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Portal Hypertension with Hepatoportal Sclerosis (HPS) in a Human Immunodeficiency Virus (HIV)-1 patient

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Abstract

Woman, 57 years old, with known HIV-1 infection since 2002 and treatment with lamivudine, didanosine (DDI) and nelfinavir for 2 years (2002-2004). Six years later, the patient presented with low volume hematemesis and melena and the upper gastrointestinal tract endoscopy (UGTE) identified large sized esophageal varices and hypertensive gastropathy. The study demonstrated normocytic and normochromic anemia, thrombocytopenia, ferropenia and cholestasis. Morphological features of chronic liver disease (CLD), splenomegaly and thrombosis in the portal vein (TPV) were observed in imaging exams. Potential causes of CLD and thrombophilia were excluded. The patient had a score of 8 kPa in liver elastography. HPS was identified in liver biopsy and the diagnosis of noncirrhotic portal hypertension (NCPH) secondary to DDI was made. The treatment consisted of oral β-blockers and endoscopic elastic band ligation. Anticoagulation did not eliminate the thrombus. After 12 years the patient remain stable.

Keywords

Didanosine; Hepatoportal sclerosis; Human immunodeficiency virus; Noncirrhotic portal hypertension.

Abbreviations

ART: Antiretroviral therapy; CLD: Chronic liver disease; DDI: Didanosine; HIV: Human Immunodeficiency Virus; HPS: Hepatoportal sclerosis; NCPH: Noncirrhotic portal hypertension; TPV: Thrombosis in the portal Vein; UGTE: Upper gastrointestinal tract endoscopy.

Introduction

DDI is a nucleoside reverse transcriptase inhibitor that was frequentely used in the past in combination with other antiretroviral drugs in HIV treatment [1]. There is an association between DDI liver toxicity and NCPH [2]. NCPH is a rare liver disease with significant morbidity and mortality [3]. Prolonged exposure to DDI is the most important risk factor to the development of NCPH [4]. The clinical presentation

consists of signs of portal hypertension such as ascites, hepatosplenomegaly and esophageal variceal bleeding. In the majority of cases liver function is preserved [5]. Endothelial damage in the portal system causes occlusion of the terminal branches of the portal venules. Portal vein thrombosis occur frequentely with portal hypertension [6]. The histology shows absence of cirrhosis with lack of portal veins and focal fibrous obliteration of small portal veins (HPS) [7]. Prognosis is usually poor [8].

Next we will describe a clinical case of a woman with NCPH secundary to DDI.

Case Presentation

A 57 years old woman was diagnosed with HIV-1 infection in 2002, transmitted by her husband. She didn't have other comorbidities. She received lamivudine, DDI and nelfinavir for 2 years (2002-2004), switching to zidovudine, tenofovir disoproxil and efavirenz duo to side effects such as lipoatrophy and diarrhoea. In June 2010 the patient presented with low volume hematemesis and melena. Large sized esophageal varices with no stigmata from recent bleeding (Figure 1) and hypertensive gastropathy were detected by UGTE.

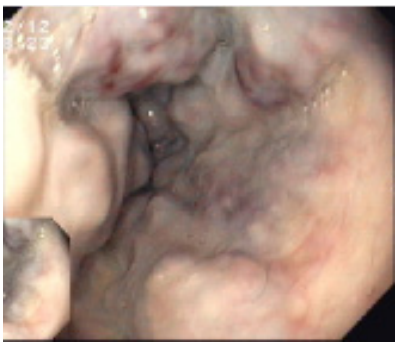

Figure 1: Large sized esophageal varices observed in UGTE.

Laboratory test found the following: normocytic and normochromic anemia (hemoglobin 9.4 g/dL), thrombocytopenia (platelets count 108000/µL), ferropenia (iron 16 µg/dL) and cholestasis (gamma-glutamyltransferase 132 U/L and alkaline phosphatase 191 U/L). HIV-1 viral load was undetectable and CD4+ T cell count was 335/µL. Abdominal doppler ultrasound and computed tomography scan (Figure 2) reported the presence of morphological features of CLD (right lobe atrophy and left lobe and caudate lobe hypertrophy), splenomegaly of 16 cm, low volume ascites and TPV.

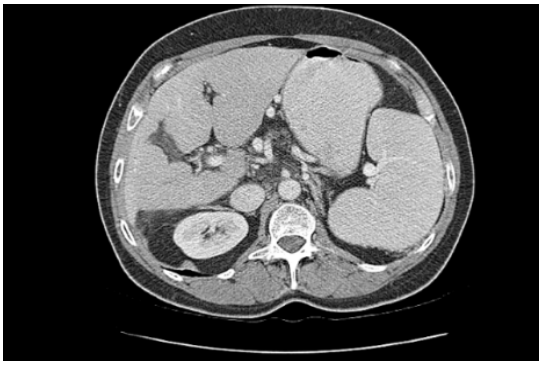

Figure 2: Abdominal computed tomography scan showing right lobe atrophy and left lobe hypertrophy), splenomegaly and low volume ascites.

Alcohol consumption and the use of other toxics were excluded. Serological antibody tests for hepatitis B and C were negative and autoimmune disease was ruled out (the following autoimmune parameters were evaluated: anti-smooth muscle, antinuclear, anti-mitochondrial, anti-soluble-liver-antigen and anti-liver-kidney microsomal antibodies). Transferrin saturation, ferritin, ceruloplasmin and alpha-1 antitrypsin were within normal ranges. Antithrombin, protein C and protein S were also normal.

Liver stiffness was 8 kPa in liver elastography. Liver biopsy was performed revealing portal lymphocytic infiltrate, bile ducts proliferation and also portal fibrosis, sclerosis of portal venules and sinusoidal dilatation which is compatible with HPS.

Given these results, the diagnosis of NCPH secondary to DDI's liver toxicity was proposed. The administration of oral β-blockers to decrease portal hypertension was initiated. The patient had to undergo several endoscopic elastic band ligation the following 8 years because of recurrent hematemesis or by indication during follow-up with UGTE. After a period of bleeding stabilization, an unsuccessful attempt to eliminate the thrombus from the portal vein was made with six months of oral anticoagulation.

After 12 years no further complications occurred and the patient has a stable condition, without anemia and with controlled thrombocytopenia. She is now taking dolutegravir and lamivudine with undetectable HIV-1 viral load and CD4+ T cell count between 250-300/μL.

Discussion/conclusions

The presence of thrombocytopenia, hepatosplenomegaly or liver decompensation in a HIV-infected patient with a history of DDI treatment should raise suspicion of NPCH [9]. Despite DDI cessation portal hypertension remain, although the disease progression slow down [4]. Our patient's portal hypertension was unknown until 6 years after DDI discontinuation when bleeding of esophageal varices occurred. The low volume of the hematemesis was never a life-threatening to our patient. Early diagnosis is importante in order to prevent potentially serious consequences [9] and to start the control of the portal hypertension-related sequela [10]. After several years we maintain the management of portal hypertension. Despite of the actual very secure profile of antiretroviral therapy (ART), we still have to deal with adverse effects of the past ART which remain as comorbidities in survivors.

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