

Celiac Disease with Concurrent Stiff Person Syndrome

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Introduction

- Celiac disease (CD) is an immune-mediated gastrointestinal disease with a prevalence of 1% in the general population.
- It is instigated by the ingestion of gluten in genetically predisposed individuals and has strong associations with HLA-DQ2 and DQ8.
- It classically manifests with gastrointestinal symptoms, however, extra-intestinal symptoms including neurologic and dermatologic findings such as ataxia and dermatitis herpetiformis can occur.
- Stiff person syndrome (SPS) is a rare, relapsing-remitting, neurological disorder characterized by truncal and proximal limb muscle rigidity and spasms.
- We report a rare case of celiac disease in concurrence with stiff person syndrome and review additional autoimmune associations.

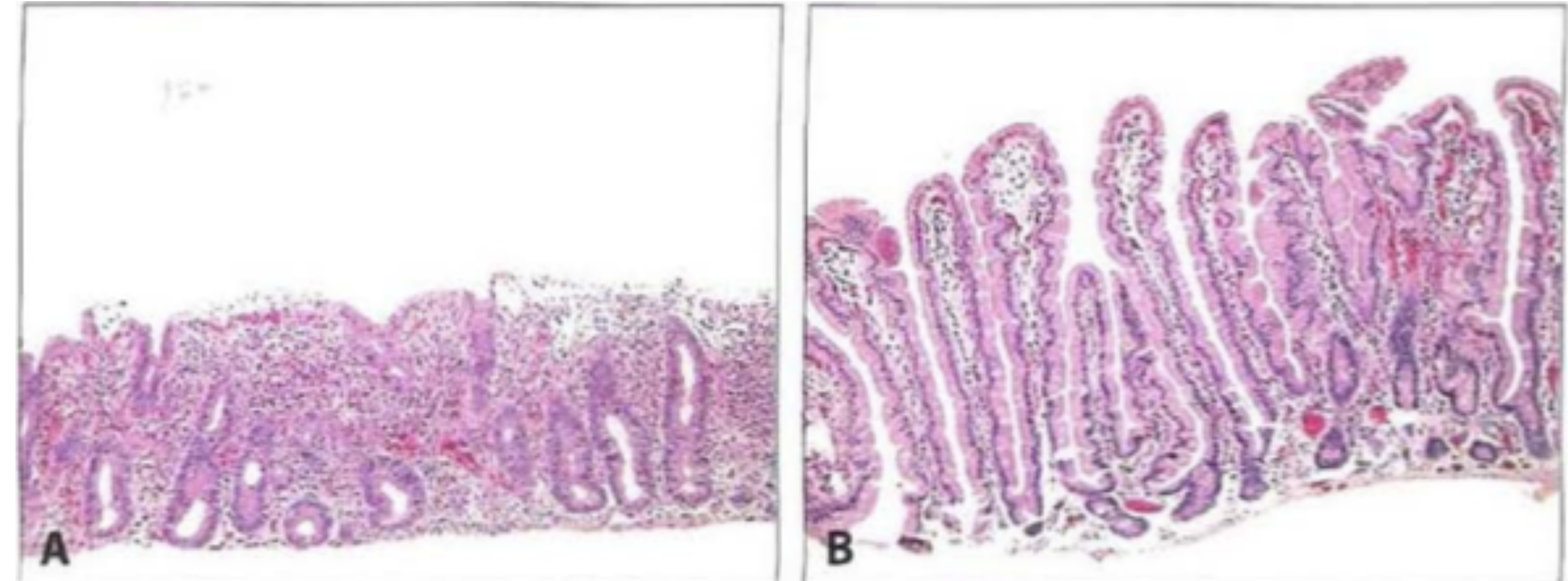
Case Presentation

A 35-year-old white female with a past medical history of celiac disease, hypothyroidism, and polycystic ovarian syndrome presented with an acute flare of stiff person syndrome. Her symptoms included painful, dystonic posturing in all extremities as well as abdominal spasms and sustained contractions for one week. She was admitted to the neurology service and was treated with plasma exchange and baclofen. Gastroenterology was consulted due to one episode of rectal bleeding with associated abdominal pain and bloating following gluten ingestion. She was placed on a strict gluten-free diet, which resulted in resolution of rectal bleeding and as well as continued improvement of neurologic symptoms.

References

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Images



A: Celiac disease, flattened villi.

B: Normal villi

Discussion

- Neurological manifestations of celiac disease have been described in the literature. These include gluten ataxia, restless leg syndrome, as well as other movement disorders such as SPS.
- Celiac disease and stiff person syndrome are strongly associated with other autoimmune diseases.
- The central genetic factors lie on the major histocompatibility complex. HLA alleles linked with celiac disease are also associated with stiff person syndrome and type I diabetes.
- Autoantibodies, specifically against glutamic acid decarboxylase, gliadin, and tissue transglutaminase, are theorized to have a significant role in these disease mechanisms.
- This case highlights the importance of a thorough gastrointestinal diagnostic workup in patients with stiff person syndrome and other movement disorders because of the concomitant nature of autoimmune diseases and disorders.
- Immune responses in celiac disease may be implicated in the pathogenesis of concurrent SPS and controlling CD may be helpful in resolving and preventing exacerbations of neurological diseases.