Bilateral Renal Artery Dissections in a Patient with Segmental Arterial Mediolysis

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INTRODUCTION

- Segmental arterial mediolysis (SAM): a rare non-inflammatory, non-atherosclerotic arteriopathy involving pairs 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 nephro lithiasis presents to the ED with a 4-day history of right flank pain
- Associated subjective chills, nausea, and non-bloody non-olentis pain
- Describes flank pain as similar to prior episode of nephrolithiasis
- No other GI, GU, or MSK symptoms; review of systems was otherwise non-specific

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, 200mg, bd; regular ACE inhibitor, no NSAID use, no OTC/supplement use

Physical Exam:
- Vital Signs: afebrile, BP 170s/100s, HR 80s, SpO2 97% RA
- Malignant Hypertension

LFTs: total bilirubin 2.8 (prior mild elevation 1.3)

Multiple aneurysms may occur in up to 1/3 of cases

Ultimately underwent abdominal angiogram:

Natural history and long term management currently unknown.

Cases of cases resolve/stabilize, some fluctuate in stability, some shift to adjacent artery segments

Multiple infectious, rheumatologic labs returned negative

Thromboembolism

Describes flank pain as similar to prior episode of nephrolithiasis

Gulf War Army veteran, stationed in Kuwait and Iraq

SAM in particular requires early recognition

Increasing awareness with development of CTA

Patient discharged on hospital day #4 on anticoagulation and anti-platelet therapy

Most commonly affects visceral abdominal arteries

May present as an aneurysm, stenosis, dissection, or occlusion of the artery

Syndrome

Ehlers-Danlos

HIV, HCV, RPR, TPHA negative

No regular aspirin or NSAID use, no OTC/supplement use

CT Angiogram Abdomen/Pelvis:

Stability of known bilateral (right > left) renal artery stenosis

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

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

Aneurysm:
- Thrombosed right renal artery with false lumen and high-grade stenosis

Dissection:
- An unruptured left renal artery stenosis

Thrombosis:
- Patient discharged on hospital day #4 on anticoagulation and anti-hyperensive 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 artery dissections due to segmental arterial mediolysis
- 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 classic clinical entity
- Differential Diagnosis
  - Fibromuscular Dysplasia
  - Marfan Syndrome
  - Ehlers-Danlos Syndrome
  - Malignant Hypertension
- Management dependent upon etiology of dissection

Segmental Arterial Mediolysis
- 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 olds)
  - Typically present with abdominal/flank pain due to commonly affected visceral arteries
- Initial mortality rate up to 50% due to spontaneous intra-abdominal/perineal hemorrhage
- Pathology
  - Non-inflammatory, non-atherosclerotic arteriopathy
  - Underlying etiology unknown
  - Involves types of the smooth muscle of the outer media arterial wall
  - Most commonly affects visceral abdominal arteries
  - Leaks commonly affect the celiac or cerebral arteries
- May present as aneurysm, stenosis, dissection, or occlusion of the artery
- Differential Diagnosis
  - Vasculitis (PAN most specifically)
  - Fibromuscular Dysplasia
  - Infection
    - Mycotic/Mycotic Arteritis
    - Bartonella, TB, syphilis
  - Malignancy
- Management
  - No standard treatment for management currently exists
  - Previously modified with coil embolization or surgical ligation or resection, uncurer beneficial to this
  - Avoid steroids (may potentially worsen SAM)
  - Expert opinion recommendation to check cardiac 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 1/3rd of cases
  - Increased risk of recurrent aneurysms at 6-18mo 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, some shift to adjacent artery segments