A Rare Case of Suspected Non-Celiac Gluten Ataxia:

Authors: Victoria S. Louwagie, DMSc, MS, PA-C1, Kari J. Mongeon Wahlen, APRN, CNS, MSN2, Paola Sandroni, MD, PhD2

1 Division of Gastroenterology and Hepatology, Mayo Clinic Health System Mankato, Minnesota

2 Division of Neurology, Mayo Clinic Health System , Mankato, Minnesota

INTRODUCTION

We present a unique case of an otherwise health 35-year-old male who despite negative tTg antibodies and negative celiac gene pair, consistently experienced ataxia within minutes to hours of consuming gluten.

PURPOSE

Physician Associates should be aware of gluten related disorders beyond simply celiac disease. Gluten ataxia (GA) is a rare neurological gluten-related condition mediated by an autoimmune response that damages the cerebellum when gluten is ingested. GA specifically results in ataxia and if not diagnosed effectively managed, can lead to permanent disability. Gluten related disordered and gluten sensitivities (GS) encompass a broad spectrum of immune-mediated manifestations triggered by dietary gluten. Gluten ataxia can be found in patients without celiac disease (CD). Less than 10% of patients with GA present with gastrointestinal symptoms. Up to 30% of patient with GA do not have HLA-DQ2.

Case

A 35-year-old male presented to his primary care provider with a two-year history of intermittent dizziness after eating. Postprandial hypotension or hypoglycemia were initially suspected, but further testing disproved those. Symptoms were more consistent with vertigo and persisted even when supine and with eye closed. Patient experienced dysmetria in addition to gait disturbance during these episodes.

Symptoms occurred within 10 minutes of eating and would resolve within hours of fasting. Events did not occur consistently with eating. There were occasional diarrhea, diaphoresis and heartburn symptoms at time of events, but no unintentional weight loss, fever, night
sweats, cough, shortness of breath, itching, dysphagia, rash, hair changes, nasal congestion, or headache.

He had no anemia or electrolyte disturbances. He was referred to the Gastroenterology (GI) Clinic for further evaluation and management of GS or CD. The patient self-initiated a strict food elimination diet avoiding wheat, dairy, caffeine, and processed sugars. He primarily ate meats, nuts, vegetables, corn, and rice. During this period, he took meticulous notes with complete resolution of symptoms on this diet. When he started to reintroduce items, he realized reintroducing even the smallest amount of gluten, resulted in a return of his symptoms. By the time of GI appointment, he had been symptom free for over 2 months on a GFD.

Physical examination revealed no abnormalities.

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Non-Celiac Gluten Ataxia

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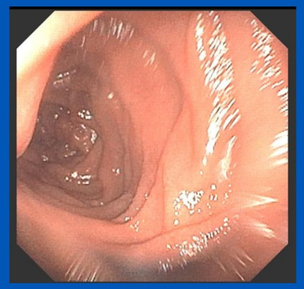
Duodenal Bowel Image and Celiac Gene Pair



CELIAC ASSOC.	
Celiac Gene Pairs Present?	No
Celiac Interpretation	see below
HLA-DQB1 Locus Molecular	03:01, 06:03
HLA-DQA1 Locus Molecular	01:03, 03

Diagnostic Testing

Diagnostic testing was comprehensive. Including CBC, point of care glucose, CMP, iron level, ferritin, TIBC with% saturation, vitamin-A level, vitamin-E level, vitamin B12 level, thiamine level, TSH, chromogranin a level, gastrin level, parathyroid hormone, A1c, GAD65 Ab Assay, insulin antibody, celiac serologies, celiac gene pair assessment, heavy metal screen which included testing for arsenic, cadmium, copper, lead, mercury and zinc. All of these diagnostics were essentially within normal limits or unrevealing. Upper endoscopy duodenal biopsy was essentially unrevealing.

patient also underwent CT of the abdomen with enterography which was Unremarkable. MR brain with and without IV contrast unremarkable. Echocardiogram was positive for a small atrial level shunt with Valsalva. Gastric emptying solid phase testing is pending.

Patient was seen by a neurologist and an allergist/immunologist. It was the Allergist/Immunologist who astutely diagnosed suspected non-celiac gluten ataxia.

Conclusion

Gluten ataxia is extremely rare. The diagnosis is not straight forward, even with on elevated anti-tTg IgA antibodies, negative small bowel biopsy and lack of gene pair but who have a strong clinical scenario for GA should be placed on a GFD indefinitely. Even with the recent description of transglutaminase 6, there is still no clear biological marker for the disease.

Stabilization/improvement of the ataxia after 1 year is a strong indicator that the patient suffers from GA. Gluten sensitivity or CD should be considered in the diagnostic workup of movement disorders (MDs) of unknown etiology in all patients.