

# Renal hemosiderosis in paroxysmal nocturnal hemoglobinuria

## Hemossiderose renal em hemoglobinúria paroxística noturna

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### ABSTRACT

A previously healthy 38-year-old man presented with history of fever, adynamia and anemia for the last 5 months. Physical examination revealed pallor. Laboratory was marked by a hemoglobin level of 9.2g per deciliter (reference range, 14.0 to 18.0 g per deciliter), and a platelet count of 50,000 cells per cubic millimeter (reference range, 140,000 to 400,000 cells per cubic millimeter). There was no iron or vitamin B12 deficiency. Reticulocyte percentage was 3.9% (reference range 0.5 to 1.5%). Lactate dehydrogenase (LDH) was 2293 U/L (reference range, 230 to 460 u/L), total bilirubin was 1.98 mg/dL (reference <1 mg/dL), with predominance of indirect fraction, which was 1.36 mg/dL (reference < 0.8 mg/dL). The direct antiglobulin test was negative, and serum protein electrophoresis was unremarkable. After admission, the patient developed persistent abdominal pain. An abdominal MRI (Magnetic Resonance Imaging) was performed and revealed segmental portal vein thrombosis (Figure 1) and the kidneys exhibited abnormally low renal cortical signal in T2-weighted sequences (Figure 2). This particular radiologic finding results from accumulation of hemosiderin in the kidneys and is typically seen in chronic intravascular hemolytic situations. His renal function was normal at admission, with a serum creatinine of 0.9mg per deciliter (reference range 0.7 to 1.3 mg per deciliter) and the urinalysis has shown hemoglobin 1+ without red cells. A flow cytometry confirmed the diagnosis of paroxysmal nocturnal hemoglobinuria (PNH).

Iron deposition in the kidneys is typically seen in cases of intravascular hemolysis. Hemosiderin is deposited in the proximal convoluted tubules, promoting an inversion of the signal intensity of the renal cortex on MR. This finding is seen as an accentuated decrease in the cortical signal intensity on T2-weighted images. Interestingly, this accumulation does not seem to affect renal function as seen in this patient.

PNH is an acquired clonal haematopoietic stem cell disorder characterized by cytopenias of any lineage, episodes of intravascular haemolysis and prothrombotic state. These manifestations are due to an increased susceptibility of the blood cells membrane to lysis activity of the complement system. Most of the morbidity and mortality of PNH is due to thrombosis, although its mechanism is poorly understood.

**Keywords:** Hemoglobinuria. Paroxysmal. Hemolysis. Hemosiderosis.

**Figure 1.** This MRI image reveals portal vein segment filling defect, finding of thrombosis.



**Figure 2.** This T2-weighted MRI sequence exhibit an abnormally low renal cortical signal, which results from accumulation of hemosiderin in the kidneys and is typically seen in chronic intravascular hemolytic situations.



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## REFERENCES

1. Jeong JY, Kim SH, Lee HJ, Sim JS. Atypical low-signal-intensity renal parenchyma: causes and patterns. *Radiographics*. 2002;22(4):833-46.
2. Queiroz-Andrade M, Blasbalg R, Ortega CD, Rodstein MA, Baroni RH, Rocha MS, et al. MR imaging findings of iron overload. *Radiographics*. 2009;29(6):1575-89.
3. Hill A, DeZern AE, Kinoshita T, Brodsky RA. Paroxysmal nocturnal haemoglobinuria. *Nat Rev Dis Primers*. 2017;3:17028.

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