Hashimoto's thyroiditis associated with idiopathic portal hypertension

To the Editor Idiopathic portal hypertension (IPH) is classified as intrahepatic presinusoidal noncirrhotic portal hypertension. The etiology of IPH is not fully understood. Such heterogeneous factors as chronic arsenic intoxication and exposure to vinyl chloride may play a causative role.[1,2] IPH is common in many Asian countries, such as India and Japan, and is rarely seen in Western countries or in Turkey.[3] Affected patients have elevated portal hypertension in the absence of hepatic histopathological changes on light microscopy.[4]

Hashimoto's thyroiditis is one of the most common autoimmune diseases, affecting up to 10% of the general population. It frequently affects women but the reason for this remains unknown. The examination of the thyroid gland on light microscopy usually reveals characteristic features of the disease, including lymphocytic infiltration, follicular destruction, and fibrosis.[5]

To date, only 4 cases of IPH associated with Hashimoto's thyroiditis have been reported, mostly by Japanese authors.[5-8] In the current paper, we present the case of a Turkish patient.

A 33-year-old woman presented to a gastroenterology clinic with a 2-week history of abdominal discomfort, fatigue, generalized weakness, and cold intolerance. The patient was born and lived in the eastern part of Turkey. The family history was negative. The patient did not take any medications at the time of admission.

On physical examination, the abdomen was soft with nonspesific tenderness on palpation and the spleen was enlarged. The neck examination showed diffuse enlargement of the thyroid gland. No other abnormalities were detected.

The laboratory analysis revealed anemia, leukopenia, and thrombocytopenia (hypersplenism) with an elevated sedimentation rate, low free thyroxine levels, and increased levels of serum thyroid-stimulating hormone (TSH) and antithyroid antibodies. Liver transaminases were slightly above the normal range. The blood levels of ammonia, total bilirubin, alkaline phosphatase, and amylase were normal. Alcohol was not detected. A moderately prolonged prothrombin time was observed. Serologic tests for detecting hepatotropic viruses such as hepatitis B, C, and D were negative.

An abdominal ultrasound showed moderate hepatosplenomegaly with an increased portal and splenic vein diameter. No obstruction was detected on the portal vasculature (FIGURE 1). A thyroid ultrasound revealed a moderately enlarged but diffusely hypoechoic thyroid gland, suggesting autoimmune thyroid disease (FIGURE 2).

The endoscopic examination showed grade 3 esophageal varices and portal hypertensive gastropathy. The pathological examination of the liver tissue from a percutaneous biopsy showed massive infiltration of lymphocytes, dilatation of the hepatic sinusoids, and periportal fibrosis, which was consistent with the diagnostic criteria for IPH (FIGURE 3).

The final diagnosis was IPH associated with Hashimoto's thyroiditis. L-thyroxine (50 µg/day) was prescribed to manage hypothyroidism. Propranolol (20 mg orally thrice a day) was also started to reduce portal hypertension.

Thus, we report here a rare case of a patient with IPH and enlarged thyroid due to Hashimoto's disease.

IPH is characterized by esophageal varices, hepatosplenomegaly, preserved liver function, noncirrhotic liver architecture, distortion of the microvascular pattern of the liver, and increased portal vein diameter as well as the lack of causative factors for parenchymal liver disease. IPH is more common in women. The diagnosis of IPH is based on laboratory tests with a radiological examination and usually confirmed by liver biopsy. IPH is diagnosed after exclusion of other causes of portal hypertension.[9]

Our patient had no family history of liver disease. Significant hypersplenism and splenomegaly were observed on admission. Negative serologic tests for viral hepatitis, evidence of portal hypertension, lack of cirrhosis and patent portal tract suggested IPH as the primary pathology.

The radiological examination could not be performed in our patient, so we decided on liver biopsy. It revealed sinusoidal dilatation with periportal fibrosis and periportal lymphocytic infiltration suggesting IPH. Further histopathological findings allowed us to establish the final diagnosis.

