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Images of Interest / Imagens de Interesse

Renal Hemosiderosis: A Typical Magnetic Resonance Imaging Pattern

Hemosiderose Renal: Um Padrão Típico em Ressonância Magnética

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Abstract

Paroxysmal Nocturnal Hemoglubinuria is a rare acquired myelodysplastic disorder characterized by increased complement-mediated intravascular hemolysis. Deposition of hemosiderin in the proximal tubules of the renal cortex follows intravascular hemolysis. MRI is the best imaging technique to depict hemosiderin deposits, demonstrating the typical signal pattern of renal hemosiderosis. We present a case of renal hemosiderosis in the context of Paroxysmal Nocturnal Hemoglubinuria, discuss its typical findings at MRI, their underlying pathogenesis, and some differential diagnosis.

Keywords

Paroxysmal nocturnal hemoglobinuria; Magnetic resonance imaging; Hemosiderosis.

Resumo

A hemoglubinúria paroxistica noturna é uma patologia mielodisplásica adquirida que se caracteriza por aumento da hemólise intravascular mediada pelo complemento. Da hemólise intravascular resulta deposição de hemossiderina nos túbulos proximais localizados no córtex renal. A ressonância magnética é a melhor técnica de imagem para detetar depósitos de hemossiderina, revelando o padrão típico de sinal da hemossiderose renal. Neste artigo apresentamos um caso de hemossiderose renal no contexto de Hemoglubinúria Paroxistica Noturna, discutindo os seus achados imagiológicos típicos em RM, a sua etiologia e patogénese e eventuais diagnósticos diferenciais.

Palavras-chave

Hemoglubinuria paroxistica noturna; Imagem por ressonância magnética; Hemosiderose.

Case Presentation

A 70-year-old female presented with high indirect bilirubin levels and mild renal dysfunction on a routine assessment. Relevant past medical history included mild chronic anemia and episodes of hematuria in the previous year, which were treated as low urinary tract infections.

Additional workup revealed increased LDH and reticulocyte count, hematuria and low haptoglobin levels, thus suggesting hemolytic anemia.

An abdominal MRI was also performed, revealing an abnormal bilateral renal cortex hypointensity, both on T1W and T2W, as well as cortical thinning (Fig.1 and Fig.2), with a relative signal increase on T1 out-of-phase compared to T1 in-phase (Fig.3). No other abdominal abnormalities were noticed, including in the renal medulla. Isolated renal hemosiderosis was therefore assumed.

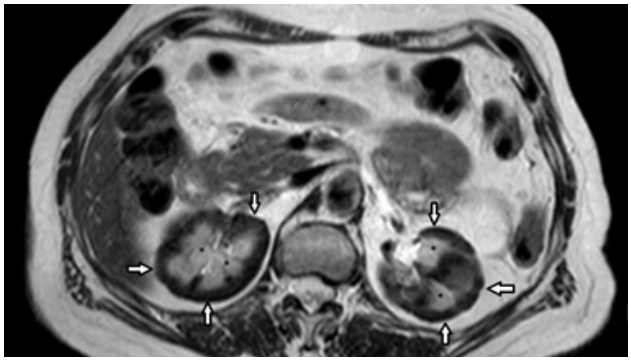

Figure 1 – Axial T2FSE. Abnormally diffuse hypointense renal cortex (arrows) and normal medulla signal (*).

Further analytical evaluation excluded autoimmune and microangiopathic hemolytic anemias. Finally, a screening confirmed the presence of the Paroxysmal Nocturnal Hemoglobinuria (PNH) clone.

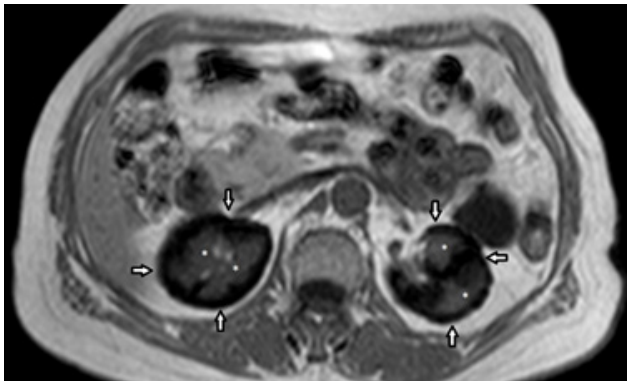

Figure 2 – Axial T1GRE (in-phase). Reversed T1 renal cortex-medulla differentiation with abnormal T1 hypointense renal cortex (arrows) and normal medullar intensity (*).

Discussion

PNH is a rare acquired myelodysplastic disorder characterized by increased complement-mediated intravascular hemolysis.[1] Clinical features of PNH include hemoglobinuria, iron deficiency anemia and venous thrombosis.[1,2] Hemosiderin accumulation resulting from intravascular hemolysis occurs exclusively in the kidney and is frequent in PNH.[1,3] Chronic renal insufficiency secondary to PNH is most likely due to microvascular thrombosis, rather than iron/hemosiderin-induced tubular damage.[2]

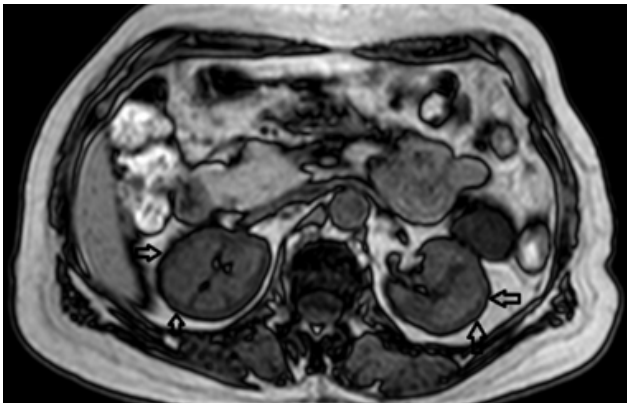

Figure 3 – Axial T1GRE (out-of-phase). Diffuse cortical hypointensity (arrows) on T1 out-of-phase. Note that on T1 GRE out-of-phase the renal cortical intensity is relatively less dark than on T1 GRE in-phase (Fig.2).

