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When Primary Biliary Cholangitis Affects Humans: Two Observations That Challenge the Standards

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Abstract | **Case Report**

Primary biliary cholangitis (PBC) is a rare cause of cholestatic jaundice in men. We report two rare cases of PBC in men presenting with chronic cholestasis (jaundice, pruritus, dark urine, pale stools, alkaline phosphatase (ALP) greater than 1.5 times the upper limit of normal, and glucocorticoids (GGT) greater than 3 times the upper limit of normal). The first case had positive anti-mitochondrial type 2 antibodies, while in the second, these antibodies were negative, with positive anti-gp210 antibodies. Histological confirmation of PBC in the first case was achieved. The second case was diagnosed at the stage of cirrhosis, based on signs of portal hypertension and liver parenchymal dysmorphism on imaging. Both patients were treated with ursodeoxycholic acid.

Keywords: Primary biliary cholangitis, Male, Jaundice, Cholestasis, Antimitochondrial antibodies type 2, Case report.

INTRODUCTION

Primary biliary cholangitis (PBC) is a cholestatic disease of autoimmune etiology, characterized by inflammatory destruction of the small intrahepatic bile ducts, which can progress to stages of fibrosis and even liver cirrhosis. It predominantly affects middle-aged women (generally between 30 and 60 years old). [1], [2] The development of this disease in men is very rare (estimated incidence of 0.7 per 100,000 person-years) with a median age at diagnosis of 62 years. [3]

We report two cases of PBC in men revealed during an evaluation for chronic cholestatic jaundice.

OBSERVATION

Case 1:

A 67-year-old male patient, with a history of COVID-19 infection 3 months ago which was treated, operated on in 2013 for a cervical disc herniation, operated on in 2014 for a meningioma, put on anticonvulsant (Lamotrigine) post-operatively due to the persistence of intermittent convulsions, with a history of chronic smoking for 20 years, quit 14 years ago, with no history of alcoholism.

Complains of permanent cutaneous-mucosal jaundice, with dark urine and discolored stools as well as generalized pruritus, dating back 2 months, without vomiting, without externalized digestive bleeding, without abdominal pain, without other manifestations, all evolving in a context of apyrexia, and alteration of general condition (weight loss of 11 Kg in 2 months, marked asthenia and anorexia).

Clinical examination revealed a conscious patient, hemodynamically and respiratory stable, afebrile, with jaundice of the skin and mucous membranes, and diffuse scratch marks. The remainder of the clinical examination revealed no other abnormalities.

The laboratory tests revealed signs of cholestasis (ALP 274 U /L (2.1 times the upper limit), GGT 79 U/L (1.1 times the upper limit), and total bilirubin 150 mg/L with a predominance of the conjugated form (140 mg/L)), associated with minimal cytolysis (AST 112 U/ L (2.2 times the upper limit) and ALT 72 U/L (1.7 times the upper limit)). Prothrombin and serum albumin levels were normal. An abdominal ultrasound was performed and showed no abnormalities, particularly no bile duct dilation. The etiological workup included serological tests for viral hepatitis (HAV, HBV, HCV, EBV, CMV), which were negative. A MRCP was

also performed, which was also negative. abnormalities were observed. The ferritin level was elevated (841 µmol /g) with a normal transferrin saturation (32%). C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) levels showed a biological inflammatory syndrome (CRP 21 mg/L and ESR 29 mg/L in the first hour). Tumor markers (alpha -fetoprotein (AFP), carbohydrate antigen 19-9 (CA 19-9), and carcinoembryonic antigen (CEA)) were negative. Immunological testing showed positive anti-mitochondrial type 2 antibodies (13 AU/ml). The remaining immunological tests (antinuclear antibodies, anti-smooth muscle antibodies, anti-cytosol 1 (LC1) antibodies, anti- Liver and Kidney Microsome 1 (LKM1) antibodies, anti-soluble liver antigen (SLA) antibodies, and assays of Total IgG was normal. The rest of the blood work, including the complete blood count, serum electrolytes, renal function tests, and metabolic panel, was unremarkable. A liver biopsy was performed and showed degenerative and regenerative hepatocyte lesions with portal fibrosis, cholestatic lesions, and bile duct destruction.

