Refractory Celiac Disease

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Abstract:
Refractory celiac disease (RCD) is when malabsorption symptoms and villous atrophy persist despite strict adherence to a gluten free diet (GFD) for more than 12 months and other causes of villous atrophy have been ruled out. RCD is considered a rare disease and almost exclusively occurs in adults. Persistent diarrhea, abdominal pain, weight loss are the most common symptoms in RCD. Also, anemia, fatigue, malaise, thromboembolic events and coexisting autoimmune disorders are frequent.

Diagnosis of RCD is based on other causes of unresponsiveness to the GFD, particularly collagenous sprue, ulcerative jejunitis, and enteropathy-associated T-cell lymphoma. Many disorders such as autoimmune enteropathy, tropical sprue, common variable immunodeficiency, and intolerance to non-gluten dietary proteins may have similar histological findings but not necessarily identical with CD and therefore should be excluded. Repeat intestinal biopsy may help to differentiate causes of non-responsive CD associated with ongoing villous atrophy (e.g., gluten contamination, small-bowel bacterial overgrowth, RCD).

There are 2 subtypes of RCD according to absence (type I) or presence (type II) of an abnormal intraepithelial lymphocyte population. RCD type 1 usually becomes better with a combination of aggressive nutritional support, adherence to GFD, and pharmacologic therapies such as prednisone, budesonide and azathioprine. For RCD type 2, more aggressive therapeutic approach is needed since clinical response to therapies is less certain and may evolve into aggressive enteropathy associated T-cell lymphoma and the prognosis is poor.

Key words: Celiac Disease, Refractory.

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