**TITLE PAGE**

**Image of The Month: Gastroenterology**

**Title: A 41 year old man with abdominal pain, deranged liver function, and fevers**

**Authors:**

**Dr James B Maurice**

Imperial College London

[james.maurice@](mailto:jamesmaurice@nhs.net)imperial.ac.uk; 07793713773;

**Dr Dominic Yu**

Royal Free Hospital, Hepatology, Pond St, London UK, NW3 2QG

**Dr Jennifer Watkins**

Royal Free Hospital, Histopathology, Pond St, London UK, NW3 2QG

**Grant support:** Nil

**Disclosures:** nothing to disclose

**Author contributions:**

**JBM:** wrote the manuscript

**DY:** provided the radiological images and approved final manuscript

**JW:** wrote the histology section of the manuscript, provided histology image, and approved final manuscript.

A 41-year-old man presented with sudden onset upper abdominal pain and fevers. His only past medical history was antiphospholipid syndrome, diagnosed after two deep vein thromboses, and discoid lupus. He had been treated with warfarin for over 10 years, but took no other medications.

On examination he had a soft but tender abdomen in the right upper quadrant. Admission blood tests revealed a bilirubin 29µM/L, alanine transaminase 36U/L, alkaline phosphatase 225U/L, creatinine 61µM/L, C-reactive protein (CRP) 314mg/L, white cell count 8x109/L and INR 6. He was transferred to the hepatology team with a provisional diagnosis of liver abscesses. Abdominal CT and liver MRI scans appeared to confirm this showing multiple low attenuation lesions, but also revealed abnormal hepatic venous anatomy with a strictured inferior vena cava; blood circumnavigated the stricture via collateral circulation from the middle hepatic vein to the right hepatic vein (Figure 1, arrow) and then into the suprahepatic inferior vena cava. There was no acute thrombus, but these collateral vessels represented possible previous hepatic venous thrombosis and chronic veno-occlusive liver disease.

MRCP showed a normal biliary tree. Despite intravenous antibiotics he continued to have fevers, right upper quadrant pain and a raised CRP. PET CT showed diffuse uptake throughout the liver (figure 2). A liver biopsy was performed (figure 3).

What does the liver biopsy show? What is the differential diagnosis for this man’s liver disease?

There is striking fibrinoid type necrosis of periportal hepatocytes (Figure 3, arrow) with associated neutrophilic inflammation. No vasculitis or thrombi are seen. The portal tracts appear oedematous and contain only scattered inflammatory cells. The was mild parenchymal steatosis.

Periportal necrosis is unusual and the differential diagnosis narrow, including eclampsia, disseminated intravascular coagulation, toxins (phosphorus, ferrous sulphate, cocaine ), systemic lupus erythematosus with antiphosphlipid syndrome and hepatic lymphomas.

Shortly after the biopsy he developed an acute kidney injury with proteinuria, followed by left side weakness, breathlessness and hypoxia. Imaging confirmed cerebral infarcts and pulmonary haemorrhage.

He was diagnosed with catastrophic antiphospholipid syndrome.

Antiphospholipid syndrome is characterised by the presence of antiphospholipid antibodies in conjunction with the clinical manifestations of venous or arterial thrombosis, or pregnancy morbidity. It is commonly associated with other systemic inflammatory diseases such as systemic lupus erythematosus. Rarely, antiphospholipid syndrome can present with rapidly evolving multiorgan failure known as *Catastrophic Antiphospholipid Syndrome*.[[1]](#endnote-1)

He was managed with plasma exchange, followed by rituximab. His liver and renal function improved, and was discharged with a mild resolving hemiparesis.

**Word Count: 432**

1. Asherson, R., Cervera, R., de Groot, P. *et al*. Catastrophic antiphospholipid syndrome: international consensus statement on classification criteria and treatment guidelines. *Lupus* 2003; 12: 530–534. [↑](#endnote-ref-1)