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Title: Selective embolization in renal angiomyolipoma with pseudoaneurysm.

Short title: Renal angiomyolipoma with symptomatic pseudoaneurysm.

Abstract

Angiomyolipomas are usually diagnosed incidentally and are usually asymptomatic. Dismorphic blood vessels in angiomyolipomas usually do not contain an internal elastic lamina, so bleeding risks are high. Particularly, lesions larger than 4 cm have a tendency to become symptomatic and can present with life-threatening retroperitoneal or urinary bleeding. Although intrarenal or perinephric bleeding is the usual complication of angiomyolipomas, a pseudoaneurysm appears unusual. In this case, we aimed to present a patient with pseudoaneurysm, an unusual complication of angiomyolipoma. In this article, a patient who presented with complaints of left flank pain and hematuria and who underwent selective arterial embolization due to pseudoaneurysmatic angiomyolipoma in the left kidney is presented. The patient's angiography showed a large hypervascular mass filling the upper and middle segments of the left kidney. A pseudoaneurysmal filling originating from a subsegmental branch was observed in the mass. The patient underwent selective embolization. After 5 years of follow-up, the mass was observed to shrink. Angiomyolipomas are benign tumors, but especially symptomatic masses larger than 4 cm and especially masses with aneurysms larger than 5 mm can cause life-threatening retroperitoneal hemorrhages. Coexistence of angiomyolipomas with pseudoaneurysm is rare, especially. Safe, effective and minimally invasive selective arterial embolization can be safely performed to prevent massive bleeding and rupture.

Keywords: Angiomyolipoma, pseudoaneurysm, retroperitoneal hemorrhage, selective angiography.

Makale başlığı: Psödoanevrizmalı renal anjiyomiyolipomda selektif embolizasyon.

Öz

Anjiyomiyolipomalar genellikle tesadüfen teşhis edilir ve genellikle semptomsuzdur. Özellikle, 4 cm'den büyük lezyonlar semptomatik olma eğilimindedir ve yaşamı tehdit eden retroperitoneal veya üriner kanama ile ortaya çıkabilir. Anjiyomiyolipomaların tipik komplikasyonu intrarenal veya perinefratik kanamadır, ancak bir psödoanevrizma görünüşü olağandışıdır. Bu çalışmada anjiyomiyolipomanın olağan dışı komplikasyonu olan psödoanevrizmalı hastayı sunmayı amaçladık. Bu makalede, sol yan ağrısı ve hematüri şikayetleri ile başvuran ve böbrekteki psödoanevrizmatik anjiyomiyolipoma nedeniyle seçici arteriyel embolizasyon geçiren bir hasta sunulmaktadır. Hastanın anjiyografisi, sol böbreğin üst ve orta segmentlerini dolduran büyük bir hipervasküler kitleyi göstermiştir. Kütlede bir subsegmental dal kökenli psödoanevrizmal dolgu gözlemlenmiştir. Seçici anjiyoembolizasyon sonrasında, kitlede 5 yıllık takipte küçülme olduğu gözlemlenmiştir. Anjiyomiyolipomaların psödoanevrizma ile bir araya gelmesi nadirdir. Büyük kanama ve yırtılma önlemek için güvenli, etkili ve minimal invaziv seçici arteriyel embolizasyon güvenle uygulanabilir.

Anahtar kelimeler: Anjiyomiyolipoma, psödoanevrizma, retroperitoneal kanama, selektif anjiyografi.

Introduction

Angiomyolipomas (AMLs) are benign mesenchymal tumors composed of adipose tissue, smooth muscle, and blood vessels [1]. They are found in approximately 0.3% of the general population and constitute about 3% of all kidney tumors [2, 3]. AMLs are often identified as solitary lesions, most commonly originating from the kidney, and less frequently occurring in the liver, lymph nodes, spleen, lungs, and retroperitoneal area [4]. While they are predominantly sporadic, around 20% of cases may be associated with tuberous sclerosis (TS) [5].

Angiomyolipomas are usually diagnosed incidentally and are usually asymptomatic [6]. Dysmorphic blood vessels in angiomyolipomas usually do not contain an internal elastic lamina, so bleeding risks are high. Particularly, lesions larger than 4 cm have a tendency to become symptomatic and can present with life-threatening retroperitoneal or urinary bleeding [7, 8]. Therefore, minimally invasive procedures such as selective renal artery embolization or nephron-sparing surgery can be performed for ineffective pain management, masses larger than 4 cm and with a risk of bleeding and whose pain cannot be relieved [6].

