ideal lesion for nephron-sparing treatment by tumour enucleation, partial nephrectomy or selective arterial embolization (1,2). However, there are limited numbers data with regard to nephron sparing surgery of renal AMLs especially in bilateral cases with TSC. We report the treatment result of bilateral nephron-sparing surgery of a patient with giant bilateral renal AMLs and TSC.

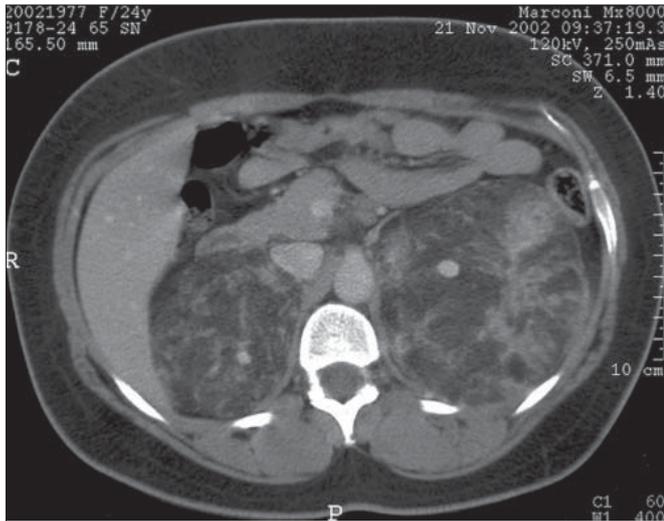
## Case report

A 25-year-old women was admitted to our clinic due to bilateral pain in costovertebral angle. On physical examination, she was found to have sebaceous adenomas. The patient's history was completely free of neurological symptoms, such as epilepsy or mental retardation. Renal sonography showed typical multiple hyperdense lesions in both kidneys. The abdominal computed tomography (CT) scan showed multiple tumour lesions in both kidneys with a varying content of fat, which was suggestive of renal AMLs (Figure 1). The multiple tu-

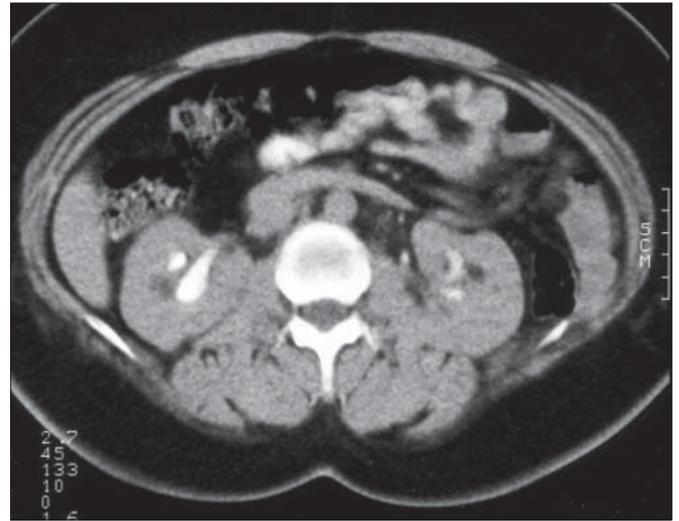
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**Figure 1.** Initial contrast-enhanced CT of the abdomen shows a faint angiomyolipoma in the left kidney with a large angiomyolipoma in the right kidney..



**Figure 2.** The postoperative CT after contrast dye administration of the abdomen demonstrates small foci of angiomyolipoma in bilateral kidneys and both kidneys are functional as well..

mours in the left kidney varied from 0.5 to 15 and in right kidney varied from 0.5 to 9 cm in maximum diameter. In cerebral CT, multiple bilateral calcifications indicated cortical tubers were revealed. Her creatinine at presentation was 0.9 mg/dl.

The patient subsequently underwent left renal nephron-sparing surgery by using extraperitoneal flank incision through 11<sup>th</sup> or 12<sup>th</sup> rib firstly. The largest resected lesion was 15x13 cm. A double-J stent was placed because of a defect that occurred in collecting system and it was removed in postoperative first month. Right nephron-sparing surgery was done in the same technique above mentioned 4 months after the first operation. The largest resected lesion was 9x7.5 cm. No stent was used in this operation. Renal lesions in the both renal units were resected with a 1 mm margin of normal parenchyma without renal arterial clamping. Smaller lesions (<5mm in maximum diameter) in the centre of the kidneys were not resected in order to preserve renal tissue. No significant intraoperative or postoperative complication was seen and blood transfusion was not necessary in these operations.

