delay. Doppler USG is an effective screening technique and should be employed early. Balloon angioplasty, along with stent implantation, should be encouraged as the preferred choice of management.

TRVS is one of the least suspected etiologies of graft dysfunction, which is potentially reversible if diagnosed without delay. Doppler USG is an effective screening technique and should be employed early in the presence of a diagnostic dilemma. Balloon angioplasty along with stent implantation should be encouraged as the preferred choice of management.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent.

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A Case of Acute Renal Infarct Secondary to Protein S Deficiency

Abstract

Renal infarction is an underdiagnosed and underreported condition with multiple etiologies. A 45-year-old man presented with acute pain in the right lumbar region, CT scan showed a wedge shaped, non-enhancing, hypodense lesion in the cortex of the upper pole of the right kidney- suggestive of infarct. A pro-thrombotic workup revealed a protein S deficiency and a heterozygous mutation for MTHFR gene. Protein S is a vitamin-K dependent plasma glycoprotein, the deficiency of which is associated with a hypercoagulable state, which in turn led to renal infarction in this patient.

Keywords: *Hypercoagulable state, MTHFR mutation, Protein S deficiency, Renal infarction, Thromboembolism*

Introduction

Renal infarction is underreported, with a reported incidence of 0.004% - 0.007%.[1] It presents with flank pain, fever, nausea, vomiting, hematuria, and sudden rise in blood pressure or can mimic pyelonephritis, renal colic, or other diseases. Delayed or missed diagnosis results in irreversible injury to the kidneys. Timely revascularization can restore renal function.

Numerous etiologies lead to the development of renal infarction, but thromboembolism of cardiac origin atheromatous disease are the most frequent. Less common causes identified include hypercoagulable conditions (sickle cell disease, thrombophilia), cocaine misuse, trauma to the kidneys, malignancies, and renal vascular disease (vasculitis, fibromuscular dysplasia, spontaneous renal artery dissection, and dissecting aortic aneurysm). Rare causes include septic emboli in systemic candidiasis or emboli in Takotsubo syndrome. The underlying cause of renal infarction may be unknown despite extensive investigations, and these are labeled idiopathic renal infarcts.[1] Only about 6% of the cases occur due to a hypercoagulable state.[2]

Case Report

A 45-year-old nonsmoker male presented with acute pain in the right lumbar region and fever, without associated chills, rashes, or joint pain. He had no nausea, vomiting,

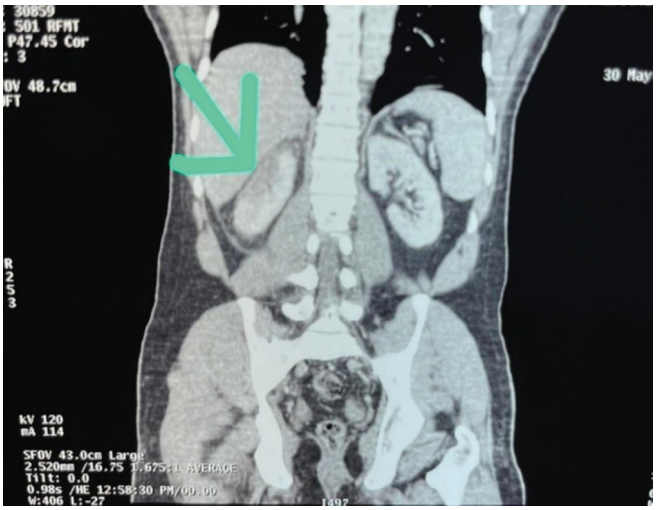

Figure 1: CT abdomen showing infarct in the right kidney. The green arrow marks the wedge-shaped hypodense lesion in the cortex of the upper pole of the right kidney. CT: Computed tomography.

abdominal distension, bowel or urinary symptoms, abdominal trauma, illicit drug use, or alcohol abuse. He gave no past history of renal stones, cardiac disease, or any chronic diseases or surgeries; there was no family history of blood disorders or malignancies.

On examination, he was conscious and oriented and the vital signs were normal. Mild tenderness was noted in the right lumbar region. Chest examination was within normal limits with no cardiac murmurs. The remaining physical evaluation was unremarkable.

Laboratory results showed normal white blood cell (WBC) count, renal function, and urinalysis, with no growth on urine culture. Abdominal ultrasound showed an enlarged fatty liver and prostate. Contrast-enhanced computed tomography (CECT) of the abdomen revealed a wedge-shaped, nonenhancing, hypodense lesion in the cortex of the upper pole of the right kidney, suggestive of infarct [Figure 1]. Renal artery Doppler was normal.

A 12-lead electrocardiogram did not reveal atrial fibrillation, and the transthoracic echocardiogram was normal. A prothrombotic workup showed protein S deficiency and a heterozygous mutation in *MTHFR* gene.

The patient was started on dual antiplatelets and anticoagulants. The pain gradually subsided, and he was discharged with dual antiplatelets and oral anticoagulation. Follow-up revealed no further episodes, and renal function remained normal.

Discussion

Renal infarction is an unusual cause of abdominal pain. Based on autopsy data, a previous study estimated the incidence of renal infarction as 1.4%;[3] however, only a few cases are reported due to the nonspecific presentation. The patients could even be asymptomatic, wherein the diagnosis is only identified incidentally on computed tomography (CT) scan.

