Idiopathic non-cirrhotic portal hypertension and portal vein thrombosis in a patient with HIV infection☆

Hipertensión portal idiopática no cirrótica y trombosis portal en paciente con infección por VIH

Idiopathic non-cirrhotic portal hypertension (INCPH) is a rare condition that involves intrahepatic portal hypertension without cirrhosis or other causes of liver disease.[1,2] It requires the presence of portal hypertension (PHT) with permeability of the splenoportal axis and suprahepatic veins. The aetiopathogenesis of this entity is unknown[3] but is associated with infections, toxins, immunological disorders, hypercoagulation and human immunodeficiency virus (HIV).

We present the case of a 39-year-old male with a pre-existing diagnosis of stage C3 HIV infection who attended A&E having presented melaena for the previous three days. He had not taken NSAIDs or other gastro-damaging agents. He presented asthenia of recent onset with no abdominal pain. He was on antiretroviral therapy, having received various drugs since 1999 (including didanosine for 42 months). At the physical examination, the patient presented pale skin and mucosa, tachycardia at 110 bpm and blood pressure of 130/90 mmHg. The abdominal examination revealed non-painful hepatosplenomegaly.

The complete blood count found haemoglobin levels of 6.4 g/dl (normal range: 13–18 g/dl) and a normal platelet count (168,000 U/ml). The clinical chemistry found bilirubin levels of 0.8 mg/dl, albumin 4 g/dl, aspartate aminotransferase 38 U/l, alanine aminotransferase 25 U/l and alkaline phosphatase 98 U/l. The coagulation study found an INR of 1. The HIV viral load was undetectable with a CD4 count of 188 cell/μl. Complementary examinations conducted included an early upper gastrointestinal endoscopy, which revealed large oesophageal varices with no signs of bleeding or recent haemostasis. Three packed red blood cell units were transfused and the patient was admitted for examination. Bloods were conducted that included: viral serologies, autoimmunity study, ferrokinetic studies, copper, ceruloplasmin, α-1 antitrypsin and porphyria, and all results were normal. The abdominal Doppler ultrasound and chest/abdominal CT scan revealed signs of PHT (hepatomegaly, splenomegaly, portal vein dilatation and significant collateral circulation). The permeability of the splenic-mesenteric-portal venous system was confirmed. A liver elastography was performed, yielding a result of 13.8 kPa. The haemodynamic study revealed a hepatic venous pressure gradient (HVPG) of 10 mmHg. A transjugular liver biopsy was performed during the procedure, with insufficient material to conduct an anatomopathological study.

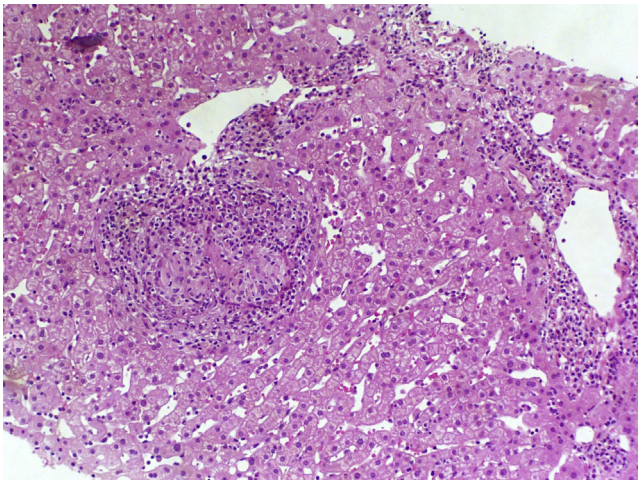

Figure 1 Histological study revealing minimal sinusoidal dilatation.

As a result, a percutaneous liver biopsy was subsequently performed. The anatomopathological study with haematoxylin and eosin staining, Masson's trichrome staining and reticulin staining did not reveal significant histological abnormalities. Only minimal sinusoidal dilatation was found (Fig. 1). Although no active bleeding was found during the endoscopy, the patient was deemed to have presented variceal upper gastrointestinal bleeding. As such, secondary prophylaxis with beta blockers and endoscopic ligation of the oesophageal varices was initiated.

