

# Bilateral Renal Artery Dissections in a Patient with Segmental Arterial Mediolyis

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## INTRODUCTION

- Segmental arterial mediolysis (SAM): a rare non-inflammatory, non-atherosclerotic arteriopathy involving lysis of the smooth muscle of the outer media wall, usually involving visceral abdominal arteries
- Presentation variable based on arteries involved but requires prompt recognition
- High initial mortality upon presentation due to risk of acute rupture and hemorrhage

## PATIENT PRESENTATION

### History of Present Illness:

- 52 year-old man with history of chronic low back pain and one prior episode of nephrolithiasis presents to the ED with a 1-day history of right-sided flank pain
- Associated subjective chills, nausea, and non-bloody non-bilious emesis
- Describes flank pain as similar to prior episode of nephrolithiasis
- No other GI, GU, or MSK symptoms; review of systems was otherwise negative

### Family History:

- Both parents and a brother and sister all alive and well

### Social History:

- No history of tobacco use, alcohol use, or other drug use
- Gulf War Army veteran, stationed in Kuwait and Iraq

### Medications:

- Gabapentin, omeprazole, tramadol
- No regular aspirin or NSAID use, no OTC/supplement use

### Physical Exam:

- Vital Signs: afebrile, BP 170s/100s, HR 80s, SpO2 97% RA
- Exam: mucous membranes dry, mild right-sided CVA tenderness, otherwise exam normal

### Laboratory Data:

- CBC: normal
- BMP: serum creatinine 1.0 (baseline 0.7), otherwise WNL
- LFTs: total bilirubin 2.8 (prior mild elevation 1.3-1.7 noted in historical labs)
- PT/INR, PTT normal
- Urine: 1 WBC, 2 RBC, 20 ketones, no protein, otherwise normal; urine drug screen negative
- ESR & CRP slightly elevated: ESR 17 (ULN 15), CRP 12 (UNL 3)
- Infectious & Rheumatologic work-up negative
  - ANA negative, complement levels normal, ANCA panel normal
  - HIV, HCV, RPR, quantiferon gold, and Bartonella testing negative

### Imaging:

- CT Abdomen/Pelvis Non-Contrast: fat surrounding right renal artery, suggest CTA for evaluation of thrombus versus vasculitis
- CT Angiogram Abdomen/Pelvis: mural thickening with luminal irregularity and severe stenosis of the right renal artery, suspicious for vasculitis; delayed right renal nephrogram due to renal artery stenosis

