CASE REPORT

Idiopathic Portal Hypertension Associated With Autoimmune Thyroiditis

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Summary

We report a case of idiopathic portal hypertension (IPH) in association with autoimmune thyroiditis occuring in a 39 year old woman. Ultrasonography revealed a normal liver echotexture. Spleno-portogram confirmed portal hypertension and liver biopsy showed features in keeping with IPH. She was also found to be biochemically hypothyroid with markedly elevated thyroid autoantibodies. These findings may suggest an autoimmune link in the pathogenesis of IPH.

Key words: Idiopathic portal hypertension, autoimmune thyroiditis.

Introduction

Idiopathic portal hypertension (IPH) is uncommon in the west but is a relatively common cause of portal hypertension in the east in particular India and less commonly in Japan[1,2]. Classically it consists of the triad of splenomegaly, hypersplenism and portal hypertension in the absence of other causes of portal hypertension. Its association with other autoimmune diseases has long been recognised and autoimmunity has been postulated as a possible aetiology[1,2]. We report a case of IPH with concomitant autoimmune thyroiditis.

Case Report

A 39 year old Malay woman presented with a two month history of ankle and abdominal swelling. Abdominal examination revealed hepatosplenomegaly and ascites. She had no stigmata of chronic liver disease. She gave no previous history of jaundice, was not known to be a hepatitis B carrier and did not consume alcohol. Clinically the patient was of short stature, mentally subnormal but otherwise had no features to suggest hypothyroidism. The thyroid gland was not enlarged.

Full blood count revealed pancytopenia with a haemoglobin of 3.0 g/dL (11.5-16.5), platelet count of 130 x 10^9/L (150-400) and white blood count of 3.2 x 10^9/L (4.0-11.0). Liver function test was normal; prothrombin time and partial thromboplastin time were also normal. Hepatitis B surface antigen was negative.

Ultrasound examination revealed hepatosplenomegaly with the liver having a normal homogenous echotexture. The main portal vein was dilated, measuring 15 mm in diameter. The splenic and superior mesenteric veins were also dilated to 10 mm each. These features were consistent with the ultrasound diagnosis of non-cirrhotic portal hypertension. Gastroscopy, however, did not reveal any varices.

Liver biopsy showed mild fibrosis and inflammatory infiltration of the portal tracts, with no extension to adjacent portal areas. One of these portal tracts had an obliterated vein adjacent to it.

Percutaneous splenoportogram revealed markedly dilated portal and splenic veins with no evidence of extrahepatic obstruction (Figure 1).

Thyroid function test showed evidence of hypothyroidism with T4 level of 59 nmol/l and TSH of 23 IU/ml. Antibodies to antimicrosomal and antithyroglobulin were elevated to titres of 1:640 and 1:160 respectively. Other autoimmune markers, in particular anti-nuclear factor, anti-mitochondrial and anti-smooth muscle anti-bodies were negative.

She was transfused with packed cells and commenced on l-thyroxine.

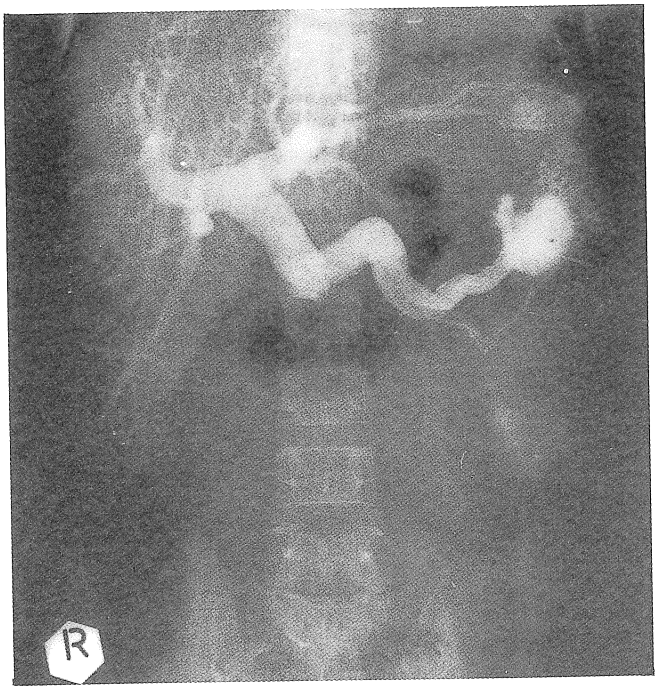

Fig. 1: Splenoportogram demonstrating markedly dilated splenic and portal veins.

Discussion

Idiopathic portal hypertension (IPH) is an uncommon condition characterised by splenomegaly, pancytopenia and portal hypertension in the absence of liver cirrhosis or extrahepatic portal obstruction[2]. It is rare in the western world but is common in Japan and India; with the majority of work in this area coming from Japan[1,2].

The aetiology of this condition remains unclear. Toxic substances (eg. arsenic and vinyl chloride) and cytotoxic agents (eg. methotrexate, azathioprine and mercaptopurine) have been implicated[3]. The possible role of infection has also been considered in view of the poor standard of hygiene in countries

like India[1,3]; the gastrointestinal tract being the portal of entry for toxins to be absorbed leading to hepatic vasculature damage[3].

The possible role of autoimmune agents have been discussed as far back as 1956 by Luxton and Cooke and more recently three cases of IPH with Hashimoto's disease have been reported amongst the Japanese[2]. IPH has also been reported in other autoimmune disorders eg. progressive systemic sclerosis; hence autoimmunity is gaining stronger grounds in the aetiopathogenesis of IPH.

In the study done by Okuda[1], it was found that the majority of these patients were female with a male to female ratio of 1:4.4. The average age of diagnosis was between 40-60 years, similar to patients with Hashimoto's disease. The commonest initial symptoms and signs were those of anaemia, gastrointestinal bleeding, mostly haematemesis and abdominal mass (splenomegaly). Most of these patients lacked the stigmata of chronic liver disease[1].

Laboratory investigations frequently showed the presence of anaemia which in the majority will be that of normochromic normocytic type. Iron-deficiency anaemia may occur with massive gastrointestinal bleed or chronic slow bleed from the varices. Leukopenia and thrombocytopenia were also described and was probably due to hypersplenism.

Liver function tests are usually normal. Histologically, most hepatic changes are confined to portal areas[1,2,3] where varying degrees of portal fibrosis may be seen associated with portal vein sclerosis or even thrombosis of the portal vein branches.

Mild to moderate inflammatory cell infiltrates in the portal areas have been described. These changes are often patchy, hence, for definitive diagnosis a wedge biopsy is preferred to percutaneous needle biopsy to avoid sampling error. In areas where schistosomiasis is endemic, this condition needs to be excluded first as the histological findings may be indistinguishable from IPH[1,2]. In the patient discussed, there were no ova seen in her stool specimen nor on liver biopsy.

Portogram is usually required in these patients to exclude extrahepatic causes of portal hypertension.

Prognosis of IPH is good with life span remaining unaltered. This good prognosis is related to the fact that liver cells and function are usually intact.

In conclusion, IPH should always be suspected in a patient presenting with unexplained portal hypertension. This index of suspicion is greater if the patient is a middle age female with a coexisting autoimmune disorder in particular thyroiditis. As it is associated with a good prognosis, treatment is only aimed at the main complication of variceal bleeding.

References

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