COMMENTARY ARTICLE

The association of primary immunodeficiency's with autoimmune liver diseases is not rare

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ABSTRACT

Autoimmune Liver Disorders (AILD) are PIDs associated with autoimmune diseases; however, it is uncertain how common PIDs are in AILD patients. The purpose of this study was to assess how strongly AILD and PIDs are related. In a tertiary hospital, we carried out this single-center, cross-sectional, and descriptive study. Eighty-two AILD patients were examined for the presence of PIDs, including 39 with Autoimmune Hepatitis (AIH), 32 with Primary Biliary Cholangitis (PBC), seven with Variant Syndromes (VS), and four with Primary Sclerosing Cholangitis (PSC). The files provided us with a thorough history of illnesses, comorbidities, family history, and laboratory information. The immunology division reviewed each patient for further testing, and PID diagnoses were established in accordance with ESID (The European Society for Immunodeficiency's) standards. PIDs were discovered in 15% (82/82 patients) of patients with AILD, including 15 patients with SIgMD, 4 patients with partial IgA deficiency (PIgAD), 4 patients with Selective IgM Deficiency (SIgMD), and 3 patients with mixed immunodeficiency (CID). PIDs were found in 29% of VS patients, 25% of PSC patients, 23% of AIH patients, and 9% of PBC patients.

Keywords: *Primary immunodeficiency; Autoimmune hepatitis; Biliary cholangitis*

INTRODUCTION

The portal and caval systems are drained by the liver. As a result, it is exposed to a wide range of autoantigens, intestinal microbiota antigens, and dietary antigens. The liver can be viewed as a "tolerogen" rather than a "reactive" organ because of the liver's substantial antigenic stimulation but low immune response. Inherent immunological mechanisms in the liver strike a balance between protection and tolerance. Autoimmune Liver Diseases (AILD), such as Autoimmune Hepatitis (AIH), Primary Biliary Cholangitis (PBC), Primary Sclerosing Cholangitis (PSC), and Variant Syndrome (VS), are caused by a loss of balance. Our understanding of the aetiology of loss of self-tolerance is restricted to a few molecular and cellular pathways, such as altered gut-liver axis and Kupffer and T regulatory cell dysfunctions. Primary Immunodeficiencies (PIDs) are an uncommon and diverse set of hereditary illnesses that have an impact on the immune system's growth or operation. Up to this point, more than 450 PID-causing genes have been discovered. Allergy, lymphoproliferation, autoinflammation, and autoimmunity are other immune dysregulation manifestations that can show clinically as PIDs in addition to infections. According to reports, autoimmune illnesses affect 26.2% of PIDs. AILD is common in PIDs as well as other autoimmune illnesses. Therefore, in a subgroup of individuals, immunological dysregulation in AILD may be brought on by or made worse by underlying PIDs. However, it is uncertain how common PIDs are in AILD patients. According to our theory, immunological dysregulation in AILD patients may contribute to PIDs, which is why AILD patients have a greater prevalence of PIDs than the general population. Determining the frequency of PIDs in established AILD was our goal in order to test our hypothesis. Clarifying the link between AILD and PID would help define individualised treatment choices and provide a better understanding of AILD pathophysiology. A difficult diagnosis is concurrent PID and AILD. Due to modifications in immunological variables such Ig levels and autoantibody titers. Numerous case studies highlight how challenging it is to diagnose AIH using conventional diagnostic criteria because CVID patients' low IgG levels. Additionally, Fukushima et al. hypothesised that certain CVID cases labelled as "non-B nonC hepatitis" who responded well to immunosuppressive medication might actually have had AIH. Similar to this, some cases characterised in the literature as "chronic hepatitis" accompanying CVID may actually be AIH patients who also have accompanying PID.

CONCLUSION

We came to the conclusion that PIDs are more common (18%) in AILD patients, despite the fact that its overall estimated frequency in the general population is just 1 in 10,000. This is the first study to show that PIDs are underdiagnosed in AILD patients. In order to create novel, tailored treatment options, additional research is required to determine the prevalence and clinical implications of PID in AILD patients.

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