Posted on Authorea 23 May 2022 | The copyright holder is the author/funder. All rights reserved. No reuse without permission. | https://doi.org/10.22541/au.165330015.59870220/v1 | This a preprint and has not been peer reviewed. Data may be preliminary.

Chorea revealing celiac disease: cause or coincidence?

Mariem Mhiri[1], Anis Hassine[2], Rihab Ben Dhia[1], Salma Naija[2], Mouna Aissi[1], Sana Ben Amor[2], and Mahbouba Frih-Ayed[1]

[1]Fattouma Bourguiba University Hospital of Monastir
[2]Sahloul University Hospital

May 23, 2022

Abstract

A number of neurological disorders have been reported to be associated with celiac disease.We reported the case of a patient aged 43 years presents for choreic movements. All the etiologic report was negative except serology of celiac disease positive etiology and gastric biopsy confirming the disease

Chorea revealing celiac disease: cause or coincidence?

Mariem Mhiri [1], Anis Hassine [2], R.ben Dhia[1], Salma Naija [2] ,Mouna Aissi [1],Sana BenAmor [2], Mahbouba Frih-Ayed[1]

[1]Fattouma Bourguiba Hospital, neurology, Monastir/Tunisia

[2] Sahloul university hospital, neurology, Sousse/Tunisia

Author correspondant: Mariem Mhiri

Tel: +21699115114

Mail: mariemmhiri@yahoo.com

Abstract

Introduction: A number of neurological disorders have been reported to be associated with celiac disease including ataxia, epilepsy, peripheral neuropathy and rarely chorea.

Case report: I reported the case of a patient aged 43 years presents for choreic movements of brutal appearance. All the etiologic report was negative except serology of celiac disease positive etiology and gastric biopsy confirming the disease.

The evolution was marked by the disappearance of chorea following the gluten-restricted diet.

Discussion: Despite the nature of association is unclear, celiac disease may be a treatable cause of chorea.

Keywords : Chorea, celiac disease, anti-gliadin antibodies.

Competing interests: No conflict of interest.

Introduction: The incidence of neurological in celiac disease or associated with gluten-sensitivity without gastrointestinal complaints was 10% (1). Cerebella ataxia and peripheral neuropathy was considered to be the commonest neurological symptoms of gluten sensitivity (2). But, these manifestations include epilepsy, myoclonus,dementia, and some cases of chorea are described.

Posted on Authorea 23 May 2022 | The copyright holder is the author/funder. All rights reserved. No reuse without permission. | https://doi.org/10.22541/au.165330015.59870220/v1 | This a preprint and has not been peer reviewed. Data may be preliminary.

The onset of chorea in adulthood has many causes, such as metabolic, genetic, and autoimmune factors. Huntington's disease is the commonest (3). Despite, a patient chorea free of family history of movement disorders or dementia and association with autoimmune disease may suggest diagnosis of celiac disease. It's so easy when a patient has gastrointestinal complaint. So, our patient is original case.

Case Report

A 38-year-old woman was referred for evaluation ofabnormal movements ofarms and legs. It was suddenly installed since one month and rapidly progressive. In her history, she had one fœtal death in utero and had a brother with diabetes mellitus type 1. She was on no medication.She complained from chronic headache but she hadn't any digestive symptome such as diarrhea or abdominal pain. On examination, she had a chorea on her four members (see video, segment1) without other neurologic syndrome. She had a BMI at19 and no othersystems' abnormality.

Laboratory evaluations showed none absorption syndrome with mixed deficiency anaemia, hypoproteinemia and hypoalbuminemia without proteinuria, hypocalcaemia,vitamin D deficiency and some episodes of hypoglycaemia.Whereas, she had normal blood level of magnesium, triglyceride, cholesterol and homocysteine and normal creatin kinase and thyroid function. On the other hand, she had hypogammaglobulinemia, small cytolysis and no lymphopenia.