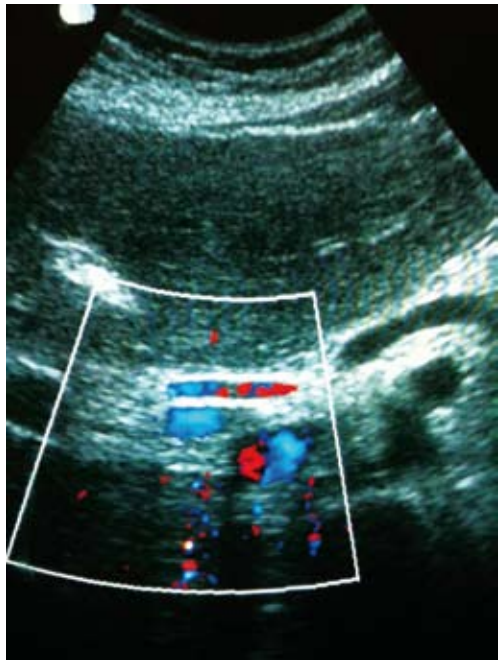

FIGURE 1 Abdominal ultrasound revealed patent portal vein with noncirrhotic liver

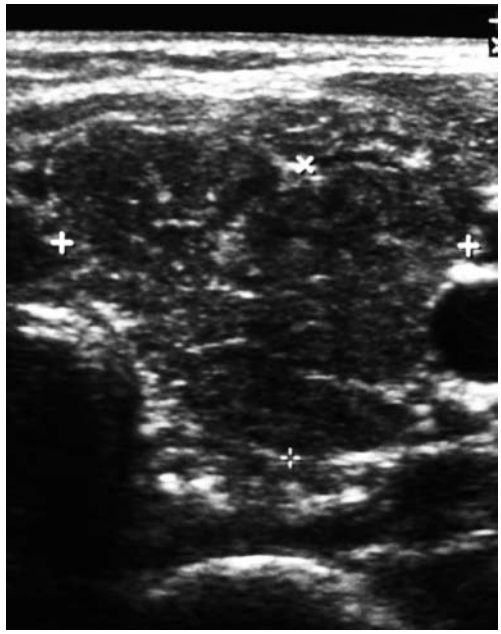

FIGURE 2 Thyroid ultrasound revealed an enlarged and hypoechogenic thyroid gland with fine septae

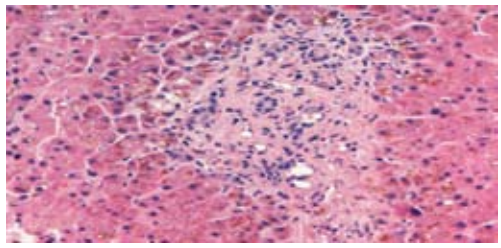

FIGURE 3 On histological examination of liver biopsy specimens, sinusoidal dilatation with periportal fibrosis and periportal infiltrate of lymphocytes were observed

In patients with advanced grade esophageal varices, sclerotherapy or band ligation in combination with β-blockers, such as propranolol, might be required to prevent esophageal variceal bleeding.[4] Therefore, we decided to start a long-term therapy with propranolol in our patient.

Hashimoto's thyroiditis (autoimmune thyroiditis) is the most prevalent autoimmune disease in the world. The diagnosis of Hashimoto's thyroiditis is usually established on the basis of biochemical and radiological tests. Elevated TSH levels, low thyroxine and triiodothyronine levels, and the presence of autoantibodies to thyroid antigens, including thyroid peroxidase and thyroglobulin, are the serological markers of the disease.[5]

Our patient had a number of signs and symptoms suggesting chronic Hashimoto's thyroiditis, including heterogeneity of the enlarged thyroid gland, serologic evidence of autoantibodies to thyroid antigens, and hypothyroidism. An enlarged and diffusely hypoechoic gland with fine echogenic septae on ultrasonography are the key findings for Hashimoto's thyroiditis.[10]

An association between an immune phenomenon and IPH has also been reported by Japanese authors.[11] In their paper, the presence of hypergammaglobulinemia, serological evidence of autoantibodies, and higher rates of chronic thyroiditis were considered the main immune characteristics of IPH. In another Japanese study,[12] a reduction in suppressor T lymphocyte activity, increase in Leu3a/Leu2a, and the presence of some autoantibodies, for example, antinuclear or antilymphocyte antibodies have been reportedly associated with IPH, and these findings were largely attributed to an obscure immune mechanism.

In summary, IPH and Hashimoto's thyroiditis may share common etiology due to their immune background. Interestingly, the Turkish population, which originates from the Asian population, may share similar immune features with other Asian nations, including the Japanese population. The current case may contribute to the further understanding of the disease and suggests that the immune mechanism is involved in the pathogenesis of IPH.

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