Renal Angiomyolipoma Presenting as Acute Abdominal Emergency: A Case Report

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This Paper was Presented at the Nigerian Urological Association Conference 1998.

ABSTRACT

A 47-year-old woman presented as an acute abdominal emergency with rapid progression to shock following spontaneous rupture of a left renal angiomyolipoma. The diagnosis was missed clinically and on computerized tomographic scan. At different stages of management based on her pattern of symptoms, signs and radiologic appearances of her lesion, she was thought to have ruptured ectopic pregnancy, torsion of a left ovarian cyst, acute salpingitis, spontaneous rupture of the spleen, leaking abdominal aortic aneurysm and eventually left renal cell carcinoma until the histology of her left nephroureterectomy specimen became available. Even though a rare entity, with a high index of suspicion and a good knowledge of the pathognomonic features on ultrasound and computerized tomographic scanning, the diagnosis should be more easily made to facilitate quicker decisions on management.

KEY WORDS: Renal Angiomyolipoma, Acute Abdomen

Introduction

Renal angiomyolipomas are rare benign tumors of the kidney. [1] They are more commonly associated with tuberous sclerosis. [2] When discovered during routine investigations, they are followed up and elective decisions taken on their management. However, when they do present clinically, it is usually as an acute abdominal emergency often with rapid progression to shock. [3]

Case Report

A 47-year-old woman presented at the accident and emergency department of

the University College Hospital, Ibadan with a 24hour history of severe aching left upper quadrant abdominal pains radiating to the back associated with nausea and 4 episodes of vomiting. Her last menstrual period was 45 days prior to presentation

On examination, she was in severe pains and was moderately pale. Her pulse rate was 88/min; her blood pressure was 110/70 mmHg. Her abdomen was full, moved minimally with respiration, with guarding, marked tenderness and positive rebound tenderness in the left half. The presence or otherwise of organomegally could not be ascertained because of abdominal tenderness and guarding. She had positive cervical excitation tenderness on the left side on vaginal examination. A provisional diagnosis of ruptured left ectopic pregnancy was made. The gynecologists reviewed her and added a possible differential diagnosis of torsion of a left ovarian cyst.

While awaiting exploratory laparotomy, she went into shock with a systolic blood pressure of 50mmHg. Her packed cell volume was 30%. She underwent emergency exploratory laparotomy via an infra umbilical midline incision under general anesthesia by the gynecologists for a possible ruptured ectopic pregnancy. Intraoperatively, the uterus and both fallopian tubes and the ovaries were normal. A huge left-sided broad ligament and retroperitoneal hematoma was observed. The general surgeons were invited. At this time, before the general surgeons arrived, the gynecologists had explored the left broad ligament hematoma creating the impression of intraperitoneal bleeding. The general surgeons therefore entertained the possibility of spontaneous rupture of the spleen and extended the abdominal incision to a cape to coast incision. The spleen was found to be normal. Further exploration revealed a very extensive left sided retroperitoneal hematoma, extending from the diaphragm above to the pelvis below and across the midline to the right side along the transverse mesocolon. The aspects of the hematoma in the anticipated location of the left kidney was cystic, suggesting the possibility of a ruptured left renal mass, but the hematoma seemed otherwise pulsatile suggesting the possibility of a leaking abdominal aortic aneurysm. The urologists and the cardiothoracic teams were invited at this stage. At this point too, the patient had become very pale. She had no blood left in the blood bank, and no possibility of more blood being crossmatched for her that night. Her blood pressure was beginning to fall. It was therefore decided to close the rent in the retroperitoneum at this stage and all further explorations suspended, and the abdomen closed. In the recovery room, she was transfused with 3 units of fresh blood on the first postoperative day, and an abdominal ultrasonography, intravenous urogram and a CT scan were ordered. The emergency intravenous urogram and abdominal ultrasound scan was not very useful, but the CT scan showed a ruptured huge left renal mass, extensive retroperitoneal hematoma and a normal right kidney (Figures 1 and). It also showed some features to suggest a ruptured abdominal aortic aneurysm, which included indistinct outline of the abdominal aorta, anterior displacement of the left kidney, high-density collection

in the paracolic areas and obscuration of the psoas muscle outline. With 6 more units of blood available 24 hours after her initial operation, she underwent a left nephroureterectomy for a presumed diagnosis of a left renal cell carcinoma. She was transfused with a total of 8 units of blood.

