Bilateral Angiomyolipama of the kidneys with shock due to rupture: Can conservative management be an option?

Alemayehu Tegene, Shemssie Shewmollo Bushira and Etenesh Tewolde Tesfahunegn

Saint Paul's Teaching Referral Hospital, Addis Ababa, Ethiopia.

*Corresponding author. E-mail: alett2005@gmail.com.

ABSTRACT

Renal angiomyolipoma is a benign tumor of the kidney which is composed of fat, smooth muscle and thick walled vessels in varying proportions. It is the most common cause of spontaneous renal hemorrhage often present with classic triads of abdominal pain, palpable mass and hypovolemic shock called Wunderlich syndrome. Once the diagnosis of ruptured angiomyolipoma with unstable hemodynamic state is made, the treatment recommendation is either surgery or therapeutic embolization. The case we report is a 20 year old female presented with classic triads of abdominal pain, giant palpable mass and hypovolemic shock who was successfully managed conservatively.

Keywords: Renal angiomyolipoma, AMLs, CT scan, tumor of the kidney.

INTRODUCTION

Renal angiomyolipoma is a benign tumor of the kidney which is composed of fat, smooth muscle and thick walled vessels in varying proportions. It is a rare parenchymal renal mass that accounts for <0.5% of all renal tumors (Siroky et al., 2004). Angiomyolipoma is the most common renal tumor associated with spontaneous hemorrhage, followed by RCC (Campbell-Walsh urology, 2012).

The risk of hemorrhage is related to the size of the tumor and is significantly higher in lesions greater than 4 cm in diameter (Grainger and Allison's Diagnostic Radiology, 2008).

About 80% of AMLs are sporadic and 20% are associated with tuberous sclerosis. Those cases associated with tuberous sclerosis are characterized by young age at onset, multiplicity and laterality and symptomatic in nature (Murat et al., 2006). They occur spontaneously in the general population and affect mainly women. In patients with tuberous sclerosis they occur at a much younger age and are frequently multiple, with an incidence of 50 to 80% (Grainger and Allison’s Diagnostic Radiology, 2008).

Angiomyolipoma is the only benign renal tumor confidently diagnosed on cross-sectional imaging by the presence of fat on non-enhanced thin-cut CT. Fat-poor angiomyolipoma may still be confused with RCC but if it is suspected preoperatively then percutaneous biopsy is eminently capable of providing the diagnosis with positivity for HMB-45 typically seen with angiomyolipoma and the treatment of choice in cases of acute hemorrhage is selective renal angioembolization (Campbell-Walsh urology, 2012).

CT is currently the most accurate means of diagnosing AMLs. The large amount of fat within an AML produces areas of low radiographic density characterized by a negative CT attenuation coefficient as measured in Hounsfield units. A Hounsfield unit value of <10 is considered diagnostic of AML (Grainger and Allison’s Diagnostic Radiology, 2008). The benign nature of AML makes it an ideal lesion for nephron sparing treatment by tumor enucleation, partial nephrectomy or selective arterial embolization.

However, there are limited numbers of data with regard to nephron sparing surgery of renal AMLs especially in bilateral cases with TSC (Murat et al., 2006). Here, we reported the outcome of conservative treatment of a patient with giant bilateral renal AMLs with rupture and presented as hypovolemic shock.

CASE REPORT

The case is a 20 year old Ethiopian female admitted to our urologic ward with hypovolemic shock due to giant ruptured AML after having trivial trauma to her abdomen.
during a fight with her husband. She was punched by her husband over her abdomen following which she started to have severe abdominal pain and lost consciousness for about 5 h. In addition, she also has progressive abdominal distension, severe fatigability and decreased urine output. On Physical examination she was hypotensive with blood pressure of 80/30 mm Hg. Pulse rate of 126 beats per minute which was feeble, respiratory rate of 24 breaths per minute, temperature of 36.5°C and arterial saturation by pulse oximetry of 79% without oxygen support. Her conjunctiva was severely pale and on the abdomen there was a 14 × 16 cm ill-defined mass arising from the left flank and subcostal area with mild tenderness over it. Chest examination showed bilateral lower 3rd lung field dullness with absent air entry. There were skin lesions in the malar, nasolabial and back region. Other systems were unremarkable. With the assessment of hypovolemic shock due to massive retroperitoneal hemorrhage from ruptured bilateral renal angiomyolipoma, the patient was resuscitated with 4 L of normal saline and transfused with seven units of whole blood while simultaneously being investigated with laboratory, ultrasound and computed tomography of the abdomen. The diagnosis of bilateral angiomyolipoma of the kidneys with rupture on the left side was made by the typical hyper echoic ultrasound and hypodense with negative 10 HU on non-contrast abdominal CT (Figures 1 to 6).