The search for other associated autoimmune diseases, particularly celiac disease (negative anti-transglutaminase antibody serology), autoimmune thyroid dysfunction (anti-thyroperoxidase antibodies (Ath) anti-TPO) and anti-TSH receptor antibodies (anti-TSH receptor antibodies), Sjogren's syndrome, scleroderma, rheumatoid arthritis was negative.

The diagnosis of primary biliary cholangitis was made, and the patient was put on ursodeoxycholic acid (UDCA) at a dose of 13 mg/kg/day.

2nd case:

A 47-year-old male patient, followed for 23 years for type 1 diabetes, put on Insulin (Basal Bolus Protocol).

Complains of permanent cutaneous-mucosal jaundice, with dark urine and discolored stools as well as generalized pruritus, evolving for 3 months, without vomiting, without external digestive bleeding, without abdominal pain, without other manifestations, all evolving in a context of very marked asthenia.

Clinical examination revealed a conscious patient, hemodynamically and respiratory stable, afebrile, with marked jaundice of the skin and mucous membranes, and diffuse scratch marks. The remainder of the clinical examination revealed no other abnormalities.

The laboratory tests also revealed signs of cholestasis (ALP 479 U/L (3.7 times the upper limit), GGT 246 U/L (3.4 times the upper limit), and total bilirubin 49 mg/L with a predominance of the conjugated form (44 mg/L)), associated with minimal cytolysis (AST 56 U/ L (1.3 times the upper limit) and ALT 50 U/L (1.1 times the upper limit)). Prothrombin and serum albumin levels were normal. The etiological workup included serological tests for viral hepatitis (HAV, HBV, HCV, EBV, CMV), which were negative. An abdominal ultrasound showed a heterogeneous liver with irregular contours, dysmorphic in appearance consistent with chronic liver disease, without detectable nodular lesions and with no dilation of the bile ducts. Biliary tract. Tumor markers (alpha -fetoprotein (AFP), carbohydrate antigen 19-9 (CA 19-9), and carcinoembryonic antigen (CEA)) were negative. An immunological workup, including antinuclear antibodies, anti-smooth muscle antibodies, anti-mitochondrial type 2 antibodies, anti- Sp 100 antibodies, anti-cytosol 1 (LC1) antibodies, anti-Liver and Kidney Microsome 1 (LKM1) antibodies, anti-soluble liver antigen (SLA) antibodies, and total IgG levels, was normal. Only the anti-GP 210 antibody was strongly positive. The rest of the laboratory workup, including the complete blood count, serum electrolytes, renal function tests, and metabolic panel, was unremarkable.

Non-invasive assessment of liver fibrosis by elastometography revealed fibrosis classified as F4. An esophagogastroduodenoscopy was performed as part of the search for endoscopic signs of portal hypertension and showed the presence of small esophageal varices without red signs.

The search for other associated autoimmune diseases, particularly celiac disease (negative anti-transglutaminase antibody serology and upper endoscopy with unremarkable duodenal biopsies), and autoimmune thyroid dysfunction (anti-thyroperoxidase antibodies (Ath) anti- TPO) and anti-TSH receptor antibodies (anti-TSH receptor antibodies), Sjogren's syndrome, scleroderma, rheumatoid arthritis was negative.

The diagnosis of primary biliary cholangitis complicated by hepatic cirrhosis at the stage of portal hypertension was made, and the patient was put on ursodeoxycholic acid (UDCA) at a dose of 15 mg/kg/day, associated with prophylaxis of hemorrhagic complications by beta-blockers.