Although intrarenal or perinephric bleeding is the usual complication of angiomyolipomas, a pseudoaneurysm appears unusual. Intrarenal pseudoaneurysm is also a well-known complication of penetrating renal injuries, renal surgery and percutaneous renal procedures [9, 10].

In this case, we present the clinical follow-up and treatment process of a patient who was evaluated for flank pain and gross hematuria and underwent selective arterial embolization (SAE) due to a pseudoaneurysmal left renal AML, in line with the current literature.

Case presentation

A 46-year-old female patient presented with complaints of left flank pain and hematuria. Upon follow-up examinations, a mass suggestive of AML was detected in the left kidney, she was referred to our department. The patient had no significant medical history except for a previous lumbar hernia surgery. Physical examination revealed an immobile lobulated mass in the left upper quadrant of the abdomen. There were no signs of tenderness or defense in the abdomen. Arterial blood pressure was measured as 100/70 mmHg, and the patient had a body temperature of 37.1°C. Chest X-ray and electrocardiography did not reveal any abnormalities. Laboratory findings showed a white blood cell count of 14.25 K/uL, hemoglobin level of 10.3 g/dL, hematocrit of 30.7%, and creatinine level of 0.69 mg/dL. Other routine biochemical values and coagulation parameters were within normal limits.

Abdominal magnetic resonance imaging (MRI) was performed after intravenous contrast agent (IVCA) injection. The MRI revealed a well-defined mass measuring 100x80x83 mm in size, located exophytically in the upper pole of the left kidney. The mass displaced the tail segment of the pancreas upward and anteriorly, and its borders could not be distinguished from the left suprarenal gland. The mass extended to the renal hilum and showed heterogeneous enhancement. The mass contained macroscopic fat (Figure 1), hemorrhagic signal intensity changes, and a pseudoaneurysm (Figure 2).

During the interventional radiology procedure, the patient's angiography revealed a large hypervascular mass filling the upper and middle segments of the left kidney. Within the mass, a pseudoaneurysmal filling originating from a subsegmental branch was observed (Figure 3). The microcatheter was advanced into the feeding vessel, and embolization was performed superselectively using four 3mm and two 4mm diameter coil embolization devices. Embolic particles with a size of 400 microns were used to perform embolization in the vascular bed supplying the mass through that branch, and achieving complete stasis (Figure 4).

The patient, who did not experience any complications during the post-procedure follow-up, was discharged after one week. During the 5-year follow-up period, the patient remained asymptomatic, and no bleeding was observed in the mass. Imaging studies revealed a decrease in the size of the AML in the left kidney. 5 years after the procedure, the patient's creatinine level was measured as 0.79 mg/dL. Abdominal computed tomography (CT) was performed after the injection of IVCA showed densities that were likely artifacts related to the embolic agents in the posterior upper segment of the left kidney. A well-defined hypodense, similar density to fat, smoothly contoured lesion measuring approximately 4.8x3.3 mm, was observed in the upper segment of the left kidney (Figure 5).

Discussion

Although angiomyolipomas are mostly asymptomatic, they can pose a threat to life due to their fragile nature, leading to spontaneous bleeding or serious complications such as vena cava compression in large tumors [7, 8]. AML is the most common cause of spontaneous retroperitoneal bleeding, and factors such as larger size (>4 cm), multifocality, and association with tuberous sclerosis increase the risk of bleeding [11]. "Wunderlich Syndrome," also known as massive retroperitoneal hemorrhage, is the most serious complication of renal angiomyolipoma and has been reported in more than 10% of patients [11]. Wunderlich Syndrome manifests with the Lenk triad, which includes flank or abdominal pain, palpable mass, and gross hematuria. If it is untreated, Wunderlich Syndrome can lead to significant morbidity and potential mortality [11, 12].

Selective arterial embolization and percutaneous radiofrequency ablation are prominent minimally invasive options in the treatment of AML. Surgical options such as partial/radical nephrectomy are preferred in more severe cases and selected patients [13]. The presence of symptoms such as tumor size increase, bleeding, and pain, as well as suspicion of cancer on imaging, constitute the main indications for the treatment of angiomyolipoma [14]. In cases of bleeding AML, embolization is implemented as the first-line treatment method and serves as a preventive treatment for masses at high risk of bleeding [15].