The final pathological examination confirmed AML. Postoperative creatinin was 1.1mg/dl and CT did not show contrast dye extravasations (Figure 2). Only a few small AMLs were seen in both kidneys and the renal functions were normal at the first year of follow-up.

## Discussion

Renal AML is a clonal neoplasm, apparently part of a family of neoplasms derived from perivascular epithelioid cells (3). It is a commonly benign unilateral (>80%) renal tumour that occurs predominantly in women between the

4<sup>th</sup> and 7<sup>th</sup> decade of life and most of them are asymptomatic (3). Tuberous sclerosis complex (TSC) is an autosomal dominant disorder transmitted by TSC1 and TSC2 genes characterised by seizures, mental retardation and hamartomatous lesions, including facial angiomyolipoma, subependymal giant cell astrocytoma, cardiac rhabdomyoma and renal angiomyolipoma (4). The hallmark lesion of the central nervous system is a superficial cortical hamartoma of the cerebrum, which sometimes looks like hardened gyri, creating the appearance of a tuber (root). Up to 40% to 80% of patients with TSC develop renal AMLs. Renal AMLs associated with TS most often occur at a younger age (mean age 17 y), develop bilaterally, with an approximately equal prevalence among male and female patients, are of larger size and these tumours are more frequently symptomatic than patients with sporadic disease (5).

Treatment is recommended for patients with pain, retroperitoneal haemorrhage, hematuria, hypertension, palpable mass and anaemia (1). It is suggested that renal AMLs that are less than 4 cm in diameter tend to be asymptomatic and generally do not require intervention and should be observed every 12 months with CT or US (6). These lesions are symptomatic in approximately 23% of cases. When renal AMLs are >4 cm in size, the incidence of symptoms increases to 82% and these patients have substantial risk of bleeding (7). AMLs that are more than 8 cm in diameter increase the risk of morbidity significantly. Therefore, it is suggested that renal AMLs greater than 8 cm and symptomatic renal AMLs of any size should be managed surgically to prevent associated complications such as rupture and haemorrhage. In addition, all suspicious renal lesions for malignancy have to be explored.

Because of the benign nature, the principles of management are resolution of symptoms and prevention of fatal morbidity without compromising renal function, except for rare cases of the malignant epithelial type (4). Selective arterial embolization and nephron-sparing surgery are renal preserving treatment modalities available for patients with these benign neoplasms. Selective arterial embolization is recommended for patients with solitary kidneys and/or haemorrhagic AML (2). But re-embolization or secondary surgery requires in about 25% of patients due to recurrence of symptoms or re-bleeding (4,8). In addition, a risk of unexpected large ischemic change should be kept in mind when treating a tumour with multiple feeding arteries (6).

Nephron-sparing surgery is preserves as much parenchyma as possible and, therefore, it should be performed whenever possible in these benign lesions. However, the studies related to NSS included only a small number of patients with TSC (3 of 55) (1,9). Heidenrich performed NSS in 28 patients 2 of whom had bilateral renal AML and they

reported excellent outcome in terms of symptom control and stable renal function (8). Tongaonkar et al. described 5 cases of bilateral angiomyolipoma and nephrectomy was performed in 4 cases and partial nephrectomy in only one case (9). The operative mortality of NSS is comparable to partial nephrectomy (10). Since these lesions are benign, the risk of residual microfocal disease has less long-term significance. Therefore, in cases of smaller, residual AML after surgery, follow up examinations are recommended in intervals of 3 months for the first 2 postoperative years to recognize changes in the size and morphology of AML, even if malignant transformation is extremely seldom.

Every effort should be made to preserve renal tissue to treat renal AMLs especially in bilateral cases. NSS may be performed safely with low morbidity, preservation of renal function, a low local recurrence rate and high patient satisfaction in patients with multiple and large bilateral angiomyolipomas. Renal function can be preserved, symptoms are controlled immediately and no secondary interventions for recurrence are necessary by NSS.

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