Renal haemosiderosis: A cause for ‘black’ kidneys on MRI

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Section: Abdominal imaging
Area of Interest: Abdomen Kidney
Imaging Technique: MR
Case Type: Clinical Cases
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Patient: 37 years, male

Clinical History:

A 37-year-old man with a history of chronic haemolytic anaemia and haematuria was electively admitted to the medicine department for endoscopic band ligation of oesophageal varices. An abdominal magnetic resonance imaging (MRI) examination was requested. Non-enhanced computed tomography (CT) was also performed for additional clarification of MRI findings.

Imaging Findings:

The patient had a dysmorphic liver with hypertrophy of the left lobe, caudate lobe and segment IV, and atrophy of right lobe, with regular surface contours. This appearance suggests a vascular liver disease rather than a cirrhotic liver (Fig. 1). The main portal vein and the spleno-portal confluent were permeable. Nevertheless, there was a filiform hypointense thrombus within the portal vein bifurcation that was calcified on the CT image, suggesting chronic portal vein thrombosis (PVT) (Fig. 2). Exuberant splenomegaly with hypointense Gamma-Gandy bodies was evident owing to portal hypertension (Fig. 1 and Fig. 3A). A peripheral wedge-shaped T2-weighted hypointense line was noted, associated with capsule retraction, related with previous splenic infarction (Fig. 3B). The kidneys had normal morphology and size but demonstrated marked low signal intensity of the renal cortex on T1-weighted in-phase gradient-echo and T2-weighted images and normal signal intensity of the medullary area (Fig. 3), suggesting iron deposition in the renal cortex.

Discussion:

Portal vein thrombosis (PVT) can be caused by diverse local or systemic pro-thrombotic states[1]. Chronic PVT is usually asymptomatic but may present as portal hypertension with digestive bleeding[2]. The liver is dysmorphic, with hypertrophy of caudate lobe and atrophy of right lobe. The presence of smooth contours and normal or enlarged segment IV helps to distinguish from liver cirrhosis. Anomalous parenchymal hyper-enhancement is usually observed on arterial phase followed by homogenous enhancement on portal venous phase.

The low signal intensity of the cortical kidneys on T1 and T2-weighted images is suggestive of iron deposition in the renal cortex. Haemolytic conditions, such as paroxysmal nocturnal haemoglobinuria (PNH), sickle-cell disease and mechanical haemolysis from a dysfunction prosthetic cardiac valve are the main diagnosis associated with this radiologic finding [3]. Unlike other haemolytic anaemias, in PNH the levels of iron deposits in the liver and spleen are usually normal. Nevertheless, if PNH patients need several blood transfusions, the iron levels may increase, and the liver and spleen may show decreased signal intensity on MRI due to haemosiderosis[4].
In our case, the presence of chronic haemolytic anaemia, haematuria and imaging signs of iron deposition in the renal cortex suggested the diagnosis of PNH, which is a prothrombotic condition that may lead to PVT. Further laboratory investigation was made and the negativity of Coomb’s test against the positivity of Ham test confirmed the diagnosis.

PNH is a rare acquired myelodysplastic stem-cell disorder characterised by acute or chronic intravascular haemolysis due to an increased sensitivity to complement-mediated haemolysis[5], which may induce chronic renal failure [6]. Due to repeated episodes of microvascular thrombosis in the renal cortex and haemoglobin saturation, free haemoglobin is filtered by the glomeruli and partially reabsorbed in the proximal tubules. There, some iron is incorporated into haemosiderin deposits and the rest is excreted as haemoglobinuria. The typical histological finding is deposition of haemosiderin in the proximal convoluted tubules of the renal cortex[7]. As the renal medulla is not affected in PNH, it has normal intensity on MRI. The T1-weighted gradient-echo images are the best pulse sequence to demonstrate iron deposition once they are more sensitive to the magnetic susceptibility effect than spin-echo sequences [8].

Teaching Points:

- MRI is the best imaging technique to demonstrate haemosiderin deposition in the renal cortex
- PNH should be considered in the face of “dark” renal cortex and signs of venous thrombosis

Differential Diagnosis List: Renal haemosiderosis due to paroxysmal nocturnal haemoglobinuria and chronic non-cirrhotic portal vein thrombosis, Renal haemosiderosis due to paroxysmal nocturnal haemoglobinuria and liver cirrhosis, Renal cortical necrosis and liver cirrhosis, Renal haemosiderosis related to sickle-cell anaemia and liver cirrhosis

Final Diagnosis: Renal haemosiderosis due to paroxysmal nocturnal haemoglobinuria and chronic non-cirrhotic portal vein thrombosis

References:

Figure 1

Description: (A) Coronal and (B) axial T2-weighted images show a dysmorphic liver, with regular contours, hypertrophy of the caudate lobe and atrophy of the central and peripheral areas (“atrophy-hypertrophy complex”) due to chronic PV thrombosis. Massive splenomegaly is also noticed. 

Origin: Department of Radiology, Centro Hospitalar e Universitário do Porto, Porto, Portugal
Description: (A) Coronal and (B) axial T2-weighted images show a dysmorphic liver, with regular contours, hypertrophy of the caudate lobe and atrophy of the central and peripheral areas (“atrophy-hypertrophy complex”) due to chronic PV thrombosis. Massive splenomegaly is also noticed. Origin: Department of Radiology, Centro Hospitalar e Universitário do Porto, Porto, Portugal
Description: (A) Axial post-contrast T1-weighted image and (B) axial non-enhanced CT image show a filiform thrombus in the portal vein bifurcation (arrow), proved to be calcified on CT image, indicating partial chronic PV thrombosis. Origin: Department of Radiology, Centro Hospitalar e Universitário do Porto, Porto, Portugal
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Description: (A) Axial in-phase gradient-echo T1-weighted image shows splenomegaly with multiple small siderotic nodules (arrows) known as gamma-gandy bodies, related to portal hypertension. (B) Coronal T2-weighted image demonstrates a notch and peripheric hypointense wedge-shaped splenic focus (arrow) due to spleen infarction. In both images (A and B), it is also evident the marked low signal intensity of the renal cortex compared with the medullary zone that maintains the normal intermediate signal intensity. This characteristic radiological finding is due to the iron deposition in the cortical kidney.

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