The intravascular hemolytic process releases free hemoglobin into the plasma that either attaches to haptoglobin or remains unbound. The unbound plasmatic hemoglobin is filtered in the glomerulus and reabsorbed at the renal cortical proximal tubules, leading to the deposit of heme iron as hemosiderin in the renal cortex. If the renal tubular absorptive capacity is exceeded, the filtered hemoglobin is excreted as hemoglobinuria.[2]

MRI is the most sensitive imaging method to detect hemosiderin deposits.

In normal kidneys, the cortex appears more intense than the medulla on T1W MRI images, and both the cortex and medulla have high signal intensity on T2W.3 (Fig. 4) In renal hemosiderosis, the renal cortex appears dark on T1W and T2W and the medulla retains its normal signal, resulting in T1 reversed cortex-medulla differentiation;1 on T1 GRE out-of-phase the renal cortical intensity is relatively higher than on T1 GRE in-phase.

Because of its superparamagnetic properties, high concentrations of hemosiderin dramatically reduce T2 relaxation time and decrease signal in both T1W and T2W.1 Gradient-echo imaging is also sensitive to T2 signal loss because it lacks the 180º refocusing pulse of the spin-echo sequences.[2,3] On gradient-echo sequences, the longer the echo time the greater the T2 decay, which explains the differences between T1 in-phase and T1 out-phase.[4]

Although renal hemosiderosis is characteristic of PNH, it has also been reported in other hemolytic anemias.[2,3] However, opposed to what occurs in most hemolytic anemias (e.g., sickle cell disease, autoimmune hemolytic anemia, hereditary spherocytosis), in PNH the reticuloendothelial system

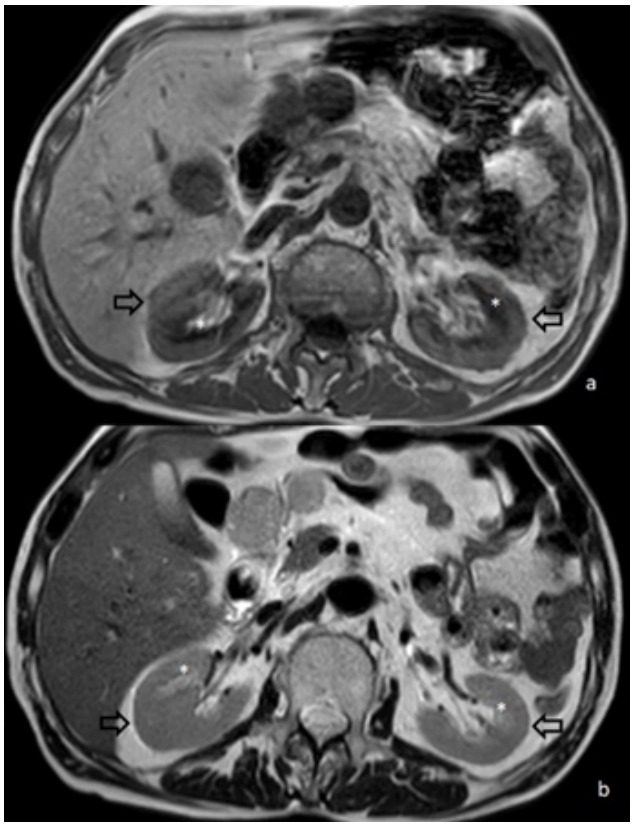

Figure 4 – Normal kidney signal at MRI (another patient from the hospital database). T1W image (a) displays normal cortex-medulla differentiation, with the cortex (arrows) appearing more intense than the medulla (*). T2W (b) depicts both medulla (*) and cortex (arrows) normal high signal.

contributes little to the red cell destruction. Therefore, the signal intensity in the liver and the spleen is generally normal in PNH, unless transfusion siderosis or vascular thrombosis have occurred.[2]

Isolated renal hemosiderosis without reticuloendothelial iron overload is very suggestive of PNH and mechanical valvular hemolysis is a differential diagnosis to be considered.[3]

PNH is a chronic, progressive, and life-threatening disease. About 4 or 5 decades ago the 10-year survival after diagnosis was only 50 percent, with thrombosis as the leading cause of death.[5] Presently the treatment options for PNH include supportive care (RBCs transfusion, immunosuppressive treatment, etc.), allogeneic stem cell transplantation or eculizumab (a complement blockade antibody). The advances in treatment in the last decades (i.e., eculizumab) have significantly improved long term survival in PNH.[6]

Ethical Disclosures / Divulgações Éticas

Conflicts of interest: The authors have no conflicts of interest to declare.

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Confidentiality of data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.

Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.

Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Protecção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

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