Autoimmune research showed the presence of IgG anti-reticulin antibodyin indirect fluorescent antibody assayon liver on the surch of antinuclear antibody.IgA anti-endomisium antibodywas found on stomach by the same assay. By technique of ELISA, we detected height levels of IgA and IgG of anti- desamidaded peptide of gliadine and IgA of anti-transglutaminase.IgA anti-phospholipidantibody had firstly positive with no significant level but was negative on the control after 3 months.CD was diagnosed and we completed by a small bowel exploration. Endoscopy was normal without villous atrophy but biopsy showedsub-total villous atrophy, intraepithelial lymphocytes (IEL), villous height to crypt depth ratio,which are specific of CD. Abdominals can founds omemesenteric adenopathy without digestive thickening or mass. No vascularitis nor cerebral venousthrombosis was detected by MRI with vascular exploration. Genetic study of Huntington's disease was negative. No etiology for chorea was diagnosed other than CD spetially Wilson disease and neuroancothocytosis was eliminated by laboratory evaluations. Treatment was started with low doses of neuroleptic and gluten free diet was started one week later. Celiac serology was negative for her three kids and her brother with diabetes mellitus. The patient had difficulties for gluten free diet's compliance. Evolution was partial regression of chorea and headache. Two months later, she developed extrapyramidal syndrome because of neuroleptic treatment

Discussion

The main evidence suggesting CD as the etiology of this patient's choreais the none improvement seen with neuroleptic and the large negative exploration. Certitude diagnosiswill be based on the improvement of chorea after strict gluten free diet.

Case reports of the benefit to neurological symptoms of a gluten-free diet are inconsistent, but the only systematic controlled study of the effect of a gluten-free diet did show neurological improvement(4).In that study,43 patients with ataxia attributed to celiac disease were identified and studied between 1996 and 2003.Twenty six patients (treatment group) adhered to the gluten-free diet and had evidence of elimination of antigliadinantibodies by one year. Fourteen patients refused the diet (control group). Three patients had persistently raised antigliadin antibodies despite adherence to the diet and were there for eexcluded from the analysis. After one year there was improvement in ataxia reflected in all of the ataxia tests in the treatmentgroup. This was significant when compared with the control group (4).

Only two publications about chorea attribuated to celiac disease was found in the litterature. The first was a series of four cases published on 2004 from United Kingdom (1). Theywere four women aged between 48–77, with no significant family history of the disease. Chorea was associated with otherneurologic manifestations and had an other wiseun explained neurological disorder. Diagnosis of coeliac disease has been made on

Posted on Authorea 23 May 2022 | The copyright holder is the author/funder. All rights reserved. No reuse without permission. | https://doi.org/10.22541/au.165330015.59870220/v1 | This a preprint and has not been peer reviewed. Data may be preliminary.

serological and/or biopsy grounds and who have an other wiseun explainedneurologicaldisorder. In all cases, there was clear improvement in chorea and ataxia with treatment with a glutenfreediet. They suggest that gluten sensitivity may cause chorea in some patients(1).

Next publicationwas a case from United States of Americareported on 2011(3).

He was a 59-year-old man. As a childhehad been diagnosed with CD. He followed a gluten-free diet as a childbut as an adulthe returned to a normal diet. He reported recurrence of abdominal pain and diarrhea. Involuntary movements were noted. On examination he had generalized chorea with dystonic facial movements and right hand posturing. IgG and IgA anti-gliadinantibodies were positifs. He was advised to adhere to a gluten-free diet.Three months later his involuntary movements weres ignificantly reduced and antigliadinantibodies level decreased. This case provides further evidence supporting celiac disease as a potentially reversible cause of chorea(3).

Conclusion

Our case provides evidence supporting that celiac disease is as reversible cause of chorea. Determination of gluten sensitivity shoud be considered in patients with chorea of indetermined etiology espetially if had a concurrent gastrointestinal symptoms.

References

(1)Pereira AC, Edwards MJ, Buttery PC, et al. Choreic syndrome and coeliac disease: a hithertoun recognised association. MovDisord 2004;19:478–482

(2)Khalafalla O, Bushara. Neurological presentation of coeliac disease. Gastroenterology 2005 ;128 :S92-S97.

(3)Walker RH and al, Chorea in CeliacDisease, TremorOtherHyperkinetMov 2011;1:*http://www.tremorjournal.org/article/view/32*

(4)Hadjivassiliou M, Davies-Jones GA, Sanders DS, et al. Dietary treatment of gluten ataxia. J NeurolNeurosurgPsychiatry 2003;74:1221–1224.

(5) Holmes. Neurological complications of celiac disease. Postgrad med 2002 ;78 :393-398.