At two-months follow-up (which found the patient to be asymptomatic), an abdominal Doppler ultrasound was performed, showing partial thrombosis of the proximal portion of the right portal vein and superior mesenteric vein. A thrombophilia study was ordered, which revealed factor V Leiden heterozygous mutation. As a result, anticoagulation therapy was started, initially with LMWH and then with acenocoumarol. Following initiation of anticoagulation therapy, subsequent CT scans revealed correct opacification of the splenic-mesenteric-portal venous system with a persistent minor peripheral repletion defect in the right portal branch smaller than found in previous studies. The patient is currently asymptomatic at 48-months follow-up. Endoscopic band ligation was performed six times and the patient continues to be treated with propranolol and acenocoumarol.

INCPH is a rare condition of uncertain aetiology characterised by PHT without liver disease. Although it can be found all over the world, it is most prevalent in developing countries and is associated with a lower socioeconomic status.[1,4] The aetiology of INCPH is unknown. A potential cause that has been hypothesised is the presence of persistent digestive tract infections or exposure to certain toxins (arsenic, vinyl, copper sulphate or azathioprine). HIV infection has been associated with the pathogenesis of INCPH, particularly in patients who have received antiviral therapy (notably following prolonged exposure to didanosine and/or stavudine), or as a direct result of HIV itself.[2,5,6] It has recently been postulated that these patients are more susceptible to prothrombotic abnormalities.[7,8] Portal vein thrombosis is more prevalent than in patients with liver cirrhosis, especially in patients with HIV-related INCPH.[1,2]

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A subgroup of patients with HIV and PHT without liver disease has recently been identified. These patients are characterised by preserved liver function and a lack of histological cirrhosis.[9] Significant splenomegaly is also typical. It is noteworthy that these patients present normal or slightly elevated HVPG values in the haemodynamic study, which underestimates the actual portal venous pressure. This is due to increased presinusoidal hepatic resistance and the presence of intrahepatic venous collaterals.[1] Despite the fact that our patient manifested unequivocal signs of PHT (oesophageal varices, splenomegaly, collaterals), an HVPG of 10 mmHg was found. Liver biopsy is vital to rule out cirrhosis and other causes of PHT. Histological abnormalities are varied and are not pathognomonic. Those that typically manifest include portal vein sclerosis, sinusoidal dilatation and nodular regenerative hyperplasia.[1,10]

There is no specific treatment for the condition, which is limited to managing the complications of portal hypertension.[7] Although the natural history of this condition is unknown, mortality in these patients is usually caused by variceal bleeding. Mortality rates are lower than for cirrhosis, probably due to preserved liver function.

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Dysphagia lusoria as a differential diagnosis in intermittent dysphagia☆

Disfagia lusoria como diagnóstico diferencial de la disfagia intermitente

The term dysphagia lusoria refers to symptoms that arise from vascular compression of the oesophagus.[1] This clinical entity was first reported by Bayford in 1787 in a woman with a long history of dysphagia, in whom an aberrant right subclavian artery (ARSA) was found during autopsy.[2] This artery abnormality was known as *luxus nature* before the term dysphagia lusoria was adopted. The prevalence of ARSA in the general population is estimated to be between 0.4% and 0.7%. Dysphagia lusoria is caused by the abnormal regression or failure to develop of the right fourth aortic arch during embryonic development.[3] In these cases, the right subclavian artery of the aortic arch runs towards the right arm behind the oesophagus and trachea in 80% of cases, between the trachea and the oesophagus in 15% of cases and in front of both in 5%.[4]

It may be asymptomatic in childhood or manifest with respiratory symptoms during ingestion owing to tracheal compression, as its elasticity is greater in infancy. Symptom onset is rare in adults and tends to manifest in the form of dysphagia arising from greater oesophageal rigidity and arteriosclerosis. Treatment will depend on symptoms, age, associated comorbidity as well as other concomitant vascular anomalies.[5]

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