Bilateral Renal Angiomyolipoma

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Bilateral renal angiomyolipoma is a rare entity, usually associated with tuberous sclerosis. Five cases of bilateral renal angiomyolipoma, all females, with a mean age of 34.6 years, are reported. All patients had symptoms attributable to lesions only on one side, with flank pain and mass the commonest symptoms encountered. Only one case had clinical evidence of stigmata of tuberous sclerosis. The mean size of the lesions on the symptomatic side was 17 cm, while that on the asymptomatic side was 2 cm. Three patients were diagnosed correctly as having angiomyolipomas preoperatively with the use of ultrasonography and computed tomography (CT); two of these were treated with total nephrectomy and the third with partial nephrectomy. The other two cases were seen before the availability of CT and were only diagnosed intraoperatively. Both patients were treated with total nephrectomy. The lesions on the asymptomatic side were kept on close surveillance. Two patients developed an increase in the size of the lesions in the contralateral kidney 1 year and 3½ years after the first surgery. Both patients were treated with partial nephrectomy. All patients are alive at 3–11 years (mean 6.6 years) after operation.

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INTRODUCTION

Bilateral angiomyolipoma of the kidney is a rare entity, usually associated with tuberous sclerosis. Very few cases of bilateral renal angiomyolipoma without the stigmata of tuberous sclerosis have been reported. We herewith report five cases of bilateral renal angiomyolipoma of which only one had clinical evidence of tuberous sclerosis.

CLINICAL FEATURES

Twenty-three patients with histologically proved renal angiomyolipoma of whom five had bilateral lesions were seen at the Tata Memorial Hospital, Bombay, between 1968 and 1990. All five patients were females, ranging in age from 26 to 48 years (mean 34.6 years).

All patients had symptoms related to one side only, the other side being totally asymptomatic. The most commonly encountered symptoms were flank pain (five patients), flank mass (three patients), haematuria (one patient), and fever (two patients). One patient presented with severe haemorrhage leading to hypovolemic shock. Patients were examined and investigated to look specifically for the stigmata of tuberous sclerosis. However, only one patient had typical features of tuberous sclerosis associated with renal angiomyolipoma.

Four patients had symptomatic lesions situated in the left kidney, while the fifth had a right-sided symptomatic lesion. The size of the lesions on the symptomatic side varied from 10 to 27.5 cm (mean 17 cm). Four of these tumours occupied almost the whole kidney, while the fifth had a predominantly upper pole tumour. The size of the lesions on the asymptomatic side varied from 1 to 3 cm (mean 2 cm).

The diagnostic modalities used in these cases varied widely, as these cases spanned a 23-year period. Three cases were correctly diagnosed preoperatively with a combination of computed tomography (CT) and ultrasonography, while the remaining two patients were seen before the availability of CT scan and only underwent

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excretory urography and selective renal angiography. In the latter two cases, the definitive diagnosis of angiomyolipoma was obtained only intraoperatively.

Figure 1 shows the ultrasonographic appearance of the highly echogenic angiomyolipomatous lesions in the kidneys of one of the patients. Figure 2 demonstrates a large rounded low-density mass with some isodense areas within, arising from the lower pole of the right kidney, displaying distinctly negative attenuation values. Multiple small nodules of fat density are also seen in the remaining portion of the right kidney. The left kidney also shows a lobulated mass of 3 cm at the lower pole and a mass in the peripelvic region of about 2 cm displaying similar characteristics. Figure 3 shows the sagittal view of the CT scan of the same patient.

The two patients without a preoperative diagnosis of renal angiomyolipoma were treated with total nephrectomy and at operation were found to have angiomyolipomatous lesions of the contralateral kidneys. Of the three patients with a definitive diagnosis of angiomyolipoma preoperatively, two required total nephrectomy due to large and/or multiple lesion(s), which made a renal parenchyma-conserving surgery impractical. The third patient, however, had a partial (upper polar) nephrectomy for complete removal of the angiomyolipoma. The histopathological examination of the excised specimens of the kidney with the tumour revealed the typical features of a renal angiomyolipoma. Figure 4 shows the postoperative CT scan of the patient taken 3 years later showing stable lesions in the other kidney. All patients were followed up at regular intervals with clinical examination and ultrasonography to monitor the lesions in the contralateral kidney. Two patients developed an increase in the size of the lesions in the contralateral kidney 1 year and 4 years after the first surgery. Both patients were treated with partial nephrectomy of the contralateral kidney and are alive and disease free to date. The other three patients have been free of symptoms, attributable to the lesions in the other kidney; in none of them, the size of the lesions exceeded 3 cm on ultrasonography at their last follow-up. No new lesions have appeared in the contralateral kidneys of the patients who were treated primarily with partial nephrectomy. All five patients are alive, asymptomatic and with either no new lesions or stable lesions in the contralateral kidneys, between 3 and 14 years (mean 6.6 years) after operation.