DISCUSSION

Primary biliary cholangitis (PBC), the new name for primary biliary cirrhosis, is a cholestatic disease of autoimmune etiology, characterized by immune-mediated inflammatory destruction of the small intrahepatic bile ducts, which can progress to fibrosis and potentially lead to liver cirrhosis. This condition predominantly affects women (with a female-to-male ratio ranging from 9:1 to 22:1) and the average age at diagnosis is generally between 30 and 60 years. [1], [2]

Certain factors, including genetic, environmental, and infectious factors, are considered necessary for the development or progression of PBC. [3]– [6] The pathophysiological mechanism of PBC appears to be based on a dysregulation of T lymphocytes, [7]– [10] as well as molecular mimicry and cross-

immunity involving microbial or self-antigens responsible for a destructive inflammatory reaction of the epithelial cells of the small intrahepatic bile ducts. [8]

The modes of presentation of the disease are variable: patients may be asymptomatic at the time of diagnosis, or present with nonspecific symptoms frequently including asthenia, jaundice of the skin and mucous membranes, generalized pruritus… The disease may also be revealed at late stages, particularly during compensated liver cirrhosis or at the stage of decompensation. [7], [9], [10]

Biologically, PBC manifests as chronic cholestasis (elevated alkaline phosphatase (ALP) > 1.5 times the upper limit of normal and gamma-glutamyl transferase (GGT) > 3 times the upper limit of normal). An increase in bilirubin levels is neither constant nor necessary for diagnosis. It is often associated with a moderate elevation of transaminases.

In 10% of cases, PBC is associated with autoimmune hepatitis, resulting in an overlap syndrome. In this particular form, laboratory tests show a significant elevation of transaminases (greater than 5 times the normal level), associated with hypergammaglobulinemia with elevated IgG and the presence of anti-smooth muscle antibodies. [11]

Anti-mitochondrial antibodies are the gold standard test for the diagnosis of PBC, with a sensitivity and specificity of 90% and 97%, respectively [2]. If this test is negative, further testing should be performed to detect PBC-specific antinuclear antibodies (anti-gp210 and anti-sp100 antibodies). [12]

Histological confirmation by liver biopsies can provide information on the stage of the disease, which was classified by Scheuer into 4 stages : non-suppurative destructive cholangitis in stage 1, peri-portal ductal proliferation in stage 2, extensive septal fibrosis in stage 3 and cirrhosis in stage 4. [13]

The development of PBC in males is very rare. Its incidence has been estimated at 0.7 per 100,000 person-years with a median age at diagnosis of 62 years. [3] Several studies have failed to demonstrate significant sex differences in the biochemical, immunological, and histological characteristics of PBC; however, intense pruritus and systemic manifestations have been reported to be more frequent in women than in men. Conversely, jaundice, signs of portal hypertension, including gastrointestinal bleeding, and the development of hepatocellular carcinoma are more commonly described in men. [8], [14], [15]

Ursodeoxycholic acid (UDCA), with a dosage of 13–15 mg/kg/day. The efficacy of UDCA treatment is assessed by the biological response 1 year after the start of treatment. The most widely used definition of biological response is that of the Paris II criteria [16]: ALP < 1.5 times the upper limit of normal and AST < 1.5 times the upper limit of normal and normal bilirubin 1 year after the start of treatment. [16] In case of non-response to treatment with UDCA, poor adherence, associated autoimmune hepatitis (overlap syndrome) or another autoimmune disease should be investigated.

Obeticholic acid has been granted marketing authorization as a second-line treatment, either as a replacement for ursodeoxycholic acid (UDCA) in cases of poor tolerance, or in combination with UDCA in cases of lack of biological response to UDCA. Other molecules such as fibrates and budesonide also constitute second-line treatments. [12]

CONCLUSION

Primary biliary cholangitis (PBC) is a rare cause of cholestatic jaundice in men. Its clinical, biological, immunological, and histological profile is the same in women. However, the development of signs of hepatocellular insufficiency (particularly hemorrhagic complications) and hepatocellular carcinoma are more frequent in men. This may be due to delayed diagnosis. Hence the importance of considering this condition to prevent progression to cirrhosis and its complications.

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