Case Report

A Giant Angiomyolipoma of the Kidney: A Case Report

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ABSTRACT

Angiomyolipoma (AML) is a rare benign renal tumor. Most frequently, it takes the form of small single tumor occurring sporadically. In some cases, the tumor may reach a very large size and cause serious complications. Herein, we present the case of a 25-year-old female patient, suffering from left loin pain, in whom a ruptured giant AML was diagnosed involving the left kidney and treated successfully with selective arterial embolization.

KEYWORDS: angiomyolipoma, embolization, renal tumor

INTRODUCTION

Renal Angiomyolipoma (AML) is a benign tumor, arising from mesenchymal elements of the kidney. It is four-fold more frequent in women and the peak of incidence falls between the 30th and 50th years of life[1]. It can occur sporadically, which is often solitary, and accounts for 80% of the tumors, or may be associated with tuberous sclerosis complex in 20% of cases. It has an incidence of 0.3 - 3% and is increased due to advancement in imaging modalities[2]. Histologically, it is composed of varying proportion of mature adipose tissue, smooth muscle and abnormal thick walled blood vessels[2].

CASE REPORT

A 25-year-old single Asian female patient presented to surgical casualty with recurrent left loin pain since five days and one day fever with no associated lower urinary tract symptoms or hematuria. She has no history of chronic disease. Clinically temperature was 38 °C with tenderness and fullness of left renal angle. Her Hb was 9.3 gm/dl, normal renal function tests. Abdominal ultrasound revealed well defined hyperechoic lesion with central hypoechogenicity in the lower pole of left kidney with a large eccentric component measuring about 12.5 x 8.5 cm with no increased vascularity on Doppler study (Fig 1a). Triphasic computed tomography (CT) of abdomen and pelvis reported large soft tissue mass lesion arising from lower pole of left kidney 12.5 x 10.5 x 8.5 cm in dimensions, with heterogeneous texture but mainly of fat density, moderately enhanced after contrast administration with moderate to marked stranding of perinephric fat planes coupled with mild perinephric hematoma (Fig 1b). The findings were suggestive of ruptured large AML. Additionally noted multiple small left renal cortical lesions (almost only fat CT density) largest is midzonal eccentric and measures 10 mm features of other smaller left AML. DMSA-Renogram study reported that left kidney contributes to 48% of total renal function with no cortical defect. Patient was initially managed conservatively with bed rest, intravenous injection of a broad spectrum cephalosporin antibiotics (cefotaxime sodium, 1 grm every eight hours for five days), IV fluids and blood transfusion. Since there was a suspicion of severe bleeding from a huge renal angiomyolipoma, we prepared the patient for open left nephrectomy, but she became clinically stable and she refused this planned surgical intervention. We consulted our interventional radiologist for selective arterial embolization of this renal mass, initial renal arteriography showed tortuous, hypervascular, and aneurysm-forming angiogenic components without extravasation of the contrast agent (Fig 2). Immediate complete obliteration with metallic coil was technically successful and her hemodynamic status was stabilized a week later without any complication in the post-operative period (Fig 2).
was discharged symptomatically for follow up in our outpatient clinic. Serial follow up with nuclear study of the renal function, Ultrasound and CT (Abdomen, Pelvis) after 3 and 6 months showed stable renal function and significant reduction of the size of the renal AML. One and half years later, the ultrasound study showed only single 5 mm left renal AML and disappearance of the above mentioned renal mass (Fig 3). The patient is still under our care in out-patient clinic with a very good general condition.

DISCUSSION

Angiomyolipoma (AML) is a rare benign hematoma renal tumor that contains various proportions of
smooth muscle, adipose tissue and blood vessels. Most frequently these tumors are unilateral, small and singular[3]. In the remaining case, they coexist with tuberous sclerosis and they may achieve significant size and could be multiple and bilateral[3]. The diagnosis of angiomyolipoma, which is most often found incidentally, depends on CT scan imaging which helps in identifying fat in the renal lesion[4]. Also these lesions need to be differentiated from malignant renal tumors which often contain areas of calcification, whereas calcifications are rare in the benign angiomyolipoma[5]. Annual follow up with CT scan or ultrasound can be performed to assess disease stability versus progression[6].

Renal AML usually grows slowly; therefore, depending also on the location of these lesions, the patients with smaller renal AML have no apparent clinical symptoms[7]. Asymptomatic and small lesions less than 4 cm often do not require any interventions[7]. When a renal AML enlarges, it usually becomes more vascular, with tortuous vessels; some of them have insufficient elastic layer and muscle wall, leading to the formation of aneurysms that are prone to spontaneous rupture, resulting in hemorrhage[8].

Patients may have sudden lower back pain, resulting in dramatically increased tumor volume. When the tumor growth reaches the renal capsule, the patients may have an acute abdomen or hemorrhagic shock caused by retroperitoneal hemorrhage, which may cause life-threatening hypovolemic shock that requires nephrectomy or nephron-sparing surgery[8]. It is generally agreed that AML which is bigger than 10 cm and affects a whole organ is considered a ‘giant’ AML and asymptomatic lesions more than 8 cm should be treated similarly to those with symptomatic lesions more than 4 cm[9].

The therapeutic options include surgery (partial or total nephrectomy) and selective embolization of these lesions which are clinically important due to their propensity to bleed[10]. The improvements and increasing skills of techniques of selective transcatheter arterial embolization led it to being widely used to treat acute hemorrhage caused by ruptured renal AML either alone or in conjunction with surgical intervention[11]. The advantages of this method include the preservation of functional renal parenchyma, and avoiding the need of anesthesia. Various materials have been used, including pure alcohol, absorbable gelatin sponged, polyvinyl alcohol particles and metal coils[12]. Complications of post embolization syndrome, which is widely thought to be an inflammatory response to necrotic tissue after embolization, can include fever and flank pain[13,14].

In our case, the patient was a young lady, presented with acute retroperitoneal bleeding, due to a ruptured huge renal angiomyolipoma. As the patient became dramatically hemodynamicaly stable only with blood transfusion and parenteral antibiotics, and she had refused any surgical intervention including nephrectomy, we advised her to do selective arterial embolization of the AML to prevent the possible spontaneous rupture in future and its consequences, especially hemorrhage. When she agreed, the above mentioned technique was performed successfully using a metal clip. The patient was almost asymptomatic in the post embolization period, and subsequent follow up with imaging showed stabilization of the disease and gradual decrease in the size of the lesion with only a small non-significant residual AML with preservation of the function of the involved kidney. Regarding the management of such cases of huge renal angiomyolipoma, and in the light of the previous published studies in literature, we found the technique of selective arterial embolization of a giant AML as a safe, attractive, minimally invasive and cost-effective method, which may help stabilization and treatment of these tumors. As we have a significant number of relatively small sized renal AML cases, which was managed conservatively, we will try in the future, when it’s possible to collect more cases of huge AML, which may need this technique that help us to establish such management in our department.

CONCLUSION

Renal angiomyolipoma is a tumor with a generally benign nature, which can be managed conservatively. Sometimes the giant sized type of this tumor may take a complicated course with spontaneous rupture and subsequent bleeding, that can be managed with selective transarterial embolization, which we found to be a well-tolerated procedure and is associated with minimal adverse reaction to preserve the function of the involved kidney. However further studies with more cases are needed to establish such a management.

REFERENCES


