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# Renal Manifestation in Paroxysmal Nocturnal Hemoglobinuria: A Case Report from Northeast India

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**Abstract**

Paroxysmal Nocturnal Hemoglobinuria (PNH) is an infrequent, acquired hematological disorder characterized by intravascular hemolysis and a spectrum of complications, including renal impairment. A 25-year-old male with established PNH presented with abdominal discomfort, emesis, dark-hued urine, and diminished urine output following substantial alcohol consumption. Laboratory investigations revealed severe anemia accompanied by a pronounced elevation in serum creatinine levels. Despite the implementation of supportive care, renal function deteriorated, necessitating hemodialysis. Renal biopsy demonstrated acute tubular injury with significant hemosiderin deposition. Following treatment with corticosteroids, dialysis, and blood transfusions, renal function was fully restored. This case underscores the peril of severe acute kidney injury in patients with PNH and accentuates the importance of renal biopsy in ambiguous clinical scenarios. It also prompts contemplation regarding the

frequency with which acute kidney injury is overlooked in this patient population.

**Keywords:** Paroxysmal Nocturnal Hemoglobinuria (PNH), Acute Kidney Injury, Hemodialysis

## Introduction

Paroxysmal Nocturnal Hemoglobinuria (PNH) is a rare & acquired hematological disorder caused by mutations affecting the PIGA gene.[1–3] This mutation leads to a deficiency of key surface proteins that protect blood cells from complement-mediated destruction. Patients with PNH experience chronic hemolytic anemia and pancytopenia.[4–6] They are at increased risk for thrombosis. These complications are well described but the impact of PNH on kidney function has received less attention.[2,7]

Renal involvement in PNH can range from mild, subclinical changes to severe acute kidney injury. Intravascular hemolysis releases free hemoglobin and iron - which can deposit in the kidneys and cause tubular injury. Kidney complications in PNH are frequently underrecognized.[2,7–9] This is partly because their presentation may overlap with other causes of renal dysfunction. Abdominal pain, dark urine and a sudden decline in renal function may point to various possible diagnoses. This can make early recognition of PNH-related kidney injury challenging.[2]

This case report delineates the clinical presentation of a young man afflicted with Paroxysmal Nocturnal Hemoglobinuria (PNH) who experienced a profound acute kidney injury subsequent to a hemolytic crisis. The clinical manifestations and histopathological insights underscore the critical necessity of incorporating PNH into the differential diagnosis of idiopathic acute kidney injury, particularly in the presence of hemolysis.

## Case Presentation

### Patient Description

The patient was a 25-year-old male diagnosed with Paroxysmal Nocturnal Hemoglobinuria since 2018. This diagnosis was substantiated by flow cytometry, which revealed the absence of CD55 and CD59 on the surface of the red blood cells. He had undergone multiple blood transfusions in the past.

### Case History

He presented with abdominal discomfort and recurrent emesis subsequent to a period of excessive alcohol consumption. He reported the passage of dark-hued urine and noted a considerable diminution in urine output.

### Physical Examination Results

Upon examination, the patient presented with pallor. Blood pressure was stable. There were no signs of jaundice, edema, or lymphadenopathy. Both cardiovascular and respiratory system assessments yielded unremarkable findings. The abdominal examination indicated mild tenderness, yet no organomegaly was detected.

### Results of Pathological Tests and Other Investigations

Laboratory evaluations conducted upon admission revealed a hemoglobin level of 6.5 g/dL, a platelet count of 130,000, and a total leukocyte count of 5,340. Blood urea was measured at 131 mg/dL, while serum creatinine levels escalated from 3.3 mg/dL to 15.5 mg/dL during the course of hospitalization. Lactate dehydrogenase (LDH) was significantly elevated at 4,976 U/L. Total bilirubin was recorded at 4.09 mg/dL, predominantly in the unconjugated form. Urinalysis indicated 1+ proteinuria, with 1–2 red blood cells (RBCs) and 6–8 white blood cells (WBCs) per high power field. The 24-

hour urinary protein excretion was quantified at 1.23 g. An ultrasound examination revealed a heteroechoic pancreas alongside kidneys of normal size exhibiting a subtle increase in echotexture.

