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Case Report

Giant Renal Artery Pseudoaneurysm Caused by Rupture of Renal Angiomyolipoma Following Pregnancy: Endovascular Treatment and Review of the Literature

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ABSTRACT. Renal angiomyolipoma is a hamartomatous, benign tumor composed of blood vessels, fatty tissue and smooth muscle cells, and is often detected incidentally. It can also be associated with the tuberous-sclerosis complex (TSC). Pregnancy and use of oral contraceptives are known to be associated with an increased risk of tumoral rupture and bleeding. Herein, we report a unique case of renal angiomyolipoma associated with TSC who presented with hypovolemic shock as a result of spontaneous rupture of a giant renal pseudoaneurysm, immediately after pregnancy. Emergency endovascular treatment was successful with sparing of most of the affected kidney as demonstrated by follow-up computed tomography imaging.

Introduction

Renal angiomyolipoma (AML) is a benign neo-plasm composed of thick blood vessels, fatty tissue and smooth muscle cells.[1,2] Most cases of renal AML present as an isolated lesion while approximately 20% of patients have an association with the tuberous-sclerosis complex (TSC) and other phacomatoses such as neurofibromatosis.[1,3,4] Renal AMLs associated with TSC tend to be multifocal, bilateral and larger. These lesions are more prone to rupture and tend to be symptomatic rather than the isolated lesions.[4,5] Renal AMLs are hormone-sensitive tumors.[4-7] Moreover, pregnancy induces tumor growth and rupture.[8] In this report, we report a case of renal AML associated with TSC who was admitted to the hospital for spontaneous rupture of a giant pseudoaneurysm, which had occurred as a complication of AML, following pregnancy. The patient was successfully managed with emergency endovascular embolization.

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Case Report

A 25-year-old puerperal female patient was admitted to an outside hospital with a sudden onset of abdominal pain, dizziness and fatigue. She had delivered a baby two weeks earlier. She was hypotensive (around 70/40 mm Hg) and had tachycardia with a heart rate over 120/min.

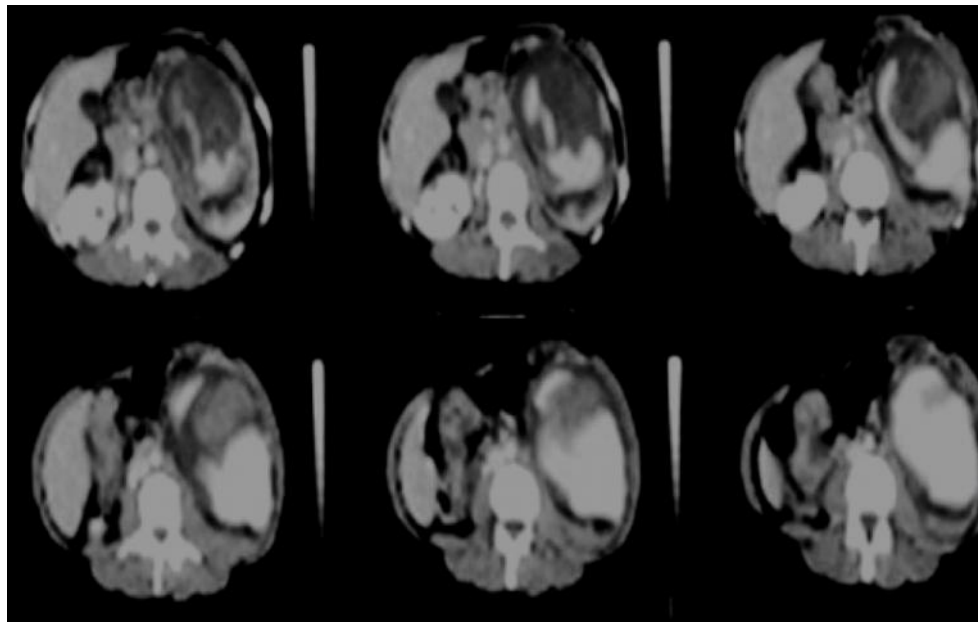

Figure 1. Abdominal computed tomography images obtained at an outside hospital showing giant pseudo-aneurysm originating from the left kidney. Multiple hypodense lesions are also seen suggestive of renal angiomyolipomas.

A diagnosis of hypovolemic shock due to blood loss was made on the basis of a drop in hemoglobin level to 8 g/dL from 14 g/dL two to three weeks earlier. Abdominal ultrasonography (US) revealed bilateral renal lesions suspicious of AMLs and a giant cystic mass originating from the lower pole of the left kidney. Subsequent computed tomography (CT) of the abdomen revealed a giant pseudoaneurysm originating from the lower pole of the left kidney with a diameter of 12 cm × 15 cm (Figure 1). The patient was immediately referred to our hospital for possible endovascular treatment.

