Hiding in Plain Sight:
Medullary Sponge Kidney

Michael Mohan, Harvard Medical School Year IV
Gillian Lieberman, MD

September 17, 2010
Our patient: History

Ms. C, a 49-year-old woman with a history of recurrent nephrolithiasis, presents to the emergency department with intermittent flank pain.

She is afebrile and has no significant findings on physical exam. Labs are notable for microscopic hematuria.

You suspect renal stone disease.
What imaging would you order to further evaluate our patient?

Please continue for discussion…
Choice of imaging in suspected nephrolithiasis

Noncontrast CT:
- The current standard of care
- 95% sensitive and 98% specific for stone disease
- Can look for dilation of the collecting system (hydronephrosis/hydroureter) which may signal partial or complete obstruction; may also allow diagnosis of a passed stone
- Accurate detection of stone size and location allows estimation of likelihood of spontaneous passage
- May see other intraabdominal pathology in the differential diagnosis

Ultrasound:
- May be first line test in pregnant women or patients in whom biliary and/or gynecologic pathology are high on the differential
- Excellent for diagnosing obstruction (hydronephrosis, hydroureter, and perirenal fluid); however, there is a time lag
- Difficult to see ureteral stones

Abdominal plain film:
- Will not see radiolucent stones
- Cannot evaluate for obstruction
- Not used when CT is available except in patients with recurrent radiopaque stones

Intravenous urogram (IVU):
- Previous gold standard, now replaced by noncontrast CT which is more sensitive, avoids need for contrast, is more rapid, and can identify alternative diagnoses
You have read about the benefits and drawbacks of different imaging modalities for suspected nephrolithiasis. For our patient, the choice of imaging made in the emergency department was noncontrast CT.

Please continue to see the images…
Our patient: Please evaluate the noncontrast abdominal CT
Our patient: Please evaluate the noncontrast abdominal CT

PACS, BIDMC
Coronal C- CT

PACS, BIDMC
Sagittal C- CT
You have had a chance to evaluate our patient’s abdominal CT. Please continue to see the conclusions of the radiologists who read the film…
Our patient: Official read of noncontrast abdominal CT

- No hydrenephrosis
- No cysts or soft tissue masses
- **1.8 cm stone** in the left renal pelvis, nonobstructing
- **Bilateral nephrocalcinosis** (calcium deposition in renal perenchyma and tubules)
Q: In what part of our patient’s kidneys is the calcinosis?
Let us review renal anatomy to help illustrate the location of our patient’s nephrocalcinosis…
Renal anatomy


Our patient’s nephrocalcinosis is localized to the medulla. Please continue to view our patient’s images next to a diagram of renal anatomy to illustrate the medullary location of our patient’s nephrocalcinosis...
Our patient: **Medullary calcinosis** on axial CT
Our patient: **Medullary calcinosis on plain film**

Our patient: Medullary calcinosis on plain film
Our patient: Medullary calcinosis on abdominal CT with contrast
You have seen images of our patient’s medullary calcinosis (also called medullary nephrocalcinosis). What is the differential diagnosis for this condition?

Please continue for a discussion…
Medullary nephrocalcinosis: Differential diagnosis

- Hyperparathyroidism
- Medullary sponge kidney
- Distal renal tubular acidosis
- Less commonly:
  - Milk-alkali syndrome
  - Hypervitaminosis D
  - Sarcoidosis
  - Others
We have seen the differential diagnosis for medullary calcinosclerosis. Our patient has no electrolyte abnormalities and her nephrocalcinosis is longstanding. She has a diagnosis of medullary sponge kidney (MSK).

Please continue to learn more about medullary sponge kidney…
Medullary Sponge Kidney: Facts

- **Irregular dilatations of the medullary and papillary collecting ducts (cortex is spared)**
- Congenital but family history usually absent; exact etiology unknown
- Common but frequency unknown because usually benign and discovered incidentally
- 70% bilateral at diagnosis, usually affects all papillae of an affected kidney
Medullary Sponge Kidney: Clinical Manifestations

- Usually asymptomatic and found incidentally on abdominal imaging
- Urinary stasis and perhaps impaired calcium resorption predispose to nephrolithiasis
  - MSK found in 12-20% of patients with stones
- Stasis in cysts predisposes to urinary tract infections (UTIs)
- May give microscopic or gross hematuria
Medullary Sponge Kidney: Management and Prognosis

Treatment is symptomatic:
- Drink fluids to prevent stones
- Treat UTIs

Prognosis is good – renal function unaffected unless recurrent stones or UTIs
You have learned about the clinical presentation and management of medullary sponge kidney. Please continue to learn about the characteristic features of medullary sponge kidney on various imaging modalities…
Companion patients 1 and 2: Appearance of MSK on plain film

- **Medullary calcinosis** or clusters of **pericalyceal stones**
  (in the medullary tubules)
- Appearance on plain film is nonspecific

http://www.accessmedicine.com
Frontal abdominal plain film

Tanagho EA, McAninch JW: Smith's General Urology, 17th Edition:
http://www.accessmedicine.com
Frontal abdominal plain film
Companion patient 3: Appearance of MSK on ultrasound

- Hyperechoic medullary region due to calcifications
- Pyramids often dilated but pelvis and calyces normal
- Also nonspecific


Transabdominal ultrasound
Companion patients 4 and 5: Appearance of MSK on intravenous urogram

- Pyelogram phase most useful – shows collecting system
- Accumulation of contrast in distended collecting ducts of medullary pyramids seen as “brush” or “blush” appearance radiating outward from calyces

Frontal view intravenous urogram

Frontal view intravenous urogram
Companion patient 6: Appearance of MSK on CT

- Without contrast: medullary calcinosis or pericalyceal stones
- With contrast: may see characteristic “blush” pattern from ectatic collecting ducts
You have learned about the characteristic imaging features of medullary sponge kidney. Let us return now to our patient Ms. C to see how she is doing...
Our patient: Clinical course

- No obstructing stone found on imaging
- However, she was found to have a UTI, with 50,000-100,000 colonies/mL *Pseudomonas aeruginosa* on urine culture
- Predisposed to UTIs by medullary sponge kidney, has had many UTIs in the past
- Currently undergoing treatment with antibiotics
Conclusions

- Noncontrast CT is generally the first line test for suspected nephrolithiasis
- Medullary sponge kidney is common and predisposes to recurrent stones as well as UTIs
- Medullary sponge kidney is often diagnosed when medullary calcinosclerosis is found incidentally
- Other common causes of medullary calcinosclerosis are hyperparathyroidism and renal tubular acidosis
Acknowledgements

- Dr. Gillian Lieberman, Course Director
- Dr. Rich Rana, Senior Resident in Radiology
- Emily Hanson, Educational Coordinator
- Larry Barbaras, Webmaster
References