Analgesics, antiemetic, antibiotics and potassium supplementation was also started and progress monitored with clinical, laboratory (mainly serial hemoglobin,
Figure 3: A well-defined hyperechoic lesion on left renal mid pole region.

Figure 4: Longitudinal ultrasound of the left kidney: There is an ill-defined heterogeneous echogenic lesion on left kidney which measured 6 cm × 7 cm which suggests hematoma collection.

Table 1: The laboratory monitoring.

<table>
<thead>
<tr>
<th>Date</th>
<th>WBC</th>
<th>HCT (%)</th>
<th>Hgb</th>
<th>Platelet</th>
<th>BUN</th>
<th>Creatinine</th>
<th>Na+</th>
<th>K+</th>
<th>Ca++</th>
<th>PT</th>
<th>PTT</th>
<th>INR</th>
</tr>
</thead>
<tbody>
<tr>
<td>17/03/17</td>
<td>4700</td>
<td>9.5</td>
<td>2.6</td>
<td>19000</td>
<td>104</td>
<td>1.9</td>
<td>146</td>
<td>4.6</td>
<td>1.01</td>
<td>18.4</td>
<td>28.4</td>
<td>1.57</td>
</tr>
<tr>
<td>18/03/17</td>
<td>16200</td>
<td>19</td>
<td>7.3</td>
<td>42000</td>
<td>86</td>
<td>2.3</td>
<td>170</td>
<td>4.36</td>
<td>2.29</td>
<td>18.9</td>
<td>28.4</td>
<td>1.5</td>
</tr>
<tr>
<td>20/03/17</td>
<td>1200</td>
<td>21.1</td>
<td>6.5</td>
<td>55000</td>
<td>98.8</td>
<td>2.25</td>
<td>150</td>
<td>4.91</td>
<td>2.97</td>
<td>17.6</td>
<td>27.8</td>
<td>1.53</td>
</tr>
<tr>
<td>21/03/17</td>
<td>13300</td>
<td>19.4</td>
<td>5.9</td>
<td>88000</td>
<td>60.5</td>
<td>1.78</td>
<td>146</td>
<td>3.66</td>
<td>2.13</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>22/03/17</td>
<td>1200</td>
<td>18.1</td>
<td>8.1</td>
<td>127000</td>
<td>58.1</td>
<td>1.65</td>
<td>138</td>
<td>3.7</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>23/03/17</td>
<td>10700</td>
<td>24.5</td>
<td>8.2</td>
<td>250000</td>
<td>45.4</td>
<td>1.34</td>
<td>145</td>
<td>3.9</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>24/03/17</td>
<td>12500</td>
<td>24.3</td>
<td>8.5</td>
<td>324000</td>
<td>32</td>
<td>1.3</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>25/03/17</td>
<td>14700</td>
<td>26.1</td>
<td>9</td>
<td>808000</td>
<td>104</td>
<td>1.9</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>30/03/17</td>
<td>7900</td>
<td>28</td>
<td>9.5</td>
<td>406000</td>
<td>88</td>
<td>1.57</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4/03/17</td>
<td>11000</td>
<td>29.5</td>
<td>9.7</td>
<td>350000</td>
<td>92</td>
<td>1.1</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>18/03/17</td>
<td>12000</td>
<td>31</td>
<td>10.1</td>
<td>450000</td>
<td>87</td>
<td>1.3</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Creatinine, liver enzymes), abdominal ultrasound and CT-abdomen. Initially, operative intervention was considered but her Creatinine was elevated even after normalization of her vital signs with fluid and blood, also the lesion was found to be bilateral and angioembolization service was not available and transplantation is at its infancy, hence, aggressive monitoring and conservative management was pursued and on the next day the vital signs became normal. Table 1 shows her progress in terms of laboratory and radiological improvements. Figures 1 to 6 shows the
radiologic findings. Both ultrasound and CT abdomen showed findings consistent with bilateral renal angiomyolipoma with left side rupture.