In our hospital, the patient was taken to the angiography unit and abdominal aorta and selective renal artery injections were obtained. Abdominal aortography demonstrated contrast extravasation from the left segmental renal artery (Figure 2). Subsequent selective left renal artery injections confirmed the diagnosis of giant pseu-doaneurysm arising from the segmental branch of the renal artery (Figure 3). Using a micro-catheter, this segmental artery was embolized with 33% diluted n-butyl cyanoacrylate (nBCA), avoiding filling the sac itself (Figure 4). Follo-wing this, control angiography documented no further filling of the pseudoaneurysm sac and approximately 90% of the left renal parenchyma was preserved (Figure 5). Six weeks after the procedure, follow-up abdominal CT showed thrombosed pseudoaneurysm with no evidence of

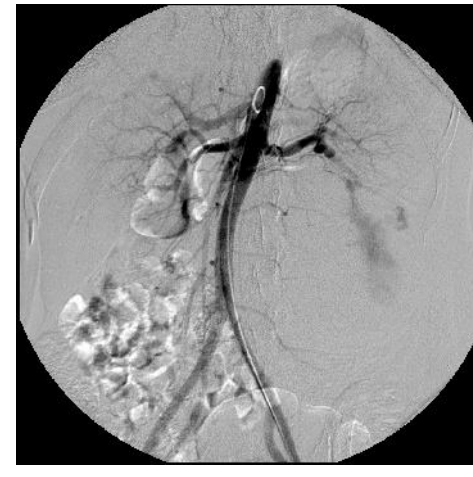

Figure 2. Abdominal aortogram showing filling of the giant pseudoaneurysm located in the left lower quadrant of the abdomen. Also, note the intestinal gas displacement to the right lower quadrant.

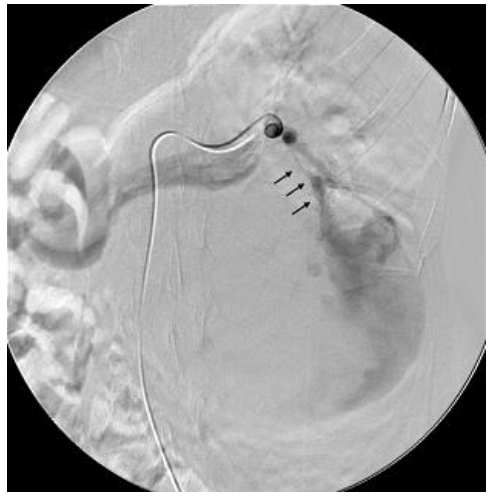

Figure 3. Selective injection into the left renal artery showing the segmental renal artery branch directly opening into the giant pseudoaneurysm (arrows).

active bleeding. Also, the diameter of the sac had decreased from 15 cm to 10 cm (Figure 6). The patient is on regular follow-up and is doing very well.

Discussion

Renal AMLs are generally benign and clinically silent lesions and diagnosed incidentally on US or CT examination performed for other clinical reasons.[9] However, they also tend to grow and be complicated with aneurysm formation and rupture,[10] which may cause life-threatening hemorrhage. In symptomatic patients, the classic triad of renal AML is flank pain, a palpable tender mass and hematuria due to intra-capsular or retroperitoneal bleeding.[11] There was no retro-peritoneal bleeding in our case as documented by CT imaging prior to the endovascular treatment. Although

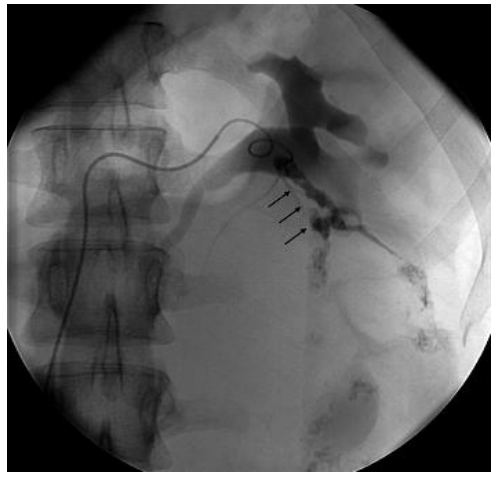

Figure 4. Scout film after nBCA embolization demonstrating the glue cast filling the segmental artery (arrows).

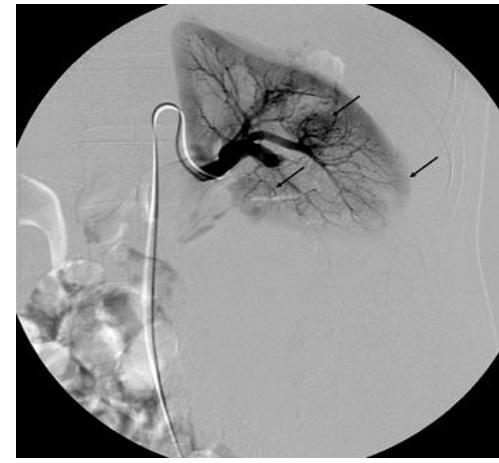

Figure 5. Selective left renal artery injection after endovascular treatment demonstrating the mostly preserved left kidney with other smaller AMLs (arrows).

