Kaplan USMLE Step 1: Flank pain in patient with sickle cell disease

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If you’re preparing for the United States Medical Licensing Examination® (USMLE®) Step 1 exam, you might want to know which questions are most often missed by test-prep takers. Check out this example from Kaplan Medical, and read an expert explanation of the answer. Also check out all posts in this series.

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This month’s stumper

A 25-year-old man with sickle cell disease comes to the emergency department because of a one-day history of sharp left flank pain associated with nausea and vomiting. Three days ago, he began seeing blood in the urine without symptoms of pain or fever. He does not have a history of significant kidney disease. His temperature is 37.2°C (99.0°F), pulse is 110 beats per minute, respirations are 20 breaths per minute, and blood pressure is 140/95 mm Hg.

Physical examination shows a young man in distress from pain and left costovertebral angle tenderness. The remainder of the physical examination shows no abnormalities. Laboratory studies show hemoglobin 8.5 g/dL, hematocrit 23.5%, leukocyte count 9,000/mm³, platelet count 300,000 mm³, serum urea nitrogen 36 mg/dL and serum creatinine 1.8 mg/dL. Urinalysis shows 4+ blood, trace protein, and negative leukocyte esterase and nitrite. A CT scan of the abdomen and pelvis is ordered.

Which of the following is the most likely cause of this patient’s symptoms?

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A. Nephrolithiasis.
B. Pyelonephritis.
C. Renal medullary carcinoma.
D. Renal papillary necrosis.
E. Ruptured renal cyst.
The correct answer is D.

Kaplan Medical explains why

Patients with sickle cell disease may suffer from various manifestations of renal injury, collectively termed sickle cell nephropathy (SCN). The inner renal medulla and vasa recta capillaries are relatively hypoxic and hyperosmotic with increased hyperviscosity and slow blood flow. This environment is ideal for the sickling of erythrocytes that cause obstruction of blood flow and eventual loss of the vasa recta.

The hallmark of SCN is the combination of impaired renal concentrating capacity and normal diluting capacity due to the loss of the countercurrent exchange mechanism in the inner renal medulla and loss of the vasa recta capillaries and loops of Henle of the juxtamedullary nephrons. Other manifestations of SCN include impaired urinary acidification and potassium excretion, proteinuria from focal segmental glomerulosclerosis, hematuria, and chronic kidney disease.

Hematuria is a common clinical manifestation in sickle cell disease. Renal papillary necrosis (RPN), a frequent cause of hematuria in this population, arises from sickling erythrocytes that lead to microthrombotic infarction and vasa recta obliteration. Incidence of RPN ranges from 20% to 65% in studies of sickle cell disease. The most common presentation of RPN is painless macroscopic hematuria. Renal colic caused by the passage of necrotic papillae resulting in urinary tract obstruction and post-renal acute kidney injury can also be seen with RPN, and is the most likely scenario explaining this patient’s presentation in this clinical vignette.

In addition to sickle cell disease, RPN can also be observed in patients with diabetes mellitus, acute pyelonephritis, and chronic analgesic abuse. Definitive diagnosis of RPN is made with CT scan.

Why the other answers are wrong

Choice A: Nephrolithiasis can present with gross hematuria, renal colic, and post-renal acute kidney injury. However, nephrolithiasis is not a commonly associated renal manifestation in sickle cell disease.

Choice B: Pyelonephritis and urinary tract infections can be more commonly encountered in sickle cell disease compared to the general population because of the functional asplenia and increased susceptibility to bacterial infections seen in sickle cell patients. Pyelonephritis can present with gross
hematuria and flank pain, and renal papillary necrosis is a known complication. This patient’s lack of fever, elevated leukocyte count or elevated leukocyte esterase and nitrite on urinalysis makes this option less likely.

Choice C: Renal medullary carcinoma has been recognized as a specific entity in sickle cell nephropathy. It is an aggressive form of renal cell carcinoma that can present with gross hematuria, flank pain, and weight loss, and is usually metastatic at diagnosis. This disease is rare compared to renal papillary necrosis in sickle cell disease and makes this option incorrect.

Choice E: A ruptured renal cyst can present with gross hematuria and flank pain. Ruptured renal cysts are more commonly encountered in patients with autosomal dominant polycystic kidney disease. Renal cysts are not associated with sickle cell nephropathy.

Tips to remember

- Hematuria is a common clinical manifestation in sickle cell disease.
- Renal papillary necrosis in sickle cell disease arises from sickling erythrocytes that lead to micro-thrombotic infarction and vasa recta obliteration.
- The most common presentation of renal papillary necrosis is painless macroscopic hematuria, and renal colic caused by the passage of necrotic papillae resulting in urinary tract obstruction and post-renal acute kidney injury can also be seen.
- Renal papillary necrosis is also observed in patients with diabetes mellitus, acute pyelonephritis, and chronic analgesic abuse.

For more prep questions on USMLE Steps 1, 2 and 3, view other posts in this series.