DISCUSSION

Renal angiomyolipomas (AMLs) are benign neoplasms composed of mature adipose tissue, thick walled blood vessels and smooth muscle in varying proportions (Bennington and Beckwith, 1975). These tumors account for 0.3 to 3% of renal masses and 1% of surgically resected tumors (Kothary et al., 2005). The chance of malignant transformation is exceedingly rare. Tuberous sclerosis–associated angiomyolipoma tends to be larger, more numerous and more likely to cause spontaneous hemorrhage than the sporadic disease. Tumors that bleed also tend to be larger. CT or magnetic resonance imaging is usually sufficient for diagnosis and biopsy is rarely needed.

The primary indications for intervention include symptoms such as pain or bleeding whereas prophylactic intervention is warranted for large tumors (Tso et al., 2005). Flank pain, hematuria or palpable mass may result from its bleeding or large size. Spontaneous renal bleeding secondary to an AML usually occurs when the tumor is larger than 4 cm (Oesterling et al., 1986). Spontaneously, hemorrhagic pattern-II renal AMLs must be differentiated from a RCC or other vascular entities (Zhang et al., 2002). For this reason, a careful search must be done during CT evaluation in order to detect fat (Bosniak et al., 1988), which in our series was invariably found (Figure 5). These lesions should be distinguished from well differentiated, low grade retroperitoneal or capsular liposarcoma and the very rare RCC encasing perirenal fat. As AML is a vascular tumor, it can be distinguished from a perirenal liposarcoma on CT scans by the presence of typical internal tortuous aneurysmal vessels and a renal parenchyma defects (Figure 6); findings usually not seen
in liposarcomas (Israel et al., 2002). Although isolated cases of calcified and non-calcified RCC containing fat was described, for an evidence-based practice, all renal mass with detectable fat should be considered an AML (Adilson, 2003). The diagnostic criterion for TSC was established in 1998 at the Tuberous Sclerosis Complex Consensus Conference in Annapolis, Maryland (Roach et al., 2004). The genetic tests may be useful to confirm the clinical diagnosis, but usually the molecular findings are not required.

Clinical symptoms of TSC include: epilepsy, mental retardation, developmental delay, ungual and periungual fibromas, shagreen patches and hypopigmented macules of the skin, angiofibromas of the face, AMLs and cyst in kidney, cortical tubers subependymal nodules and subependymal giant cell astrocytoma in the brain, cardiac rhabdomyomas, retinal hamartomas, hyperinflated lungs and enamel pits (Andrzej, 2008). Our patient presented with majority of the reported TSC symptoms and diagnosis of tuberous sclerosis was made based on clinical grounds.

TSC-associated AMLs tended to be more symptomatic, multiple and bilateral. Therefore, active surveillance is strongly recommended for asymptomatic tumors < 4 cm, while angioembolization or surgical interventions with maximal parenchymal preservation including partial nephrectomy, enucleation, or wedge resection should be an alternative option for symptomatic tumors ≥4 cm. In their experience of 4 TSC-associated patients with tumors ≥8 cm who finally ended up with receiving a radical nephrectomy, Kyo et al. (2010) demonstrated that, although angioembolization could be a primary option in controlling existing or impending hemorrhages in spontaneous ruptured AMLs, a prophylactic radical nephrectomy is relatively recommended for tumors ≥8 cm or those refractory to angioembolization.

Although surgical intervention in the form of nephron sparing surgery or radical nephrectomy was the mainstay in the management of most renal tumors, a study by Kyo et al. (2010) showed a surging number of angioembolization in patients with TSC-association. Angioembolization and nephron sparing surgery for renal AMLs both offer long-term renal function preservation with low complication rates. However, characteristics of TSC-associated AMLs such as large size and multiplicity have limited role of nephron sparing surgery and are one of the reasons for increased number of angioembolization for this group of patients. Although embolization and surgical therapies can successfully treat solitary lesions, the much more frustrating clinical problem of coalescent renal angiomyolipomas that replace renal parenchyma has remained largely unaddressed (Figure 5). When bleeding occurs in this circumstance, it can be impossible to identify which lesion is the source (John et al., 2004). The described patient presented with large, multiple bilateral AML with shock and deranged renal function made surgical intervention in the form of nephron sparing and radical surgery impossible (Figure 7). With the absence of angioembolization service (not best for this kind of lesions as earlier pointed out), conservative management in the form of transfusion, fluid resuscitation and aggressive monitoring of clinical, laboratory and serial abdominal ultrasound, the patient was stabilized. She stayed in the

**Figure 7:** Photo of the patient on discharge. Consent was obtained from the patient.
ward for 28 days and discharged with advice on possibility of re-rupture and she is her 3rd month of follow up with Creatinine in the last visit of 1.3 mg/dl. The plan is to continue frequent follow up.

REFERENCES