Table 1: Serial Laboratory Results in PNH Case

| Parameter | 24/7/23 | 29/7/23 | 1/8/23 | 4/8/23 |
|---|---|---|---|---|
| Hemoglobin (g/dL) | 6.5 | 10.2 | – | – |
| Total Leukocyte Count (/mm³) | 5,340 | – | – | – |
| Platelet Count (/mm³) | 130,000 | – | – | – |
| Blood Urea (mg/dL) | 131 | 211 | 167 | 102 |
| Serum Creatinine (mg/dL) | 3.3 | 15.5 | 5.8 | 2.1 |
| LDH (U/L) | 4,976 | – | – | – |
| Serum Albumin (g/dL) | 3.7 | – | – | – |
| Amylase (U/L) | 590 | – | – | – |
| Lipase (U/L) | 2,846 | – | – | – |
| Total Bilirubin (mg/dL) | 4.09 | – | – | – |
| Unconjugated Bilirubin (mg/dL) | 3.3 | – | – | – |
| Conjugated Bilirubin (mg/dL) | 0 | – | – | – |
| 24-hr Urinary Protein (g) | 1.23 | – | – | – |

Kidney biopsy revealed the presence of 14 glomeruli, all devoid of global sclerosis. The glomeruli exhibited focal dilatation and congestion of the capillaries.

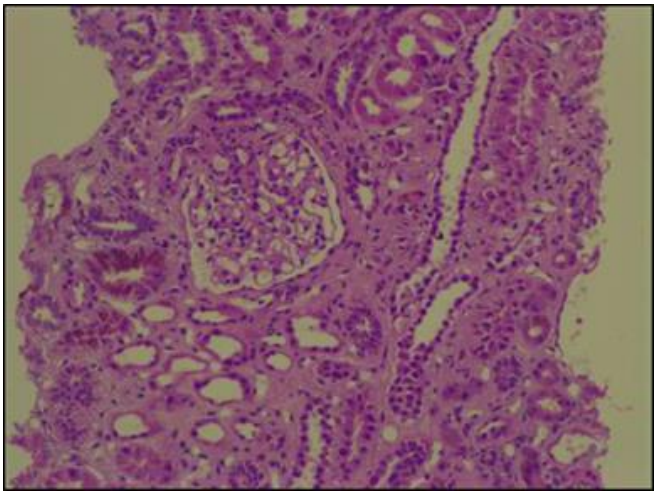

Figure 1: Section of renal cortex from the patient with Paroxysmal Nocturnal Hemoglobinuria, showing acute tubular injury and marked pigment accumulation within tubular epithelial cells. (Haematoxylin and eosin, ×400)

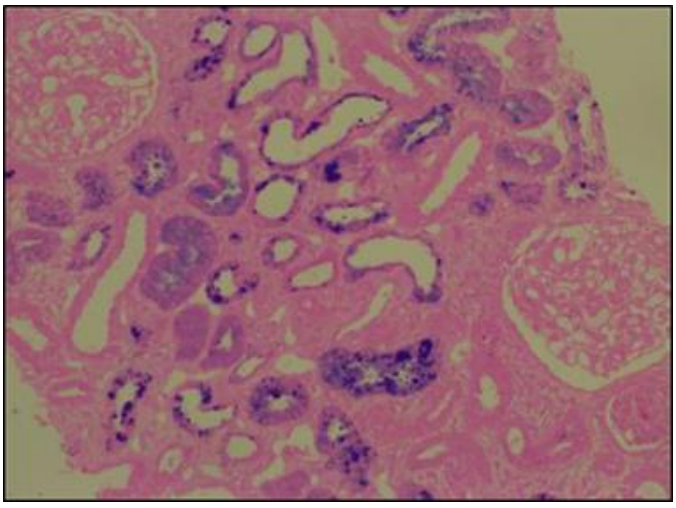

Figure 2: Perls' Prussian blue stain highlights dense hemosiderin granules within renal tubular epithelial cells - a feature of intravascular hemolysis in PNH. Glomeruli are preserved. (×400)

Tubules exhibited pronounced vacuolar alterations, acute cellular injury, and a notable loss of brush borders. Numerous tubules displayed golden-brown pigment that stained blue with Perls stain, thereby confirming the deposition of hemosiderin.

**Treatment Plan**

The patient was initiated on intravenous fluid therapy and subjected to meticulous monitoring. As renal function declined, he underwent five sessions of hemodialysis. Subsequently, he received pulse therapy with methylprednisolone, which was followed by a regimen of oral prednisolone. Additionally, he was administered three units of blood transfusion.

