EDITOR'S QUIZ: GI SNAPSHOT

Unexplained liver dysfunction in a young Chinese man with eosinophilia

CLINICAL PRESENTATION

A 37-year-old man complained of a 7 months' history of yellowish skin and sclera accompanied by fatigue, pruritus and dry mouth, with a history of subcutaneous masses on his elbow. Hard and fixed subcutaneous masses were noted on his palm and periauricular region, the largest of which was approximately 2 cm in diameter (figure 1A). Mildly tortuous veins on his abdominal wall above the umbilicus were detected (figure 1B). The patient had no history of liver disease. Family history was not contributory. Laboratory tests demonstrated a leucocyte count of 2.9×109/L (3.97–9.15×109/L), 7.9% eosinophil (0.5%–5%), a total bilirubin of 94.5μmol/L, gamma-glutamyl transpeptidase of 67.8 U/L (11–50 U/L) and serum IgG of 23.3 g/L (7–16 g/L), IgG4 of 3.48 g/L (0.08–1.4 g/L) and IgE of 256 IU/mL (0–100 IU/mL). Coagulation function tests revealed prothrombin time of 14.8 s (9.4–12.5 s), international normalised ratio (INR) of 1.33 and D-dimer of 0.14 mg/L (< 0.5 mg/L). Other laboratory tests included a normal blood urea nitrogen and creatinine; perinuclear anti-neutrophil cytoplasmic antibody (pANCA) and anticardiolipin antibody (ACA) were negative. Contrast-enhanced CT of the abdomen revealed diminished endoluminal enhancement of the right and middle hepatic veins with patchy alterations in the parenchymal perfusion, as well as signs of portal hypertension (figure 1C,D).

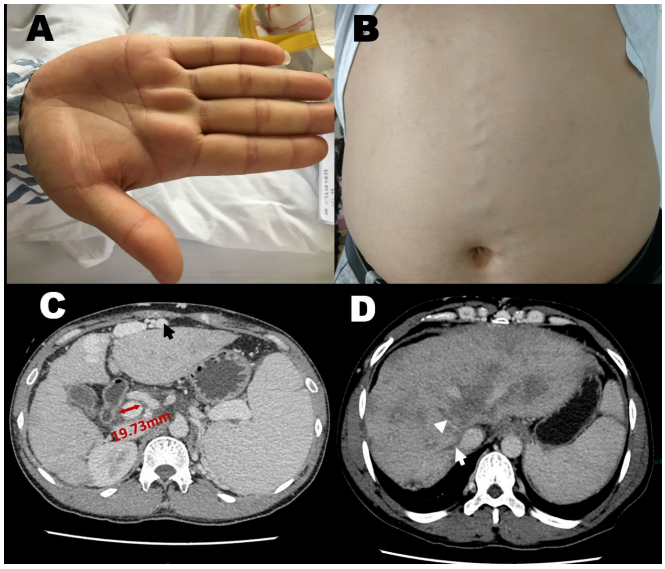

Figure 1 Clinical and radiological findings: (A) palmar subcutaneous nodules; (B) caput medusa; (C) contrast CT showing splenomegaly, dilated portal vein (19.73 mm) and formation of collateral circulation (black arrow: periumbilical vein); (D) diminished endoluminal enhancement in the right and middle hepatic veins with patchy alterations in the parenchymal perfusion (white arrow: right hepatic vein, white arrowhead: middle hepatic vein).

QUESTION

What is the possible diagnosis?

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ANSWER

Biopsy of the subcutaneous masses revealed typical histological features of Kimura disease (KD) with marked proliferation of lymphocytes and eosinophilic abscess; proliferated vessels were lined with hobnailed endothelial cells surrounded by abundant eosinophils (figure 2A,B). A liver biopsy demonstrated marked sinusoidal dilatation and congestion accompanied by hepatocytes. Diffuse perisinusoidal fibrosis and obliteration of hepatic veins were found. Vessels were short shunted from fibrous septa and proliferated into the hepatic parenchyma. Mild inflammation and IgG4-positive plasma cells were occasionally noted (<10/high power field) (figure 2C–F).

The patient was diagnosed with Budd-Chiari syndrome (BCS) associated with KD. KD, clinically characterised by a triad of painless subcutaneous masses, elevated serum IgE and peripheral eosinophilia, can cause hypercoagulation and thrombosis.[1 2] In this case, the BCS is likely related to the hypercoagulable status of KD. Although serum IgG and IgG4 were increased, immunohistochemical staining disclosed only a few IgG4-positive plasma cells in the liver, and the ratio of IgG4 to IgG plasma cells was no more than 40%, which ruled out the IgG4-related hepatobiliary disease.[3]

Our patient was treated with immunosuppressive drugs for specific treatment of KD and also anticoagulant therapy (low molecular weight heparin, 0.4 mL/d) for BCS. Both serum IgE and eosinophils returned to normal range during follow-up.

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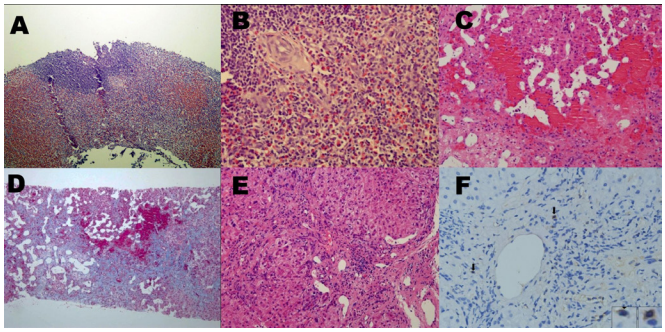

Figure 2 Histopathological findings: (A) a biopsy of the right elbow mass demonstrated marked proliferation of lymphocytes with eosinophilic abscess (H&E ×100); (B) proliferated thin-walled capillaries, infiltrated with numerous eosinophils, plasma cell and lymphocytes were lined with either flattened or low cuboidal endothelium (H&E ×400); (C) sinusoidal dilation and congestion with atrophy of hepatocytes (H&E ×200); (D) diffuse perisinusoidal fibrosis and obliteration of hepatic veins (trichrome staining ×100); (E) vessels were short shunted from fibrous septa and proliferated into hepatic parenchyma (H&E ×200); (F) IgG4-positive plasma cells are occasionally noted (arrows, IgG4 immunohistochemistry staining ×400).

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