CASE REPORT

Nephron-Sparing Surgery for Giant Angiomyolipoma of the Kidney

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Abstract

Renal angiomyolipomas are rare benign tumors known to occur sporadically and in association with genetic syndromes, tuberous sclerosis and lymphangioliomyomatosis. Surgical removal or radiographic embolisation of angiomyolipomas larger than 4 cm is usually indicated because of an increased risk of spontaneous hemorrhage. We describe successful nephron-sparing surgery for a giant angiomyolipoma and discuss relevant management issues in a patient with angiomyolipoma.

Key Words

Renal Angiomyolipoma, Nephron-sparing Surgery

Introduction

Angiomyolipoma is an uncommon benign tumor of the kidney (1) (constitute less than 3% of all renal neoplasms) .The tumor consists of smooth muscle, thick walled blood vessels and mature adipose tissue. The tumour can exist in isolation or it can be associated with tuberous sclerosis. In the first case renal AMLS are single, often asymptomatic, have female preponderance (male to female ratio of 1:4 to 1:6) and are typical of fifth and sixth decade of life. In the second case the renal AMLs are most often multiple, bilateral, symptomatic and without female or male preponderance (2).

We want to highlight the fact that being in a peripheral centre where there is no facility for angio-infarction or argon beam coagulator, we have successfully and safely performed nephron sparing surgery for a giant angiomyolipoma using basic instrumentation. Large size of the tumour alone should not be the criteria for performing nephrectomy and should be resorted to only if whole of kidney is replaced with tumour even in a peripheral centre.

Case Report

38yrs old female presented with H/O fever for 7-8 months which was intermittent and low grade. She had no urinary symptoms. Ultrasound abdomen picked up S.O.L arising from lower pole of left kidney which was highly echogenic measuring 16x12 cm. CT scan revealed large mass lesion arising from the lower pole of left kidney resulting in gross displacement of left kidney. Hounsfield density of -47 to -91 was recorded from the mass lesion suggesting the presence of fat (*Fig-1*). There was no evidence of calcification. CT findings were diagnostic of angiomyolipoma of the kidney. The patient did not have any features of tuberous sclerosis. As this tumor is considered benign a conservative nephron sparing surgery was planned.

On exploration an exophytic tumour was arising from lower pole left kidney. Although the tumor was large (16cm), most of the renal parenchyma was well preserved. After dissecting the tumour and the kidney, the renal vessels were clamped with bull dog clamps and the kidney was cooled with sterile ice slush. Tumor was excised with 1cm margin of normal renal tissue around it, without opening the pelvicalyceal system (*Fig-2*). Hemostasis was achieved and renal pedicle clamps were released after 19 minutes of clamping. Patient required no blood transfusion and made uneventful recovery. Histo-pathological examination confirmed the diagnosis of angiomyolipoma(*Fig-3*).

Patient is on follow-up for last one and half years. She is asymptomatic clinically and ultrasound examination done one year after the surgery does not reveal any tumour in either kidney.

Discussion

The term angiomyolipoma was first coined by Margum in 1951. Fischer(1911) and later on Critchley in (1932) found that 40-80% of cases of renal angiomyolipoma were associated with one or more features of tuberous sclerosis complex i.e. mental retardation , epilepsy, retinal phecomas, adenoma sebaceum, hemartoma of liver, heart or bone, cerebral angiomas and cerebral neoplasms. Our

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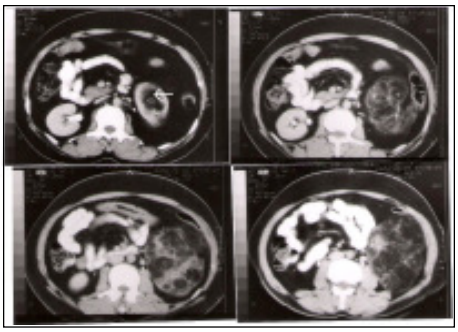

Fig 1. Contrast CT Showing Large Tumor Arising from Lower Pole of Left Kidney

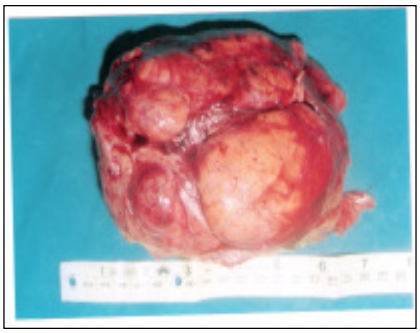

Fig 2. Excised Tumor Specimen

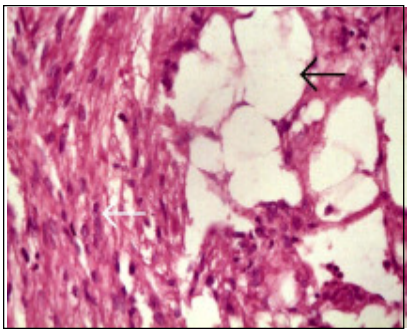

Fig 3. Histo-pathological of Tumor

case did not have any of the features mentioned above. Widespread use of ultrasound and C.T. abdomen has increased the incidental detection of isolated renal AMLs. These lesions often appear highly echogenic on ultrasound and have very low (below-30) attenuation values on C.T. scan which are most diagnostic (3).Renal angiomyolipoma with minimal fat does pose as a diagnostic dilemma. Double-echo gradient-echo(GRE) chemical shift magnetic resonance(MR) imaging can be used to differentiate AML with minimal fat from other renal neoplasms (4). Our case was diagnosed by abdominal ultrasound and CT abdomen done subsequently confirmed the diagnosis.

The most common clinical presentation is pain. The other modes of presentations include mass, hematuria, hypertension, U.T.I, and fever (5). Hemorrhage within the tumor or in the perinephric region is the most dreaded presentation and requires prompt management. Low grade fever of prolonged duration was the only mode of presentation in our case.

Management of AML is based on symptoms and size of the lesion. These tumors are considered benign hence conservative surgery preserving as much normal kidney as possible should be planned when ever possible (6). Angio- infarction of large lesions or those associated with hemorrhage can be done.

Small (<4cm) and asymptomatic tumor can be followed up conservatively by annual ultrasound examination. Large (>4cm) and symptomatic lesions require nephron sparing surgery where ever possible (7,8). Surgery may take the form of enucleation if the tumor is peripheral. Minervini A et al have reported excellent long-term local control of tumor with simple enucleation even for the larger lesions (9).Polar nephrectomy or partial nephrectomy may be the operation of choice for large polar lesions or multiple lesions limited to one pole. In other cases selective arterial embolisation in conjunction with operative intervention may be necessary (10).We performed polar nephrectomy in our case without prior selective arterial embolisation. However total nephrectomy is indicated only rarely when whole of the kidney is involved with tumor or in patients with uncontrollable life threatening hemorrhage.

Conclusion

The message we want to convey is that every effort should be made to conserve the normal renal parenchyma when the diagnosis of angiomyolipoma is established without doubt since it essentially is a benign condition. Even in a peripheral setup with minimal instrumentation, renal preservation surgery is possible if the surgeon is trained to perform such operation.

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