Figure 1: Plain Computerized Tomographic Scan Showing a Huge Left Renal Mass and Retroperitoneal Hematoma

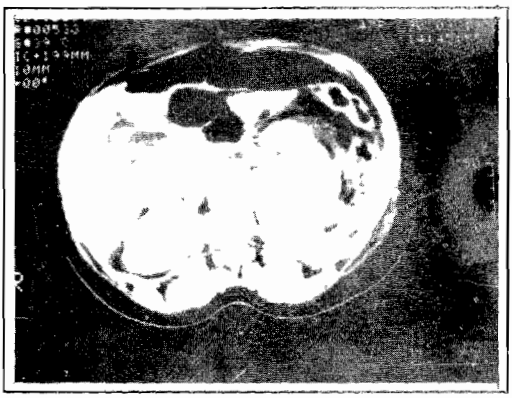
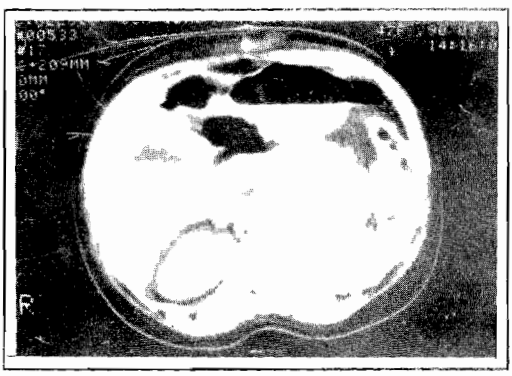


Figure 2: Contrast enhanced CT Scan, Showing the ruptured left renal mass



Postoperative recovery was uneventful. The histology of the renal mass came back as angiomyolipoma. With the benefit of hindsight afforded by the histology, she was reviewed during her follow up visit but there were no features to suggest tuberous sclerosis.

Discussion

Renal angiomyolipoma grossly [4] and radiologically [5] resemble adenocarcinoma of the kidney. As in this patient, most occur in women between the ages of 40 and 60 years. [1] The diagnosis is most often incidental during abdominal ultrasonography or computerized tomography for other unrelated problems. [6, 7] Initial symptoms are often due to sudden spontaneous rupture with perirenal hemorrhage, the patient rapidly progressing to shock [3] as in the patient presented. Angiomyolipomas are benign lesions and rarely metastasize but some may be locally aggressive as in our patient, displacing contiguous structures and parasitising local vasculature. [8] Angiomyolipoma of the kidney may occur sporadically as in our patient with no features of tuberous sclerosis on follow-up review but are much more prevalent in patients with tuberous sclerosis. [2]

A high index of suspicion is required to reach the diagnosis. The computerized tomographic and nuclear magnetic resonance imaging appearances are virtually pathognomonic, showing well circumscribed fat density mass or masses partially or totally within the renal substance. [5] In this patient, because of the sequence of

her clinical symptoms and signs, her differential diagnoses included ruptured ectopic pregnancy, acute salpingitis, torsion of left ovarian cyst, spontaneous splenic rupture, leaking abdominal aortic aneurysm and ruptured left renal call carcinoma. Predominant muscle, vascular components or intramural hemorrhage as in our patient make diagnosis of renal angiomyolipoma difficult and may explain why the diagnosis was missed in our patient on computerized tomographic scan. Consequently, our patient underwent a left nephroureterectomy on the presumed diagnosis of ruptured left renal cell carcinoma. However, in patients in whom a preoperative diagnosis is available, they are followed up and elective decisions taken on their management. When an intervention is indicated, it is aimed at renal parenchymal conservation, which can be accomplished through limited surgery or preferably by selective embolisation. [3, 6]

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