

Acute Renal Infarction With Heritable Coagulopathy and Patent Foramen Ovale

Eric M. Vilbert, M.D., and Stefan V. Franciosa, D.O.

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Abbreviations: CT, computed tomography; MRI, magnetic resonance imaging

Eric M. Vilbert, M.D. (Email: VilbertE@mlhs.org), is in the department of Radiology at the Bryn Mawr Hospital, Main Line Health System, Bryn Mawr, PA, United States of America.

Stefan V. Franciosa, D.O. (Email: FranciosaS@mlhs.org), is in the Department of Interventional Radiology of the Main Line Health System, Bryn Mawr, PA, United States of America.

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Abstract

We present the case of a 27-year-old woman who presented to the emergency department with the acute onset of left sided abdominal pain. Initial CT examination showed multiple renal infarcts in the lower pole of the left kidney, and an angiogram showed thrombus in a segmental branch of the left renal artery. Subsequent transesophageal echocardiogram demonstrated a small patent foramen ovale with bidirectional shunting, and serum coagulopathy evaluation demonstrated a G20210A prothrombin gene mutation. We conclude that the renal infarctions were caused by a paradoxical embolic event in the setting of an inherited coagulopathy and a patent foramen ovale.

Introduction

As in other organ systems, renal infarction is caused by a hypoxic insult to the kidney secondary to disruption in blood supply. This disruption can be secondary to obstruction of either the arterial or venous systems. Occlusion may be secondary to direct renal vessel thrombosis, thromboembolism, or secondary to renal trauma.

We present a case of renal infarction caused by angiographically proven renal artery embolus in a patient with multiple risk factors for thrombosis. These risk factors included a prothrombin gene mutation, oral contraceptives, and a patent foramen ovale. The most likely source was a paradoxical embolus from the deep venous system, although a definitive deep vein thrombosis was not identified on duplex evaluation of the extremities.

Case Report

A 27-year-old woman presented to the emergency department with the acute onset of diffuse abdominal pain, greatest in the left lower abdomen and flank. She had no significant past medical history, and the only medication she was taking was an oral contraceptive (Loestrin-FE). She was a nonsmoker and drank socially. At the time of presentation, she was initially evaluated with blood work and a contrast enhanced CT scan of the abdomen and pelvis with a clinical suspicion of bowel pathology. Her initial blood work showed a mildly elevated WBC count (10,200/mm³), but was otherwise unremarkable. The CT scan showed perfusion defects in the lower pole of the left kidney (Fig. 1), leading to a strong suspicion for renal infarction, or less likely a renal infection. The patient was admitted to the hospital and further imaging was obtained. A Doppler ultrasound of the abdomen and renal vasculature was unremarkable. An MRI/MR angiogram was then obtained which again showed perfusion defects in the lower pole of the left kidney; sonography with Doppler showed no renovascular abnormality (Fig. 2). A cortical rim sign was evident on both CT and MRI, but much better appreciated on MR images (Fig. 3). Concomitantly, an embolic workup was performed with a negative Doppler ultrasound of the lower extremities and a normal transthoracic echocardiogram. The patient's INR, PT, and PTT were also normal.

The patient had an elevated LDH (474 U/L), which can be seen with renal infarction, and based on the strong clinical suspicion, the patient was then sent for direct renal angiography for further investigation. At angiography, an ovoid filling defect was found in a segmental branch of the left renal artery in the lower pole, consistent with thrombus (Fig. 4). There was no evidence of vasculitis or aneurysm, and the main renal artery and vein were both patent. When thrombus was confirmed, a transesophageal echocardiogram was performed which showed a small patent foramen ovale with bidirectional shunting (Fig. 5). A serum coagulation panel was also obtained, and the patient was found to have a G20210A prothrombin gene mutation.

The patient was initially placed on a heparin drip as a bridge to Coumadin, and was discharged home in good condition for 6 months of warfarin therapy. Her oral contraception was discontinued and no further investigations are pending. The clinical diagnosis/assumption is that a small embolus, likely from a deep venous thrombosis, passed through the patent foramen ovale and caused the infarction. There are no plans for an operative closure of the patent foramen ovale at this time.