**Expected Outcome of the Treatment Plan**

The objectives of the treatment plan were to (a) stabilize renal function, (b) address the acute hemolytic crisis, and (c) facilitate recovery from acute kidney injury.

**Actual Outcome**

His urine output progressively returned to its normal parameters. Serum creatinine levels exhibited improvement, ultimately reaching 1.6 mg/dL at the time of discharge. Upon follow-up, his renal function returned to baseline values.

**Case Significance**

This case underscores the potential for severe acute kidney injury in patients afflicted with paroxysmal nocturnal hemoglobinuria during a hemolytic crisis. The precipitous decline in renal function, when coupled with distinctive biopsy findings indicative of hemosiderin deposition, contributes a significant clinical observation. The confluence of symptoms such as abdominal pain, emesis, and diminished urine output engendered initial diagnostic ambiguity. This serves as a poignant reminder for clinicians to remain vigilant for PNH-related renal impairment, even when alternative explanations appear plausible upon initial evaluation. Furthermore, this experience raises an imperative inquiry regarding the frequency with which acute kidney injury is overlooked or misdiagnosed in patients with PNH, particularly in instances where renal biopsy is not conducted.

**Discussion**

Paroxysmal Nocturnal Hemoglobinuria is a rare and acquired disorder caused by mutations in the PIGA gene. This leads to a deficiency of proteins that protect blood cells from complement-mediated destruction. Patients can present with hemolytic anemia, pancytopenia and a tendency for thrombosis. Renal involvement is not as widely recognized but can lead to significant complications.

In the case presented herein, the patient experienced a severe acute kidney injury amidst a hemolytic crisis. The precipitous elevation in serum creatinine and the requirement for dialysis underscore the significant renal involvement. Mild chronic renal alterations have been documented in Paroxysmal Nocturnal Hemoglobinuria (PNH). However, the magnitude of acute renal impairment observed in our case was atypical. The patient's manifestation of abdominal pain and vomiting could have indicated a multitude of potential etiologies for acute kidney injury, including dehydration, infection, or even acute pancreatitis, particularly given the noted pancreatic abnormalities on ultrasound. These overlapping clinical presentations possess the potential to obfuscate and prolong the diagnosis of PNH-related renal injury.

Renal biopsy was pivotal in this case. The detection of hemosiderin within tubular cells corroborated that intravascular hemolysis had precipitated substantial iron deposition, thereby contributing to tubular injury. This observation aligns with prior reports indicating that

hemosiderin accumulation is a prevalent histological characteristic in Paroxysmal Nocturnal Hemoglobinuria (PNH), despite the fact that renal biopsies are not invariably performed. The reversibility of renal dysfunction following intervention with hemodialysis and corticosteroids implies that timely treatment can facilitate complete recovery—even in instances where initial creatinine levels are markedly elevated.

This case illuminated a complication that is likely to be underdiagnosed in Paroxysmal Nocturnal Hemoglobinuria (PNH). Acute kidney injury during hemolytic episodes may be more prevalent than the literature indicates, as the majority of cases are managed conservatively without a definitive tissue diagnosis. The clinical manifestations can be nonspecific, and in the absence of a heightened index of suspicion, clinicians may erroneously attribute the renal findings to alternative etiologies. The identification of hemosiderin within the renal tissue substantiated the correlation between intravascular hemolysis and acute renal impairment in these patients.

