CLINICAL VIGNETTE

Renal Papillary Necrosis in a Pregnant Patient with Sickle Cell Disease

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Case Presentation

A 22-year-old female who was 25 weeks pregnant presented to the labor and delivery unit with hematuria. She has sickle cell disease complicated by vasoocclusive crises and acute chest syndrome. For the past day, she noticed maroon urine with occasional clots and mild dull right flank pain. She denied fevers, chills, nausea, vomiting or prior history of hematuria or nephrolithiasis. Her only medication was a prenatal vitamin. Prior to her pregnancy, she was taking voxelotor and folic acid but they have been held since she became pregnant. She denied tobacco, alcohol, illicit drug use, and family history of nephrolithiasis.

The patient's initial vitals were temperature of 37.3 degrees Celsius, heart rate of 102 beats per minute, blood pressure of 107/66 mm Hg, respiratory rate of 16, and oxygen saturation of 97%. On physical examination, her abdomen was soft, nondistended, and nontender. She had no costovertebral angle (CVA) tenderness. Laboratories included a normal basic metabolic panel (BMP) and CBC notable for hemoglobin of 8.3 g/dL, near her baseline hemoglobin level. Urinalysis was unremarkable and urine culture was negative. Renal ultrasound showed moderate hydronephrosis and heterogeneously isoechoic fluid within the dilated right kidney collecting system that likely represented blood products.

Urology was consulted and felt her hematuria was from renal papillary necrosis and the hydronephrosis was secondary to obstruction from sloughed papilla or blood clots. Urology recommended no further imaging and conservative management with bedrest. After four days, her hematuria resolved. Her hospital course was complicated by vasoocclusive pain crisis requiring intravenous (IV) hydration, opioids, and an exchange transfusion before her pain resolved and she was discharged home.

Discussion

Renal papillary necrosis (RPN) is ischemic necrosis of the papilla in the medulla of the kidneys. Contributing factors include analgesic use, diabetes mellitus, urinary obstruction, and sickle cell disease (SCD). As a complication of SCD, papillary necrosis has an incidence of 30-40%.[1] RPN in SCD results from intravascular stasis and thrombosis because of the sickling of the red blood cells, causing obstruction of the micro-circulation, resulting in ischemic necrosis.[2] It typically presents with mild, painless, self-limited bleeding. Flank and abdominal pain can occur because of sloughing of papillae, and symptoms of urinary tract infection may be present. If obstruction occurs, renal failure can also develop. Renal ultrasound is a useful initial test and shows increased echogenicity of the inner medulla. Computed tomography (CT) scans show ischemic changes with greater accuracy than ultrasound, but is not required unless there is concern for obstruction or need for urologic intervention.[3]

Pregnancy exacerbates SCD, resulting in severe complications for the mother and fetus. In pregnancy, there is an increase in metabolism, blood stasis, and coagulability, increasing the risk of cardiomyopathy or pulmonary hypertension, and SCD complications such as vaso-occlusive crisis, acute chest syndrome, and venous thrombosis. Pregnant SCD patients are also more likely to experience pregnancy-related complications such as preeclampsia, eclampsia, antepartum bleeding, preterm labor, and fetal growth restriction.[4] Studies have shown that prophylactic blood transfusions may reduce pain crisis and acute chest syndrome but have not affected maternal or fetal outcomes. Blood transfusions in pregnant SCD patients are usually reserved for complications of SCD and for severe acute anemia.[5]

Most cases of RPN from SCD are self-limited, and resolve after bedrest, hydration, and occasionally blood transfusion. Bedrest is recommended to avoid dislodging blood clots. Hydration and alkalization of the urine reduces the toxicity from heme pigment that is released by hemoglobin. In some cases, exchange blood transfusion can be performed to decrease the percentage of hemoglobin S.[6] When there is acute obstruction and urinary tract infection, immediate surgical intervention is required. In these cases, percutaneous nephrostomy is the treatment of choice, with ureteral stent placement an alternative.

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