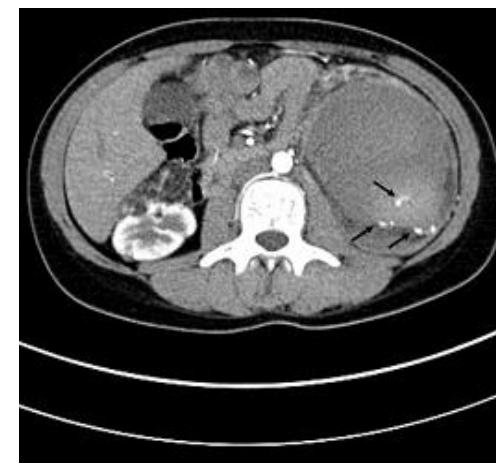

Figure 6. Six-week follow-up computed tomography image showing significant decrease in diameter of the totally thrombosed pseudoaneurysm in the left kidney. Also, note some hyperdense glue foci (arrows) within the aneurysm sac representing distal emboli during proximal parent artery embolization.

the patient did not have any baseline CT imaging prior to the bleeding, we believe that the rupture was totally intra-tumoral, which led to the formation of a pseudoaneurysm. CT imaging provides a more definitive and accurate assessment of the exact site and extent of hemorrhage in these cases.[12]

The risk of rupture of a renal AML increases with tumor size, presence of symptoms and pre-sence of TSC.[10,13] Several authors agree that lesions with a diameter of 4 cm or more have a high risk of hemorrhage.[1,13-15] The treatment of asymptomatic lesions larger than 4 cm is debatable, and some authors have suggested that they need not be treated.[14] However, some other authors have suggested that AMLs larger than 4 cm should be treated even if patients are asymptomatic; in symptomatic patients, AML of any size needs treatment.[16,17] There are two different options for treatment of renal AML. One of them is the classical surgical approach with a total or partial nephrectomy and the other is selective embolization.[5,18] Surgery should be reserved as a last measure, particularly in patients with multiple renal AMLs in both kidneys (e.g., TSC). Today, prophylactic embolization is recommended in high-risk patients including not only tumors large in size but also multiple lesions, ones associated with TSC and those seen in younger women who desire having children.[9]

The patient in this report was completely asymptomatic until the time of pregnancy. Sebaceous adenomas on her face did not draw any attention until the diagnosis of bilateral renal masses was made. Therefore, the diagnosis of TSC was made only after bleeding occurred from the renal AML, most likely triggered by pregnancy.

Several renal lesions such as AML and renal cysts are well recognized in patients with TSC.[19] Renal AML is the most common renal lesion occurring in up to 80% of adult patients with TSC.[20,21] Renal AMLs tend to increase in size over time and can distort the renal architecture, which can cause renal dysfunction and lead to renal failure in such patients.[19-21] In addition, renal AMLs contain abnormal blood vessels and aneurysms that may cause spontaneous rupture and bleed. In the present case, there were two additional risk factors such as pregnancy and TSC that increased the risk of rupture and bleeding. The life-threatening giant renal artery pseudo-aneurysm seen in our patient as a result of rupture of renal AML, which was treated by an endovascular approach, is unique and should draw attention to early diagnosis and treatment of these so-called benign lesions.[22,23]

In contrast to surgery, transcatheter embolization is minimally invasive without the need for general anesthesia, can preserve renal functions more effectively, is cheap and can be performed multiple times, as required.[24] The shorter recovery time and hospitalization are additional benefits of embolization over surgery. Several investigators have suggested that transcatheter embolization should be reserved for acute bleeding and multiple lesions in order to avoid life-threatening hemorrhage and preserve renal parenchyma.[14,18] In a patient with ruptured AML who presents with hypovolemic shock, the surgical option is usually radical nephrectomy due to impossibility to differentiate normal renal parenchyma from the huge hemorrhagic AML.[15]

There are different types of embolic agents that can be used for embolization of renal AMLs, including liquid agents such as nBCA (Onyx), polyvinyl alcohol particles, gelfoam and metallic coils. There is no consensus regarding the embolic agent of choice in the literature.[2] In the present case, nBCA was used as the embolic agent to embolize the parent artery from which the aneurysm originated, without filling the aneurysm sac itself. By this method, we aimed to achieve complete occlusion of the ruptured vessel with maximal nephron sparing and also to help thrombosis of the aneurismal sac and reduction in size with time. Reduction of size of the AML after embolization is an expected result and goal; however, efficacy should not be evaluated by only tumor size. The absence of bleeding episodes in a long-term follow-up period and the disappearance or at least reduction of the vascular component should be taken into account.[14]

In conclusion, spontaneous rupture of renal AML should be suspected in a puerperal patient with sudden onset of abdominal pain and hypovolemic shock. CT imaging should be the diagnostic tool of first choice. We suggest that trans-catheter embolization should be considered in high-risk patients with renal AML as the first therapeutic option to prevent the growth and spontaneous rupture of the lesions. Because renal failure is the most common cause of death in patients with the TSC, selective transcatheter embolization can be helpful both for preserving the renal functions and for preventing catastrophic complications.

Conflict of interest: None

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