Although SAE is a minimally invasive procedure, it can still be associated with complications such as pain, post-embolization syndrome, hematuria, vascular injury, rupture during the procedure, infection, abscess formation, renal infarction, and kidney failure. Post-embolization syndrome (PES), which occurs as a result of renal tissue necrosis, manifests itself with symptoms like nausea, vomiting, fever, abdominal pain, and leukocytosis, and it is treated conservatively [16].

In a meta-analysis study conducted by Murray et al. [17] involving 524 patients, SAE performed for AML showed a success rate of 93.3% during an average follow-up period of 39 months. Different embolization agents were used in 46.8% of cases, with 2 or more agents used in some cases. Among the patients, 20.9% required retreatment due to revascularization, no change/increase in tumor size, persistent symptoms, or retroperitoneal bleeding. SAE was associated with low mortality and a 35.9% incidence of PES.

In a cohort study involving 41 patients with AML, SAE was performed on 48 kidneys, and no cases of retroperitoneal bleeding were observed during the follow-up period. There were no significant changes in creatinine levels ($p=0.27$). Five years after SAE, the rate of avoiding surgical treatment was reported to be 94%, and the disease-specific survival rate for the entire cohort was reported to be 100% [18].

In another single-center study, the results of 23 patients who underwent SAE for retroperitoneal bleeding ($n=6$) and prophylaxis purposes ($n=17$) were retrospectively evaluated. During an average follow-up period of 20.5 months, a 26.2% reduction in AML size was observed. Three patients experienced major complications, including renal abscess in 2 patients and femoral pseudoaneurysm in 1 patient. Additionally, 14 patients reported minor complications in the form of post-embolization syndrome (PES) [19].

Due to the hypervascularity of aneurysms, the risk of tumor rupture in AML is related to the presence and size of intratumoral aneurysms. The imaging findings, defined clinically as "aneurysm", are divided into 2 pathological types: primary aneurysm and pseudoaneurysm [20].

Yamakado et al. [7] investigated a total of 29 kidneys with AML, with 8 being hemorrhagic and 21 being non-hemorrhagic. They reported that when a tumor size of 4 cm or larger and an aneurysm size of 5 mm or larger were used as rupture indicators, the probability of rupture could be predicted with higher specificity.

In a meta-analysis that examined 739 cases of AML, aneurysms were demonstrated in the interlobar and interlobular arteries in 71.4% of the cases based on selective angiography results [21]. In another study that evaluated 27 patients diagnosed with AML and 34 kidneys, aneurysms were detected in 29.4% of AML patients using CT angiography. However, in this study in which 6 kidneys were ruptured, it was stated that tumor size and aneurysm size cannot be used as indicators of spontaneous rupture of the tumor [22].

Renal artery pseudoaneurysms observed in AMLs have been defined as unusual vascular complications that can cause bleeding. Therefore, it is anticipated that appropriate treatment should be initiated based on the size of the AML [23]. Furthermore,

in a study conducted by Albi et al. [12], it was stated that the presence of an intratumoral aneurysm on CT scan can predict a higher possibility of tumor rupture.

Sayin et al. [1], evaluated a 64-year-old patient who presented with severe abdominal pain, weakness, dizziness, and localized abdominal rigidity on the left side. The abdominal ultrasound (USG) revealed a well-defined AML that nearly filled the left half of the abdomen, measuring approximately 15 x 20 x 25 cm. Contrast-enhanced abdominal CT imaging showed a hyperdense area in the central region of the mass, consistent with a pseudoaneurysm. During the patient's follow-up, there was a progressive decrease in hemoglobin levels and a deterioration in the overall condition, leading to the decision to perform a total nephrectomy. Sayin et al. [1], emphasized that for AMLs larger than 4 cm and exhibiting symptoms, measures such as SAE or partial/total nephrectomy can be employed to prevent bleeding and rupture.

Esmat and Naseri [24], colleagues evaluated a 31-year-old patient who had been experiencing left-sided abdominal pain for a week and had recently developed hematuria. The patient had a medical history of bilateral AML and tuberous sclerosis. Four years ago, the patient had undergone SAE due to retroperitoneal hematoma. Contrast-enhanced abdominal CT images revealed findings consistent with AML in both kidneys. In the lower pole of the left kidney, a lesion was observed in the arterial phase, showing homogeneous contrast enhancement and being consistent with a pseudoaneurysm, with active extravasation observed in the adjacent area. No retroperitoneal bleeding was detected, and the patient underwent successful SAE. Esmat and Naseri [24], colleagues recommended SAE treatment for bleeding AMLs and those at risk of bleeding.