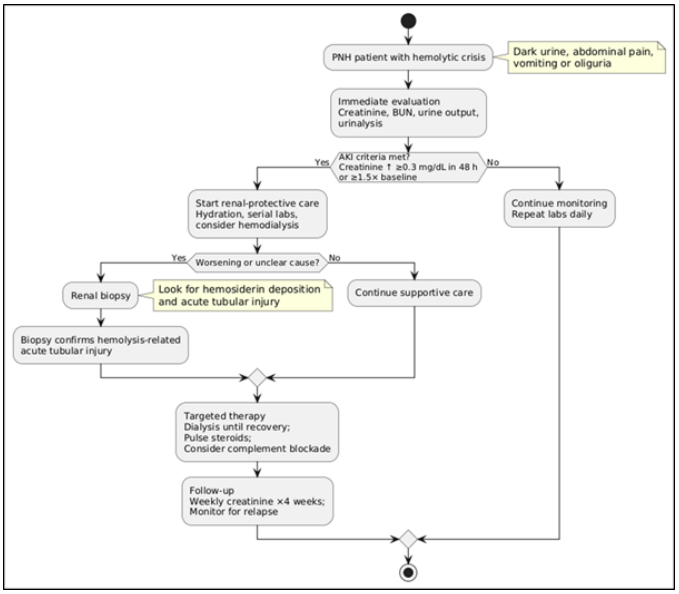


Figure 3: Management of Acute Kidney Injury in PNH Haemolytic Crisis

Given the risk of recurrent hemolysis in Paroxysmal Nocturnal Haemoglobinuria (PNH), it is imperative to meticulously monitor renal function and to contemplate kidney biopsy in instances of unexplained or severe renal impairment. Furthermore, there exists a pressing necessity for heightened awareness of PNH-associated renal injury among healthcare practitioners. The early identification and supportive management of this condition can significantly enhance patient outcomes.

Figure 3 delineates the management protocol that is advocated in such scenarios.

This case also prompts a critical examination of the broader acknowledgment of acute kidney injury (AKI) in patients with Paroxysmal Nocturnal Hemoglobinuria (PNH). There exists a notable lack of consensus regarding the optimal strategies for monitoring and managing renal complications in this patient population. A more comprehensive body of data is essential to elucidate the true incidence, risk factors, and long-term ramifications of AKI in the context of PNH. Furthermore, additional investigative efforts are warranted to explore the efficacy of disease-specific interventions, such as complement inhibitors.

Published evidence regarding acute kidney injury (AKI) in Paroxysmal Nocturnal Hemoglobinuria (PNH) remains limited. Nevertheless, an accumulating body of clinical observations offers deeper insights into its manifestation, renal pathology, and therapeutic responses.

Choi et al. (2023) described a young woman who developed severe pigment-induced AKI that required five sessions of hemodialysis; heavy tubular haemosiderin was confirmed on biopsy, and renal function returned to baseline once ravulizumab was introduced.[10] A trio of cases collated by Puri, Gandhi and

Sharma (2017) expanded the morphological spectrum: each biopsy showed moderate-to-marked siderosis, two patients needed dialysis while the third improved with conservative measures. All patients left the hospital with creatinine concentrations lower than admission values.[11] The larger series assembled by Ram and colleagues (2017) strengthened these observations; six of fourteen individuals with PNH experienced AKI, four underwent biopsy that showed acute tubular necrosis driven by hemosiderin and every one of the five patients dialyzed regained full renal function within three months.[9]

Single-patient reports echo the pattern of brisk renal deterioration followed by recovery once hemolysis is controlled. Patel and Hota (2019) documented a 50-year-old man whose dialysis-dependent AKI improved after corticosteroids and transfusion support.[8] From earlier reports, Chen et al. (2007) and Satish & Rajesh (2010) each outlined recurrent hemoglobinuria AKI with biopsy-proven pigment nephropathy; repeated episodes settled with timely supportive care and did not progress to chronic kidney disease.[12]

Complement blockade featured increasingly in recent accounts - Mauro and Gherlinzoni (2020) moved a patient from long-term steroids to eculizumab after years of recurring AKI and documented complete absence of further renal insults.[13] Ranade and co-workers (2024) reported two instances of pigment nephropathy that resolved after short-term dialysis.[14]

Across these studies, several salient themes emerge. (1) Profound hemosiderin deposition observed in biopsy specimens manifests as a consistent histological hallmark, corroborating a direct toxic effect of filtered haem-iron on tubular epithelium. (2) Timely hemodialysis preserves metabolic stability without hindering renal recovery. (3) Complement inhibition, utilizing agents such as eculizumab, ravulizumab, or emerging therapeutics, can mitigate further hemolysis, abbreviate the duration of acute kidney injury (AKI), and safeguard long-term renal function.

Our patient's clinical presentation, biopsy results, and subsequent outcomes closely align with the prevailing literature. This collective experience underscores the significance of (A) proactive creatinine monitoring during hemolytic crises, (B) a low threshold for renal biopsy when the etiology of acute kidney injury remains ambiguous, and (C) the prompt initiation of complement blockade to avert recurrent renal damage.

**Conclusion**

AKI in PNH is likely to be inadequately explored and insufficiently recognized; prompt and protocol-driven interventions can reverse even severe renal failure, as elucidated in this case report. Timely dialysis, coupled with targeted therapy, facilitated a reduction in creatinine levels to baseline, allowing for the patient's complete recovery. Incorporating our proposed decision-making framework can assist frontline teams in identifying and addressing PNH-related AKI before irreversible damage ensues.

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