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REVIEW ARTICLE

CELIAC DISEASE AS A POTENTIAL CAUSE OF IDIOPATHIC PORTAL HYPERTENSION: A RARE CLINICO-PATHOLOGIC ENTITY

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ABSTRACT

Celiac disease is an autoimmune-based digestive disease and it is an immune-mediated enteropathy due to the ingestion of a gluten containing diet. Individuals predisposed to risk are genetically predetermined. Symptoms of the disease at a large usually start at the very early part of life just after weaning, however, a small group of patient present late we called it 'latent celiac sprue". We present a rare case of young patient having celiac disease with symptomatic idiopathic portal hypertension.

INTRODUCTION

Celiac disease is a systemic disease having wide range of clinical manifestations. Syptomatic or Classic celiac disease refers to presentation with diarrhea, with or without malabsorption symdrome, where as in atypical or silent celiac disease gastrointestinal symptoms are lacking or not prominent. IPH/NCPF is a disorder of unknown etiology, clinically characterized by portal hypertension, splenomegaly and anemia secondary to hypersplenism. The characteristic portal hemodynamics include intrahepatic presinusoidal portal hypertension, increased splenic and portal vein blood flow, and increased intrahepatic portal resistance. The prognosis is generally good depending on the management of bleeding varices. Although the etiology is obscure, certain immunologic abnormalities seem to play an etiologic role in Japanese patients, and the incidence has markedly declined in recent years in Japan, indirectly suggesting a role of infection.

Case presentation

21 Years old male presented to us with hematemesis and fever, he has also past history of large bowel diarrhea with generalized weakness. On examination he was found to have short height (105CM) as per his age, weight was 26kg (Pic -1). He had generalized muscle wasting, pallor. Abdominal examination revealed large spleen (11cm below left costal margin), no free fluid was noted in the abdomen.

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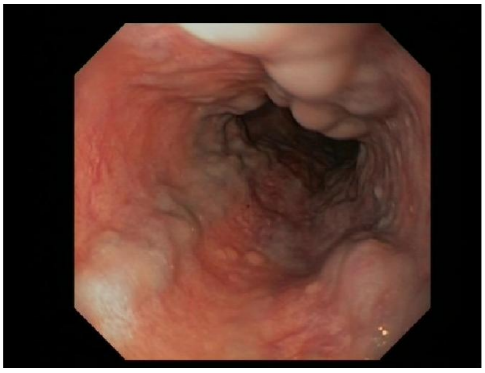
Fig.1. Muscle wasting and short height of the patient

Laboratory Investigation showed Hb -8.2g%, total count -2300 and platelet of 49,000, LFT –TB-0.83, DB-0.43, TP-6.7, ALB-4.1, SGOT-34, SGPT-37, ALP-341, GGT-62, Folate -4.6 low, Anti-TTG- >1000.Ceruloplasmin -26mg/dl, KF ring was not detected.USG abdomen –Liver span was 13cm, normal size and echotexture, spleen was enlarged with span of 17cm, PV was 13mm with hepatopetal flow and portosystemic collateral vessels however no thrombos noted. Rest of the organ was within normal limit.



Picture-2. Esophageal vaeices

Endoscopy (Pic-2) showed large esophageal varices which were banded and duodenum showed attenuation of mucosal folds biopsies of which revealed features of celiac disease (Marshal –class-111). Bone marrow showed features of hypersplenism. In view of low platelet patient was advised for a trans-jugular liver biopsy, TJLB revealed a pressure study of normal (HVPG <5 mm of Hg). A liver biopsy showed normal parenchyma, cupper stain was normal.

DISCUSSION AND CONCLUSION

This patient presented to us with feature of portal hypertention and hyperspenism and no evidence of liver parenchymal disease as evidenced by normal albumin and liver span, his HVPG was normal. IPH is a heterogeneous and multifactorial disorder with a potential genetic contribution, seen most often in the Indian subcontinent and in East Asia (**Okudaira *et al.*, 2002; Okuda, 2002; Scott and Losowsky, 1975**). Trace element chemical theory, autoimmunity theory and infection theory have been suggested in the literature, although none has been clearly proven (**M'saddek *et al.*, 2007**). It seems that celiac disease, by an increased immune reaction in the splenoportal axis, can result in the development of idiopathic portal hypertension in susceptible affected patients (**Kara and Sandikci, 2007; Sharma *et al.*, 2006**). As his liver function tests were totally normal, Celiac disease was suggested as a cause of IPH in this patient, as his symptoms improved while he adhered to a gluten free diet. As on follow up his haemoglobin was improved (9.2gm%), serum folate (12.2mg/dl) became normal. The association of celiac disease with IPH has been recently reported in the literature (**Harmanci and Bayraktar, 2007**). The improvement of portal hypertension with a gluten free diet, a rare entity reported in a case, implicates a causal relationship between portal hypertension and increased inflammatory reactions in celiac disease (**Sama *et al.*, 1971; Ichimura *et al.*, 1993**).

Abbreviations

IPH-Idiopathic portal hypertension, NCPF-Non-cirrhotic portal fibrosis, HVPG-Hepatic venous pressure gradient. TJLB-Trans-jugular liver biopsy.

Competing interests

The authors declare that they have no competing interests.

Authors' contribution. Dr K C Das, Dr Navjot Singh, Dr Sumeet David, Dr. Nitin and Dr. Rohit Masih were involved in the clinical assessment and writing the case report. All authors read and approved the final manuscript.

Consent

Full written consent was received for the manuscript to be published.

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