In this case in which SAE was performed due to pseudoaneurysm, no complications were observed during early and late follow-ups after the procedure. Additionally, there was no need for surgical intervention or any additional procedures during the follow-up period, and a significant reduction in lesion size was noted after 5 years. No renal function loss was observed during the patient's follow-up.

In conclusion, angiomyolipomas are benign tumors, but symptomatic masses larger than 4 cm and especially masses containing aneurysms larger than 5 mm can cause life-threatening retroperitoneal hemorrhages. Pseudoaneurysms seen in angioliipomas are an unusual component and should be followed carefully because of the risk of bleeding. Safe, effective and minimally invasive selective arterial embolization can be safely performed to prevent massive bleeding and rupture.

Conflict of interest: No conflict of interest was declared by the authors

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Authors' contributions to the article

A.S. and M.K., constructed the main idea and hypothesis of the study. O.L.T, developed the theory and arranged/edited the material and method section. Discussion section of the article written by M.B.D. and K.K.

Y.O. and S.C. reviewed, corrected and approved. In addition, all authors discussed the entire study and approved the final version.

List of abbreviations

AML: Angiomyolipoma

TS: Tuberous sclerosis

SAE: Selective arterial embolization

MRI: Magnetic resonance imaging

IVCA: Intravenous contrast agent

CT: Computed tomography

PES: Post-embolization syndrome

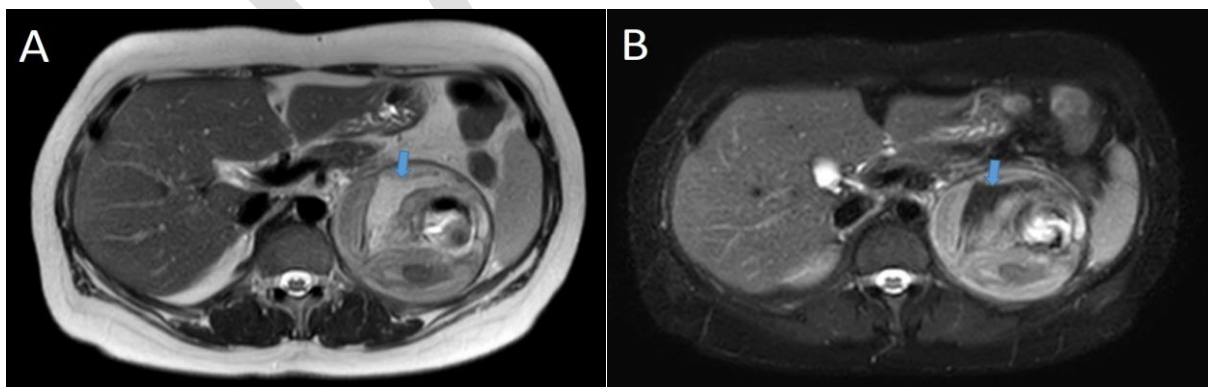


Figure 1. T2W image [A] and T2W fat saturated image showed macroscopic fat (blue arrow)

The mass contained macroscopic fat

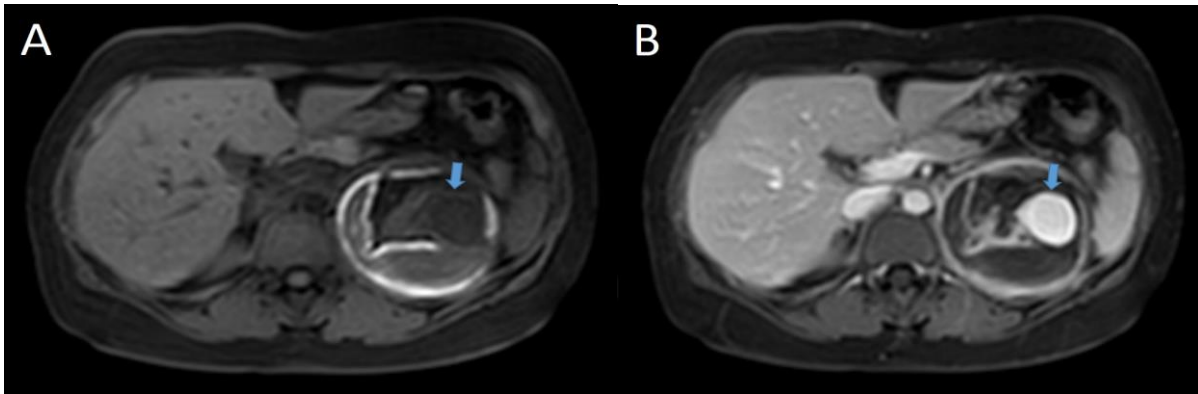


Figure 2. T1W pre-contrast fat saturated image [A] and T1W postcontrast fat saturated image showed a pseudoaneurysm (blue arrow) and peripheral hemorrhage Hemorrhagic signal intensity changes, and a pseudoaneurysm

