Gastrointestinal Manifestations of Autoimmune Diseases Requiring Critical Care

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KEYWORDS

- Autoimmune hepatitis • Autoimmune pancreatitis
- Catastrophic antiphospholipid syndrome • Systemic lupus erythematosus
- Immunosuppressive medications • Biological medications
- Disease-modifying medications

KEY POINTS

- Gastrointestinal organs have specific autoimmune diseases that can present to critical care, primarily the liver, pancreas, and intestines.
- There are systemic autoimmune diseases that present with gastrointestinal manifestations.
- Most interventions for autoimmune diseases are linked to immune pharmacology and decreasing circulating antibodies.
- Autoimmune diseases are often clinical diagnoses and require extensive laboratory testing.

INTRODUCTION

Gastrointestinal (GI) integrity, appropriate liver function, and beneficial bacterial flora are vital to overall immune system competence.[1] It is, therefore, not surprising that both autoimmune diseases (ADs) expressed within the GI tract and systemic ADs can acutely present signs and symptoms within the GI system. There are estimated to be 50 million persons in the United States with ADs.[2] Women predominate the statistics reported for each of the 80 to 100 AD spectra. This sex-associated incidence is attributed to multiple theories. One theme that is repeated through each theory is the high reactivity of the female immune system to circulating self-antigens. Additionally, AD often presents as 2 or more overlapping autoimmune processes within a single patient.[2] AD diseases are chronic and exist on a continuum of mild to severe. The intent

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of this article is to review the acute clinical presentations of each GI-associated AD and to focus on high acuity interventions. These interventions are primarily medical and grouped as antiinflammatory drugs, immunosuppression, disease-modifying medications, and biological pharmacology categories. The desired outcome of medication intervention is to decrease autoantibody formation and suppress subsequent generation of inflammatory cytokines. When caring for patients with ADs, the health care provider must focus on the correct diagnosis, correct drug, appropriate dose, and for a sufficient duration.

Three categories of GI patients require critical care for AD. These categories are organ-specific ADs, such as liver, pancreas, and intestinal. Next, there are clinical presentations of immune dysregulation, such as angioedema (AE) of the intestines leading to an acute abdomen. Finally, there are clinical presentations of systemic ADs that initially manifest with severe GI signs and symptoms. AD statistics in critical care populations describe that the frequent ADs requiring critical care were antiphospholipid syndrome (APS), systemic lupus erythematous (SLE), and rheumatoid arthritis (RA). Each of these AD types can have GI system manifestations. The mortality for patients with ADs in the critical care area ranged from 17% to 55% and was associated with high Acute Physiology and Chronic Health Evaluation scores.[3] Lungs are typically the first organ to fail in patients with ADs in critical care. Lung damage is attributed to the microcirculation damage from activated circulating cytokines.[3]

Gastrointestinal Autoimmune Diseases: Liver, Pancreas, Inflammatory Bowel Disease

Liver and normal immune function

Examining the immune role of the liver in the immune system reveals some unique aspects. The liver is more tolerant to antigens and also serves as the body's sentinel to preventing bacterial systemic spread. The highly vascularized liver is bombarded with blood-borne pathogens and antigenic substances from digestion. This workload of pathogen containment and antigen recognition is specific to the liver network of cells called antigen-presenting cells (APCs).[4] APC cells types include Kupffer cells, dendrite cells, and liver sinusoid epithelium cells. These APC cells defend the body from pathogens. Paradoxically, APCs also have the property of making the liver less immunologically reactive to antigens through T-cell suppression. In autoimmune hepatitis, for example, there is abnormal feedback or dysfunction in the APCs and T cells activate. T-cell activation leads to a production of a cascade of immune cytokines leading to hepatic cell destruction (**Fig. 1**, **Table 1**). The APC system is the reason that immunosuppressive medications can selectively be reduced after a liver transplant.[5]

Liver and autoimmune diseases

The 3 types of liver ADs are autoimmune hepatitis (AIH), primary biliary cirrhosis (PBC), and primary sclerosing cholangitis (PSC). Systemic immunoglobulin G4 (IgG4) disorder can also target the liver. AIH is a rare disease produced by the interaction between the environment and genetic vulnerability. The incidence of AIH is estimated to be about 1 per 200,000 cases in the United States, with most being women.[6] Twenty-five percent of AIH can present as acute liver failure to the critical care unit. Acute liver failure is typically recognized as a decreased level of consciousness, elevated liver enzymes, respiratory failure, and metabolic acidosis. Acute AIH is often refractory to the effects of steroids.

Severe AIH is a clinical diagnosis. Supporting patterns of findings are liver-biopsy-identified centrilobular zone 3 necrosis, autoantibodies, and hypergammaglobulinemia. AIH must be differentiated from acute viral hepatitis and drug-induced liver injury (DILI) through a scoring system of probability and tissue biopsy findings.[7] Treatment