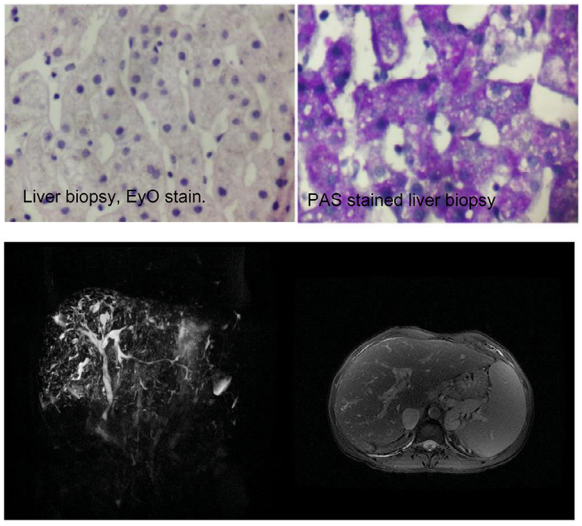
age; the clinical data will depend on the affected tissue and type of Glycogenosis. The histopathological report is the mainstay of diagnosis in this type of case and making a differential diagnosis with other entities and a genetic study.

Conclusions: Glycogen storage disorders are part of a group of rare and few suspected pathologies. It is not frequent to find them in adult patients due to their complications. The prognosis must be individualized based on the affected tissue and the subtype presented.

The authors declare that there is no conflict of interest.



Collagioresonance

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ADDRESSING THE INFILTRATIVE PATTERN: COMPLEX DIAGNOSIS

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Introduction and objective: Alterations in liver function tests are frequent, so the approach based on the predominant alteration and the patient's medical history is relevant. Carrying out a liver biopsy in cases of diagnostic doubt is imperative. The objective of this work is to describe a clinical case of an infiltrative pattern approach that culminated in the diagnosis of Hepatosplenic T-cell Lymphoma (HSCTL).

Patients and Methods: Clinical case report. Presentation. Woman, 72 years old. History of a sister with cirrhosis. Consumption of alcohol and herbalists; arterial hypertension and Sjögren. He was admitted for persistent fatigue and jaundice. Laboratories with anemia, thrombocytopenia and kidney injury; Hepatic biochemistry with a predominantly cholestatic pattern at the expense of alkaline phosphatase and direct hyperbilirubinemia. Without acute liver failure. By imaging the liver, vessels and normal bile duct; splenomegaly; Negative hepatitis viral panel, positive ANAs and Anti-actin, negative antimitochondrials, normal immunoglobulins. HAI vs. DILI / HILI is suspected. Liver biopsy reports HSTCL-type lymphoproliferative process (Figure 1). It was supplemented with bone marrow aspirate and PET-CT. He started prednisone and cyclophosphamide.

Discussion: The systematic approach to altered liver biochemistry requires integrating personal and family risk factors for liver disease. The infiltrative pattern that resembles the cholestatic one represents a diagnostic challenge as it is little recognized. In this case, we report a rare neoplasm corresponding to 5% of peripheral T lymphomas; they usually develop in young adults and in the absence of lymphadenopathy. It also has an adverse prognosis due to refractoriness to chemotherapy.

Conclusion: The HSTCL presented in this clinical case represents a complex and infrequent diagnosis. The symptoms and age group were atypical and the identification was possible through a systematic evaluation of the infiltrative pattern and differential diagnoses.

The authors declare that there is no conflict of interest.

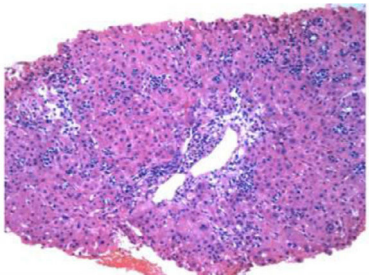

Figure 1. Atypical lymphocyte infiltrate in liver parenchyma.