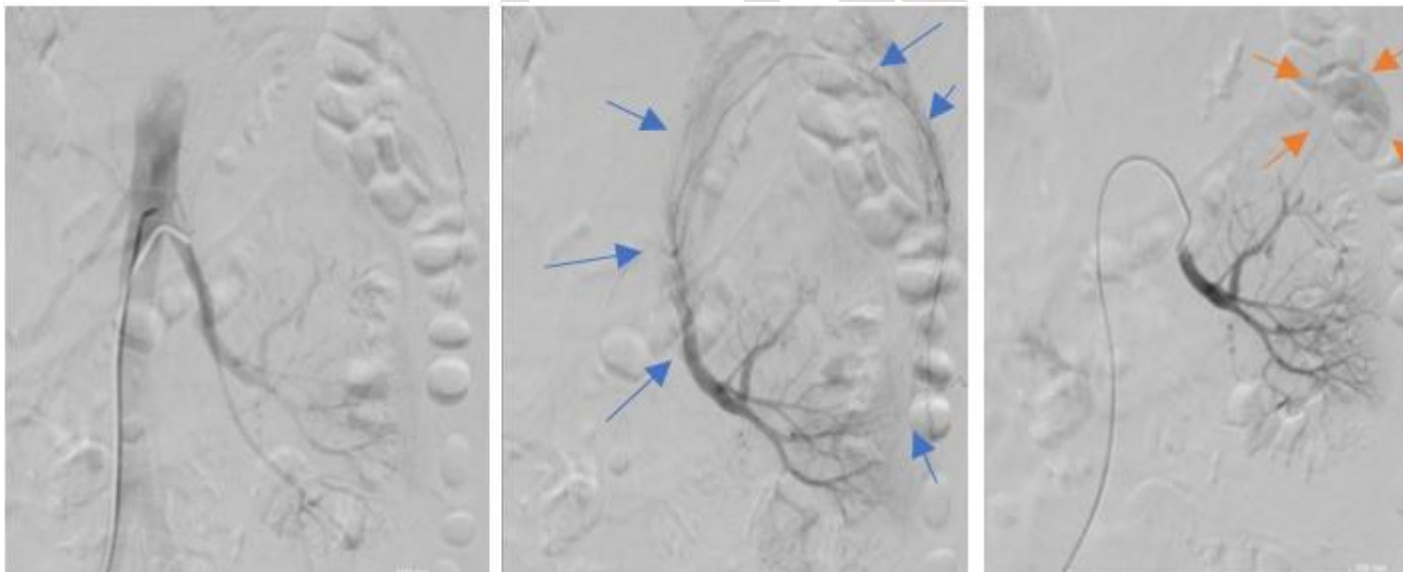


Figure 3. Angiography images after contrast agent injection through the catheter before the procedure

AML mass lesion indicated by blue arrows; area indicated by orange arrow represents the pseudoaneurysm