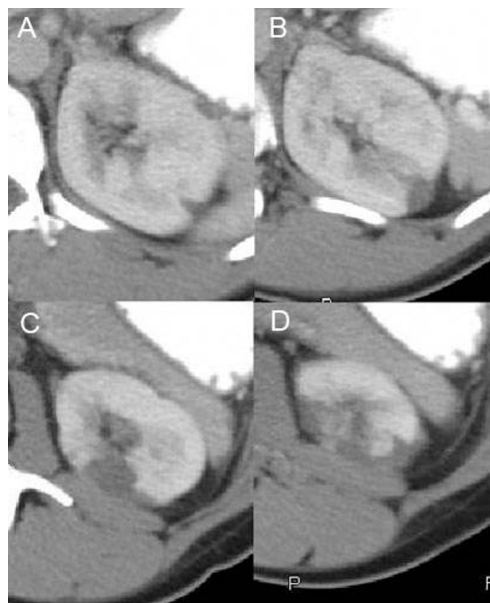


Figure 1. 27-year-old woman with acute renal infarction. (A-D) CT scan with contrast shows multiple perfusion defects in the lower pole of the left kidney.

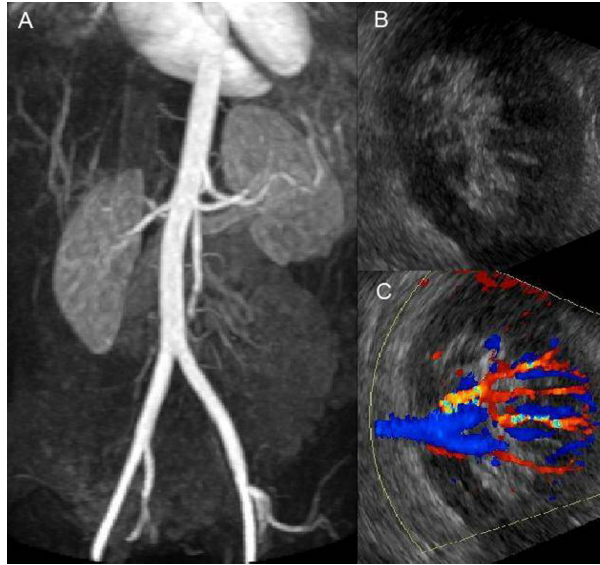


Figure 2. 27-year-old woman with acute renal infarction. (A) Coronal MRI shows perfusion defects in the lower pole of the left kidney. (B) Sonogram shows normal left kidney. (C) Doppler shows no renovascular abnormality.

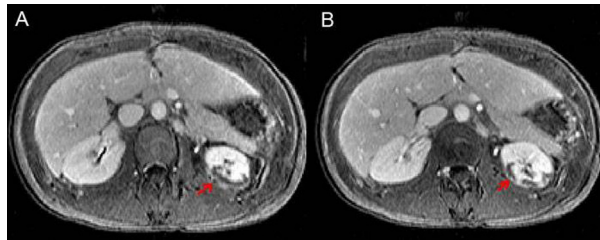


Figure 3. 27-year-old woman with acute renal infarction. (A-B) MRI shows a cortical rim sign (arrow) in the left kidney.



Figure 4. 27-year-old woman with acute renal infarction. (A-B) Angiogram shows ovoid filling defect (arrow) in a segmental branch of the left renal artery in the lower pole, consistent with thrombus.

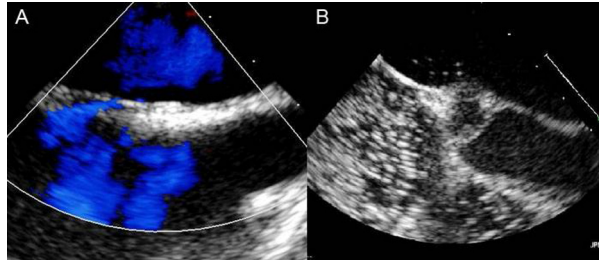


Figure 5. 27-year-old woman with acute renal infarction. (A-B) Transesophageal echocardiogram shows a small patent foramen ovale with bidirectional shunting.