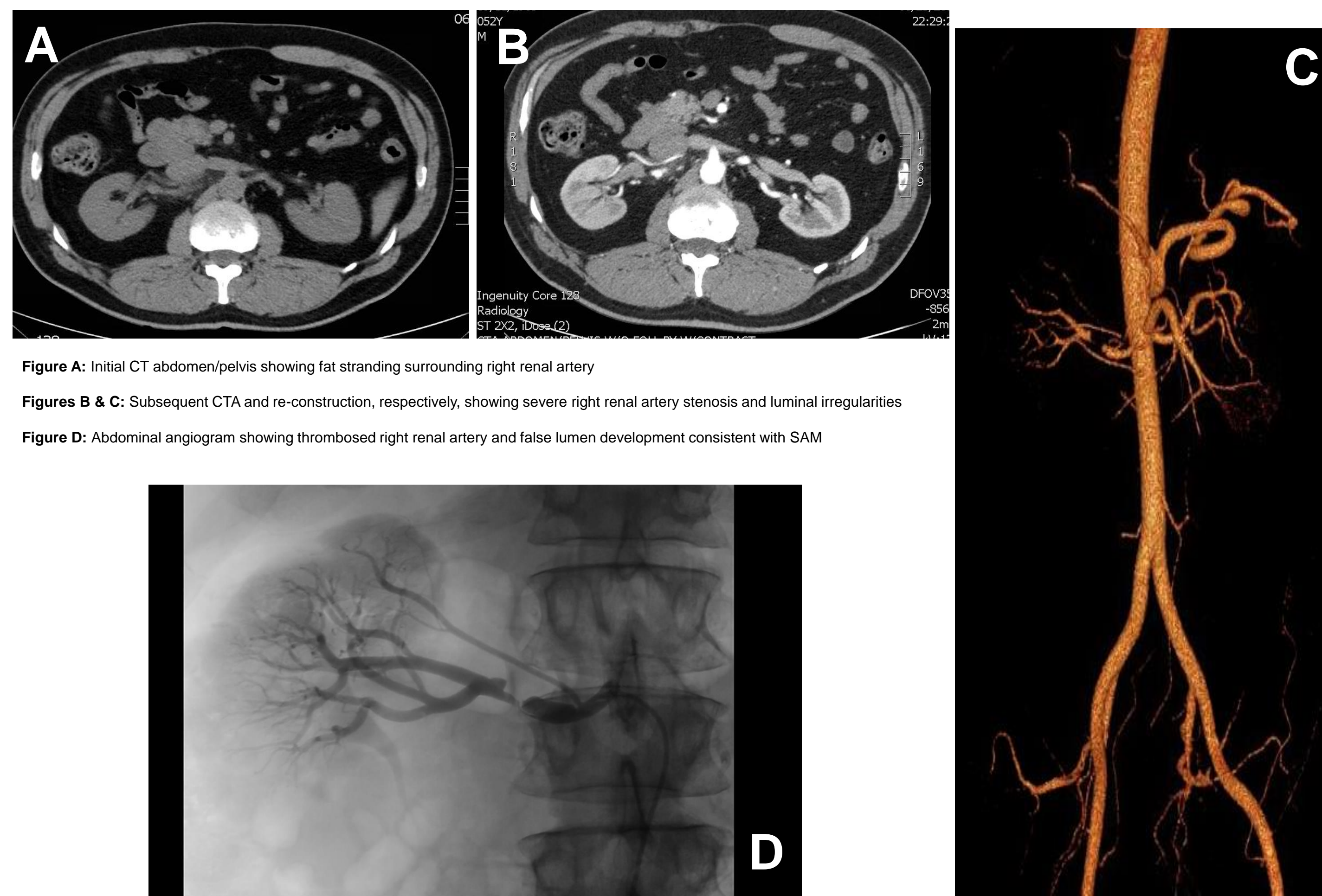


Figure A: Initial CT abdomen/pelvis showing fat stranding surrounding right renal artery

Figures B & C: Subsequent CTA and re-construction, respectively, showing severe right renal artery stenosis and luminal irregularities

Figure D: Abdominal angiogram showing thrombosed right renal artery and false lumen development consistent with SAM

## CLINICAL COURSE

- Admitted for further work-up given initial CT findings concerning for thrombus versus vasculitis
- CT angiogram as discussed also concerning for possible vasculitis
- Multiple infectious, rheumatologic labs returned non-diagnostic
- Ultimately underwent abdominal angiogram:
  - Thrombosed right renal artery with false lumen and high-grade stenosis, findings consistent with segmental arterial mediolysis
  - Two right-sided collateral renal arteries
  - An uncomplicated left renal artery stenosis
- Thereafter, in consultation with vascular surgery, patient started on anticoagulation (heparin gtt, then transitioned to warfarin with LMWH bridge)
- Post-procedural course complicated by local right femoral artery pseudoaneurysm, requiring repair
- MRA brain and U/S carotids obtained, which did not show SAM of cerebral or carotid arteries
- Patient discharged on hospital day #4 on anticoagulation and anti-hypertensive medications, with vascular and primary care follow-up
- Subsequent CT angiogram four weeks later:
  - Stability of known bilateral (right > left) renal artery stenosis
  - A new focal uncomplicated, non flow-limiting dissection of a branch of the left renal artery
- Patient's blood pressure has since normalized and is no longer requiring anti-hypertensive medications. He remains on anticoagulation.

## DISCUSSION

- This is a case of a patient presenting with bilateral renal dissections due to segmental arterial mediolysis – a rare presentation of a rare condition
- Both renal dissections and SAM have distinct but overlapping differentials, including uncommon conditions that require prompt recognition for appropriate management
- SAM in particular requires early recognition as it carries a high risk of mortality due to risk of arterial hemorrhage. Its long-term natural history remains unclear.

### Spontaneous Renal Artery Dissection

- Represents <0.05% of all arterial dissections
- Can present as a distinct clinical entity
- Differential Diagnosis
  - Fibromuscular Dysplasia
  - Marfan Syndrome
  - Ehlers-Danlos Syndrome
  - Thromboembolism
  - Atherosclerotic Disease
  - Malignant Hypertension
- Management dependent upon etiology of dissection

### Segmental Arterial Mediolyis

- Background
  - Initially described in 1976, 14 cases described since 1997, 45 cases described since 2007
  - Increasing awareness with development of CTA
  - True prevalence probably underestimated from subclinical disease
- Presentation
  - Middle-aged/older (50-80 year-old) males (1.5:1 M:F ratio)
  - Typically present with abdominal/flank pain due to commonly affected visceral arteries
  - Initial mortality rate up to 50% due to spontaneous intra-abdominal/peritoneal hemorrhage
- Pathology
  - Non-inflammatory, non-atherosclerotic arteriopathy, underlying etiology unknown
  - Involves lysis of the smooth muscle of the outer media arterial wall
  - Most commonly affects visceral abdominal arteries
  - Less commonly may affect the carotid or cerebral arteries
  - May present as aneurysm, stenosis, dissection, or occlusion of the artery
- Differential Diagnosis
  - Vasculitis (PAN most specifically)
  - Fibromuscular Dysplasia
  - Thromboembolism
  - Infection/Mycotic Aneurysm (Bartonella, TB, syphilis)
  - Ehlers-Danlos Syndrome
  - Cystic Medial Necrosis (seen in Marfan Syndrome)
- Management
  - No standard recommendation for management currently exists
  - Previously managed with coil embolization or surgical ligation or resection, unclear benefit to this
  - Avoid steroids (may potentially worsen SAM)
  - Expert opinion recommendation to check carotid and cerebral arteries, routine surveillance imaging and prophylactic treatment if aneurysms develop
- Prognosis
  - High mortality upon initial presentation due to acute rupture and hemorrhage
  - Multiple aneurysms may occur in up to 1/3<sup>rd</sup> of cases
  - Increased risk of recurrent aneurysms at 9-18mo after initial diagnosis
  - Two case reports of intra-cranial SAM, both fatal
  - Natural history and long-term prognosis unknown
    - Case reports following patients out to 4 years after diagnosis
    - Some cases resolve/stabilize, some fluctuate in stability, some shift to adjacent artery segments

### References:

- Pillai AK et al. "Segmental Arterial Mediolyis" Cardiovascular Interventional Radiology 2014 37: 604-612
- Stawicki W et al. "Spontaneous renal artery dissection: three cases and clinical algorithms" 2006 Journal of Human Hypertension 20:710-8