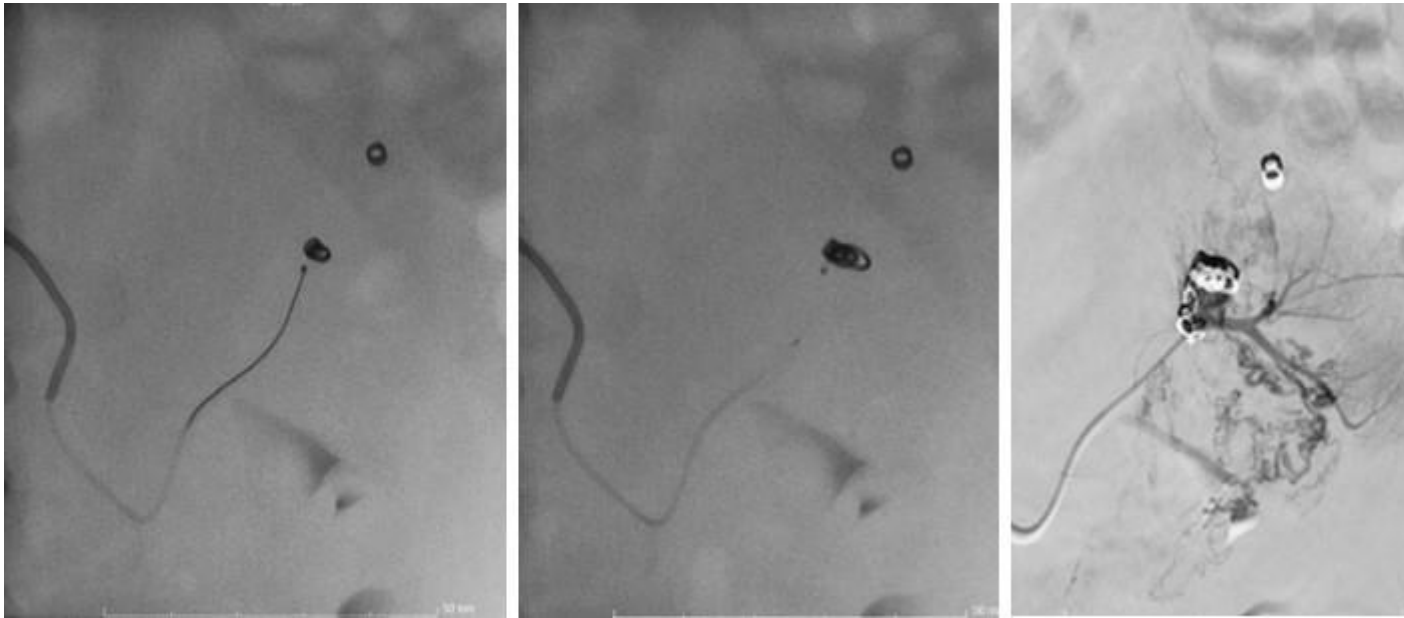


Figure 4. Angiography images after embolization

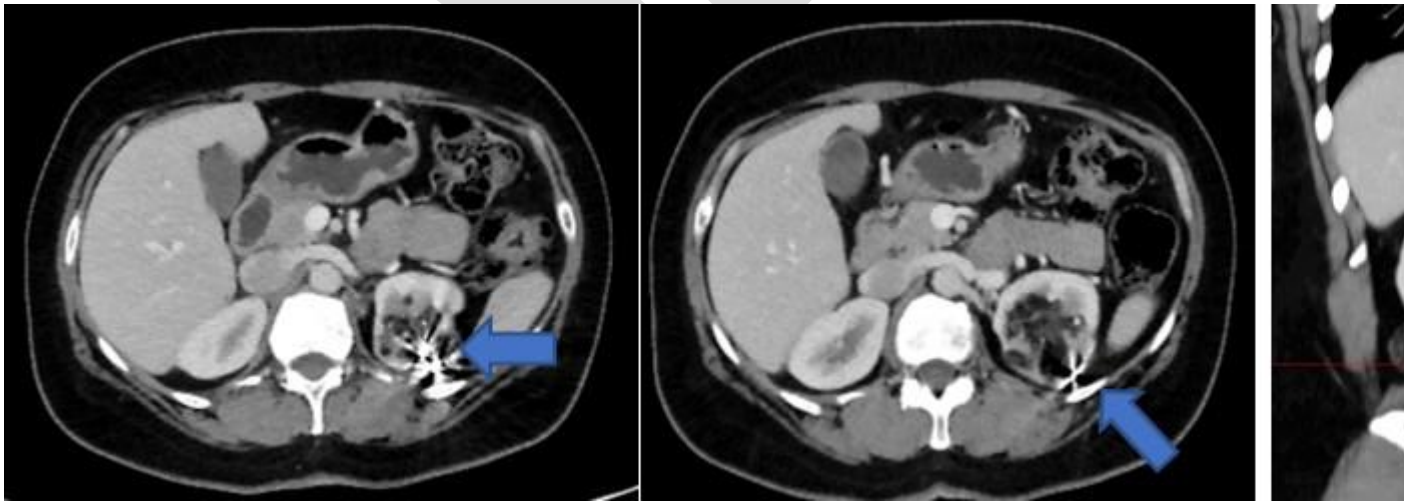


Figure 5. a. Axial section CT

b. Coronal section CT

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