Discussion

Acute renal artery embolism and infarction is relatively uncommon, and rarely initially suspected due to vague and nonspecific clinical findings. The clinical symptoms may include flank pain, abdominal pain, chest pain, nausea, vomiting, and fever [1, 2]. Diagnosis is often delayed, as these findings lead clinicians to first investigate other etiologies such as pyelonephritis, nephrolithiasis, cholecystitis, diverticulitis, lumbago, or myocardial infarction [1, 3]. Laboratory findings may include leukocytosis, elevated lactate dehydrogenase, serum glutamic-oxalacetic transaminase, serum glutamic-pyruvic transaminase, and alkaline phosphatase, as well as hematuria on urinalysis [1, 2, 4]. Elevated serum creatinine and acute renal failure are also possible findings, but not present in all cases [1, 2]. The most sensitive serum indicator for renal infarction is an elevation in serum lactate dehydrogenase, which was found to be present in almost 100% of cases in one reported series [1].

The most common source of renal emboli appears to be the heart, especially in the setting of atrial fibrillation, and the majority of these events occur in patients that are either subtherapeutically or not anticoagulated [5]. In the absence of atrial fibrillation, one case series showed that 50% of patients with noncardiac renal emboli also have a heritable thrombophilia and/or hyperhomocysteinemia [6,7], or another predisposing factor such as connective tissue disease [8]. Noncardiac embolic sources may include emboli from the suprarenal aorta [6], or much less likely a paradoxical embolus from a venous source through a patent foramen ovale or atrial septal defect [9]. Renal artery dissection is a reported but uncommon cause of renal infarction [10], as is compression from a subcapsular hematoma [11], trauma, vasculitis, and shock [12].

Thrombophilia is recognized as a risk factor for developing deep venous thrombosis, and our patient was found to have a G20210A prothrombin gene mutation. Prothrombin has both procoagulant and anticoagulant functions, and an imbalance in these factors can lead to imbalances in hemostasis [13]. The prothrombin gene mutation is the second most common heritable risk factor for thrombosis, and can impart a 3 fold or greater increase in heterozygous carriers, especially in the setting of oral contraceptives [13, 14, 15]. It is important to note that there is a risk of recurrent thromboembolism in these patients, and other risk factors such as smoking and oral contraception should be modified. Alone, the risk of recurrence is modest and lifelong anticoagulation after a single event is not currently recommended [16].

The evaluation of acute abdominal pain typically begins with a CT examination, either with or without intravenous contrast enhancement. To detect a renal infarction, imaging would ideally be performed during the corticomedullary phase [17] although nephrographic phase images can also show the findings. Imaging performed without IV contrast, such as for the evaluation for renal calculi can easily miss the diagnosis of renal infarction. The contrast enhanced CT findings of wedge shaped peripheral perfusion defects are typical of renal infarcts. These can be described as focal or global,

and are considered global if the perfusion defect encompassed more than 50% of the renal cortex [12, 17]. In some cases a “subcapsular rim sign” or “cortical rim sign” may be seen, which involves a smooth 2-4 mm ribbon of enhancing cortical tissue peripherally in an otherwise nonfunctioning kidney [12, 17, 18]. This is thought to represent enhancement of the renal capsule as well as the outer renal cortex through local collateral vessels in the absence of renal artery flow [18]. This sign has been reported in approximately 50% of renal infarctions from all etiologies, and if seen should lead to a strong suspicion of this diagnosis. It is not 100% specific though, as it has also been reported in cases of renal abscess [12].

Although challenging, it is important to rapidly and accurately make the diagnosis of renal infarction since clinical outcomes are typically very good with prompt treatment [19, 20]. Treatment recommendations currently entail anticoagulation, with medically managed patients performing better than those undergoing surgery [19, 20]. Although there is currently no defined role for thrombolysis [5, 21], there have been case reports that show favorable results with endovascular thrombolysis with outcomes comparable to direct surgical intervention [21, 22]. The ischemic tolerance of the kidney is estimated at approximately 90 minutes, and research indicates that there is no role for thrombolytic therapy after this time as it is unlikely to lead to recovery in renal function [23].

We would suggest that in the appropriate clinical setting and suspicious laboratory findings (mild leukocytosis and elevated lactate dehydrogenase), that renal infarction should be considered and weight given to the benefits of a contrast enhanced CT scan examination, even if an unenhanced examination has already been performed. We would also suggest that angiographic confirmation is probably unnecessary and should likely be reserved for equivocal cases. Confirmation does not change management and is of no definite additional therapeutic benefit at this time. Rather, a thorough evaluation for risk factors and an embolic source would more likely be of benefit in this setting.

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