DISCUSSION

Angiomyolipomas are uncommon tumours, comprising about 1–2% of all renal tumours [1]. They are benign hamartomatous lesions, having components of smooth
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There are clinically two distinct types of angiomyolipomas: those associated with tuberous sclerosis, and others that occur independent of tuberous sclerosis. Those associated with tuberous sclerosis are generally small, asymptomatic, multifocal, and bilateral and are usually found at autopsy. The other group without tuberous sclerosis is less common and is usually unilateral and focal but presents with large and symptomatic lesions. About 5–12% of lesions are bilateral, most of which are associated with tuberous sclerosis. Both types are, however, histologically indistinguishable from each other. The incidence of bilaterality in patients without tuberous sclerosis is rare [2–6]. However, we found that only one out of five patients with bilateral renal angiomyolipomas had evidence of tuberous sclerosis.

The commonest presenting symptoms of this tumour are usually a tender mass in the flank, pain that may be secondary to intrarenal and/or retroperitoneal haemorrhage and hematuria in some patients due to erosion of the pelvicalyceal system [5–7]. In our patients too, flank pain and mass were the commonest symptoms encountered. The tendency of the tumour to bleed is due to the high vascularity, structural rigidity, and inelasticity of blood vessels, which may require emergency treatment. Of the angiomyolipoma patients described in the literature, 25% presented with serious pain or shock because of hemorrhage and had to undergo emergency nephrectomy [8–11]. Only one of our patients, however, presented with shock due to serious hemorrhage. Renal angiomyolipomas have been reported to have a predilection for the female sex. Our experience was similar, since all five of our patients were females.

In the past, angiomyolipomas were difficult to diagnose preoperatively, although radiolucent areas on nephrotomograms that indicated the fatty component led to the suspicion of the lesion [12]. However, computed tomography (CT) is very useful for the preoperative diagnosis of angiomyolipoma, since the fat and nonfat interface in the tumour gives a characteristic appearance offering high accuracy for its diagnosis [13–16]. CT documents distinctly negative attenuation values, corresponding to the presence of fat within the tumour, indicating the presence of angiomyolipoma. Besides, identification of adipose tissue within an intraparenchymal renal lesion virtually excludes the possibility of carcinoma [16]. Ultrasonography, by demonstrating a highly echogenic well-defined mass within the renal cortex is as conclusive as CT [16–18]. The angiographic features are not specific.
and not of any diagnostic help but are useful in planning conservative surgery whenever indicated. Since the availability of the CT scan, we have been able to diagnose renal angiomyolipomas correctly preoperatively in all three cases, while before the CT scan era, we were unable to diagnose these lesions with confidence and differentiate them from renal cell carcinoma.

Indications for surgery in renal angiomyolipoma and its extent have been a matter of great debate. In the past, due to inability to differentiate angiomyolipomas from renal cell carcinoma the risk of massive bleeding, total nephrectomy was the primary treatment carried out. Before 1976, 93% of all reported angiomyolipomas not associated with tuberous sclerosis were treated with total nephrectomy [119]. Currently, with the correct diagnosis of angiomyolipoma with the newer imaging techniques, it is possible to plan either conservative surgery, selective arterial embolisation, or even surveillance if the lesions are small, multifocal, and bilateral [19–21]. Total nephrectomy may be justified only in the presence of uncontrollable life-threatening haemorrhage, when the entire kidney is involved or when there is a possibility of a coexistent renal cell carcinoma in the same kidney [22,23]. While treating bilateral angiomyolipomas, the decision to operate on a patient is influenced by the size of the lesions and the symptoms produced. One must be as conservative as possible in order to save maximal renal tissue and renal function. Small and asymptomatic lesions may be kept under surveillance, and large and/or symptomatic lesions should be surgically removed. Two of our five patients underwent total nephrectomy because of the lack of definitive diagnosis of angiomyolipoma preoperatively. Of the remaining three patients, two had very large masses occupying almost the entire kidney, making renal tissue-conserving surgery impractical. In only one patient was the lesion on the symptomatic side small enough to allow a conservative partial nephrectomy.

The lesions in the contralateral kidneys were being kept under observation, and although two of these five developed increase in the size of the lesions during the follow-up period, they could be managed successfully with conservative partial nephrectomy.

In conclusion, this is a report of five cases of bilateral renal angiomyolipoma in which a total or partial nephrectomy had to be done on the symptomatic side, while the other kidney was kept under close observation. Bilateral renal angiomyolipomas without evidence of tuberous
sclerosis have been reported to be rare but were a common finding in our study. Bilateral angiomyolipomas must be handled with great care; the treatment approach should be as conservative as possible to preserve maximum renal function. This can be better accomplished with a good CT scan imaging for a correct preoperative diagnosis of renal angiomyolipoma.

REFERENCES


Fig. 4. Postoperative CT scan of the patient who had undergone a right nephrectomy and has stable lesions in the contralateral kidney.


