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Editorial

# Immunoproliferative Small Intestinal Disease: Infectious Pathogen Associated Human Lymphoma

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## DESCRIPTION

The term small intestinal immunoproliferative disease (IPSID) is adopted by World Health Organization (WHO) in 1978. This IPSID is a type of lymphoma that occurs in the small intestine in the lymphoid tissue associated with the mucosa. This type of lymphoma is characterized by a large number of plasma cells infiltrated in the intestinal wall. These plasma cells exude alpha-immunoglobulin heavy chains but are not content with any immunoglobulin light chains. Most cases are characterized by the loss of the ability to synthesize light chains and the discharge of defective heavy chains.

Its histopathological feature is that lymphocytes and plasma cells infiltrate the wall of the small intestine. Infiltration produces abnormal immunoglobulin (IgA), which is a truncated alpha heavy chain with no light chain component.

Small intestinal Immuno-proliferative disease (IPSID) sometimes may resemble celiac disease (an autoimmune disease), tropical stomatitis diarrhea, or parasitic infection and the patient may have long-term emaciation (extreme weight loss) or diarrhea are due to mal-absorption syndrome (unable to absorb nutrients) and protein-losing enteropathy (Protein-losing enteropathy is a pathological condition in which there is an increased loss of proteins through the gastrointestinal tract, which leads to low serum proteins.) as a secondary symptom of lymphoma infiltrating the intestines. Most patients appear in the early stages and can be cured with oral antibacterial drugs. IPSID is considered a unique subtype of extra-nodal marginal zone B-cell lymphoma. This disease is also called Mediterranean lymphoma because these types of immunoproliferative diseases have been widely reported in the Mediterranean Basin, the Middle East, the Far East and Africa. In the Middle East, the most common site for extranodal lymphoma in the gastrointestinal tract: immunoproliferative disease of the small intestine accounts for about one-third of gastrointestinal lymphomas. The limited geographic distribution of the disease leads to the hypothesis that environmental factors may have a pathogenic effect.

IPSID (Immunoproliferative small intestinal disease) has been suggested to occur in patients with recurrent or persistent intestinal infections, causing chronic antigenic stimulation of the lymphoid tissues secreting IgA at this site, and the resulting clone population acquires mutations that cause heavy chain production, as described above. Subsequently, most cases are characterized by a loss of ability to synthesize light chains. In our case, immunofixation (measures certain proteins in the blood) of serum protein electrophoresis was performed, and the M component is typically IgA, and there is no corresponding light chain.

In early-stage immune proliferative disease of the small intestine generally responds to antibiotics. This indicates that it may be caused by a bacterial infection. Trying to use standard culture methods to identify pathogens is not very helpful. However, some studies have shown that H. pylori (a type of bacteria) and *C. jejuni* (a human diarrheal pathogen) may be potential pathogens.

Serum protein electrophoresis (SPE) with immunofixation and serum antibodies to tissue transglutaminase (an enzyme that catalyzes the formation of isopeptide bonds between proteins, TTG) is important diagnostic tests that distinguish these two entities. Patient demographics and responses to gluten-free diets also provide clues. High-grade lymphoma can be related to any type of disease, but the lymphoma that complicates celiac disease is a T-cell lymphoma that easily causes perforation (a hole that develops through the wall of a body organ).

If left untreated, the disease can turn into high-grade lymphoma. It can cause severe morbidity, mal-absorption and protein-losing of the intestine. Rapid treatment is important to control the mal-absorption and prevent the progression of the disease.

It can be difficult to distinguish non-specific chronic inflammation from early IPSID, especially in small biopsies. Conducive to IPSID, it is a dense, predominantly plasma cell infiltration that distorts the mucosal structure of patients with IPSID compatible clinical features. Immunohistochemical staining showed that only plasma cells expressing the heavy chain confirmed the diagnosis.

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