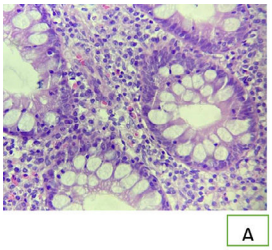
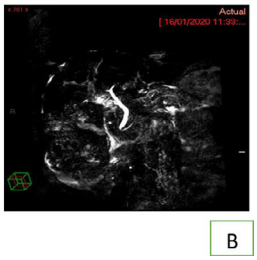
biopsy samples, reporting the presence of nonspecific proctitis, chronic colitis with focal ulceration, lymphoid aggregates, focal atrophy, and glandular distortion compatible with ulcerative colitis (UC). (Panel A) Due to the unusual association between UC and PBC, magnetic resonance cholangiography was requested, ruling out the overlap syndrome between primary biliary cholangitis and primary sclerosing cholangitis. (Panel B)

Discussion: A diverse heterogeneous group of hepatobiliary manifestations is reported in both UC and CD, and approximately 5% of adults with IBD have developed chronic liver disease. PBC is not usually associated with IBD, and concomitant reported cases are anecdotal. The presentations are different than the typical CBP without UC. PBC usually affects middle-aged women. The sex ratio is 10: 1 (female to male) and the mean age at diagnosis is 57.5 years. While the disease tends to affect men more often, with a female / male sex ratio of 2: 1 when associated with IBD. The distribution of ulcerative colitis in PBC patients is usually mild with limited bowel involvement. In a review by Tasa et al., eleven of 15 patients described left side colitis and proctitis.

Conclusion: The association between PBC and UC remains rare, as there are still few reported cases regarding the combined presentation of these diseases. Although PSC is the most specific hepatobiliary manifestation among UC patients with cholestasis, PBC should be considered in those with unexplained intrahepatic cholestasis. The use of a reliable test such as AMAs is of utmost importance to avoid misdiagnosis and/or under diagnosis.

The authors declare that there is no conflict of interest.





Panel A. Chronic and diffuse infiltrate of the basal lamina, with the presence of glandular distortion in rectal biopsies.

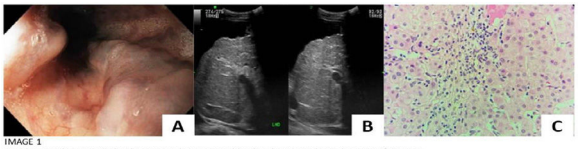


IMAGE 1
Panel A. Esophagogastroduodenoscopyshowing non-bleeding largeesophageal varices of Baveno
Panel B. Liver ultrasoundshowing the right lobe of the liver with irregularborders and a nodular pattern.
Panel C. HyE 40x staining.Hepatic parenchyma with pseudoacinartransformationis observed.

Panel B. The intra- and extra-hepatic bile duct preserved morphology without observing stenosis or dilations with normal signal intensity.

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AUTO IMMUNE HEPATITIS AS A LIVER MANIFESTATION OF COMMON VARIABLE IMMUNODEFICIENCY: A CASE REPORT

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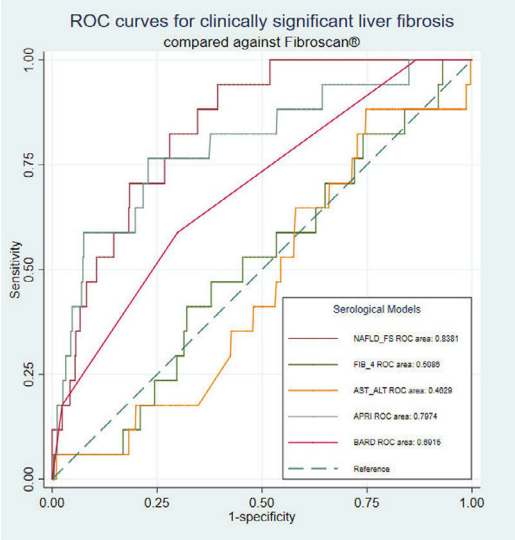
Introduction and Objectives: Common variable immunodeficiency (CVID) is a primary immunodeficiency disorder characterized by impaired differentiation of B cells with defective immunoglobulin production and paradoxically the development of autoimmune disorders. It is estimated that the prevalence of the liver disease is less than 10%. The most reported is focal nodular hyperplasia and, to a lesser extent, primary biliary cholangitis, primary sclerosing cholangitis and autoimmune hepatitis (AIH).

Objectives: present the case of a 22-year-old male who was admitted for hematemesis. He was diagnosed with immune thrombocytopenia 12 years ago without treatment and common variable immunodeficiency six years ago, currently three years without treatment. No family history of autoimmune or liver diseases. After extraction of the 4th molars, he presented swelling, redness, heat in the left submandibular region, unquantified fever, for which he received antibiotic treatment and non-steroidal anti-inflammatory drugs; 12 hours before admission, he began with hematemesis and hematochezia with data of hemodynamic instability, for which crystalloids are administered, the physical examination presents scleras with jaundice, an edematous, erythematous area with local heat in the left submandibular region, flat, symmetrical abdomen, absence of collateral circulation, Non-painful hepatomegaly, without shifting dullness. Laboratory admission with anemia, thrombocytopenia, cholestatic pattern in liver biochemistry tests, chronic viral infectious processes are ruled out and antibodies are made for autoimmune diseases, ANA 1:1200 is documented, other antibodies negative. Esophagogastroduodenoscopy where large esophageal varices of Baveno and severe portal hypertensive gastropathy (image 1 panel A). Doppler ultrasound reports diffuse liver disease, 14mm portal vein, with the presence of free fluid perihepatic and perisplenic, without biliary obstruction data (image 1 panel B). Percutaneous liver biopsy: fibrosis F2-3 on the scale of Metavir, interface hepatitis, associated with infiltrate lymphoplasmacytic integrating diagnosis of AIH (image 1 panel C).

Discussion: The prevalence of this condition is estimated at 1 in every 50,000 people worldwide. Up to 25% of patients with CVID will have an association with autoimmune diseases, representing the heterogeneous nature of the disease. The presentation of liver disease commonly reported in case series is anicteric cholestasis, and biopsies show evidence of non-cirrhotic portal hypertension. However, the association of CVID and AIH is rare; its diagnosis requires biopsies due to the lack of expression of antibodies in most cases.

Conclusions: In CVID patients with altered liver function tests, the association with autoimmune liver diseases should be ruled out to initiate timely treatment and avoid late complications.

The authors declare that there is no conflict of interest.



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