WBC counts, C-reactive protein (CRP), alkaline phosphatase, and transaminases are often raised, though not consistently. Raised lactose dehydrogenase (LDH) is sensitive, but not specific.[4] Raised serum creatinine may also be seen. Doppler has low sensitivity for identifying renal ischemia (10%).[5] Renal artery angiography is the gold standard for diagnosis. It typically demonstrates wedge-shaped patches of hypoattenuation within the renal parenchyma, indicating perfusion defect. Etiological investigation requires electrocardiogram (ECG), Holter monitoring, echocardiography, CT angiography, hypercoagulability panel, and autoantibody assay.

A small percentage is caused by hypercoagulable diseases – antiphospholipid syndrome (APS), protein C, S, or factor V Leiden deficits, hyperhomocysteinemia, and polycythemia vera. In this patient, the underlying protein S deficiency was considered the likely cause of renal infarction. Protein S is a vitamin K-dependent plasma glycoprotein, acting as a cofactor for the activated protein C (APC) and tissue factor pathway inhibitor (TFPI) pathways. In addition, it inhibits the intrinsic tenase and prothrombinase complexes, thus having direct anticoagulant actions.[6] Thus, patients deficient in protein S are at risk of developing recurrent thromboembolic events. Here, a heterozygous mutation in *MTHFR* manifested clinically, possibly contributing to the hypercoagulable state.

Treatment aims to restore blood flow to the kidneys early and prevent further embolic episodes. Treatment depends on the underlying etiology and needs to be tailored for each patient. It is mainly based on anticoagulants, and antiplatelets in select patients.

Given the nonspecific presentation, a high index of clinical suspicion should be maintained for early identification of renal infarction and its etiology, to avoid prolonged ischemia and irreversible renal damage.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent.

Conflicts of interest: There are no conflicts of interest.

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Bones, Stones, and Hematuria - Connecting the Dots

Abstract

We report a 12 yr old boy who presented with recurrent gross hematuria, polyuria and rickets with growth failure. Investigations showed bilateral renal calculi with small kidneys on ultrasonography along with hypercalciuria; hypomagnesemia and reduced kidney function. His younger sibling also had nephrocalcinosis hypomagnesemia. The genetic analysis done in view of recurrent renal calculi with chronic kidney disease showed a homozygous missense variant (c.392G>A) at exon 4 of CLDN 16 gene suggestive of Familial hypomagnesemia with hypercalciuria and nephrocalcinosis (FHHNC). The younger sibling had a similar homozygous mutation and the parents were heterozygous carriers.

Keywords: *FHHNC, Hypercalciuria, Hypomagnesemia, Recurrent gross hematuria, Rickets*

Introduction

There are various causes of recurrent kidney stones in pediatric population, such as adenine phosphoribosyltransferase (APRT) deficiency, cystinuria, Dent disease, familial hypomagnesemia with hypercalciuria and nephrocalcinosis (FHHNC), and primary hyperoxaluria (PH).[1] Lack of recognition can result in delays in treatment. Here, we report a 12-year-old boy with recurrent gross hematuria, polyuria, and rickets with growth failure, with a similar history in a younger sibling.

Case Report

A 12-year-old boy, the first born of a nonconsanguineous marriage, presented with a history of recurrent gross painless hematuria since last 4 years. There was a history of nocturia and constipation in the child from early infancy with poor weight and height gain and bowing of the legs. Two years back, the child had sustained a fracture of the distal end of the radius on trivial trauma. His investigations had shown severe hypocalcemia with radiological features of rickets and renal calculi on ultrasound of the kidneys. The younger sibling also had a history of painless gross hematuria.

Examination revealed weight and height below the third centile as per the Indian Academy of Pediatrics (IAP) growth charts.[2] Pallor and bilateral genu valgum were present. His investigations showed deranged renal function tests with hypocalcemia, hypomagnesemia, and hypercalciuria [Table 1]. There was bilateral nephrolithiasis on ultrasound of the kidneys and urinary tract [Figure 1]. The laboratory investigations in the younger sibling showed similar results, except that he had an estimated creatinine clearance of 75 ml/min/m^2.

Suspecting an underlying genetic etiology, whole exome sequencing was ordered, which showed a novel homozygous missense mutant C.392G>A at exon 4 of *CLDN 16* gene, suggestive of FHHNC. Sanger sequencing showed a heterozygous mutation in parents and a homozygous mutation at a similar location in the younger sibling [Figure 2].

Discussion

Children with recurrent stone disease have been reported to have underlying metabolic cause with progression to chronic kidney disease (CKD).[3] Our patient was detected to have FHHNC by whole exome sequencing.

FHHNC, a rare autosomal recessive disease is of two types I and II, which occur due to mutation of *CLDN 16* and *CLDN 19* genes present on chromosomes 3q27 and 1p34.2, respectively. The mutations are responsible for the defective production of claudin proteins 16 and 19, respectively.[4] These proteins that are expressed in the tight junction of the thick ascending limb of the loop of Henle play an important role in the paracellular transport