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PRIMARY BILIARY CHOLANGITIS COMPLICATED WITH ULCERATIVE COLITIS: A CASE REPORT

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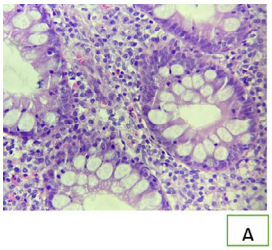
Introduction and Objectives: Liver involvement is not unusual in patients with inflammatory bowel disease (IBD), where one third of patients have abnormal liver biochemical tests, becoming a diagnostic challenge. Primary biliary cholangitis (CPB) is an autoimmune liver disease that presents with chronic cholestasis, the presence of specific antibodies and histological findings of destructive non-suppurative cholangitis. Genetic, immunological, and environmental factors that contribute to the pathogenesis of IBD may also contribute to associated hepatobiliary disorders. Objective: Present the case of a 67-year-old woman who consulted for cholestatic symptoms. Medical record of smoking for 25 years was suspended 15 years ago with a smoking index of 5. She reported a history of weakness, asthenia, self-limited palmar and plantar pruritus of 2 years of evolution, associated with abnormal liver function tests (total bilirubin 1.8 mg / dl, INR 1.40, albumin 3.2 g / dL). The initial physical examination revealed jaundice in the sclera, palmar erythema, evidence of telangiectasia in the abdomen, collateral circulation with medusa caput, positive ascitic wave, and splenomegaly. R factor is cholestatic, with alkaline phosphatase of 416 U / L, gamma glutamyl transpeptidase 660 U / L, alanine transferase 59 U / L, for which possible viral and autoimmune causes are addressed as the first possibility, reflecting antimitochondrial antibodies with high titers (278 U / L), associated with immunoglobulin G of 2490 mg / dl and immunoglobulin M of 734 mg / dl. During her one-year follow-up, she reported the onset of diarrheal stools Bristol 6, 2 to 3 episodes per day, with occasional urgency and with inflammatory characteristics, due to the presence of mucus and blood. Infectious causes are ruled out, elevated acute phase reactants are reported, followed up with a colonoscopy and

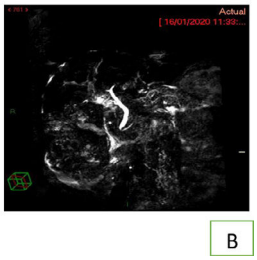
biopsy samples, reporting the presence of nonspecific proctitis, chronic colitis with focal ulceration, lymphoid aggregates, focal atrophy, and glandular distortion compatible with ulcerative colitis (UC). (Panel A) Due to the unusual association between UC and PBC, magnetic resonance cholangiography 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.

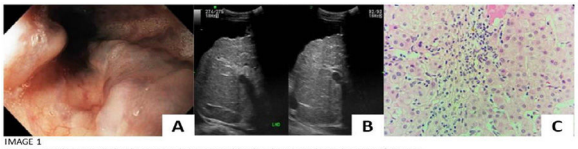


IMAGE 1
Panel A. Esophagogastroduodenoscopyshowing non-bleeding largeesophageal varices of Baveno
Panel B. Liver ultrasoundshowing the right lobe of the liver with irregularborders and a nodular pattern.
Panel C. HyE 40x staining.Hepatic parenchyma with pseudoacinartransformationis observed.

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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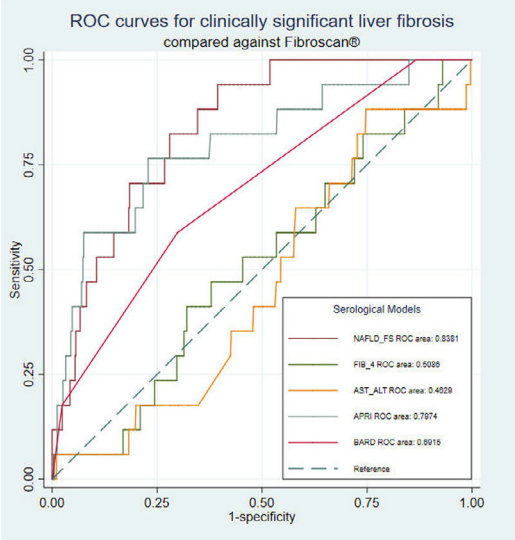
Introduction and Objectives: Common variable immunodeficiency (CVID) is a primary immunodeficiency disorder characterized by impaired differentiation of B cells with defective immunoglobulin production and paradoxically the development of autoimmune disorders. It is estimated that the prevalence of the 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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