Rapidly Progressive Cerebellar Ataxia in a Case of Unsuspected Celiac Disease: Early Diagnosis Leads to Reversibility

ARVIND VYAS*, DIVYA GOEL†

ABSTRACT

Subacute late-onset cerebellar ataxia in a patient can be due to varied causes. The common ones are infectious, drug-related, autoimmune and paraneoplastic pathologies. As majority of these causes are deemed treatable, they should be investigated in a sporadic case of ataxia. Celiac disease can have neurological complications in about 10% of cases but those are usually a secondary feature of this disease. This case report features a case of rapidly progressive cerebellar ataxia as the sole presentation of unsuspected celiac disease.

Keywords: Cerebellar ataxia, rapidly progressive, celiac disease

Cerebellar ataxia can be classified according to the age of onset. Its occurrence below 40 years of age is considered early-onset and above this is labeled late-onset ataxia. Another type of classification is done by the type of symptom onset- sudden, acute, subacute or chronic. Where sudden cerebellar ataxias of late-onset are usually due to stroke and chronic ones are mostly due to degenerative causes, the acute and subacute ones are those, which may have a treatable etiology.

The commonly postulated causes of late-onset cerebellar ataxias are infectious (e.g., prion disease), drug-related (e.g., phenytoin), autoimmune (e.g., anti-glutamic acid decarboxylase [GAD], steroid responsive encephalopathy with autoimmune thyroiditis, gluten sensitive ataxia) and paraneoplastic. A battery of tests is needed to arrive at the correct diagnosis. Celiac disease is one such cause and is rendered treatable if neurological symptoms have not advanced much. It presents with cerebellar ataxia in about 10% of patients. The ataxia is usually insidious in onset and follows abdominal symptoms. But rare presentations, like rapidly progressive cerebellar ataxia with lack of abdominal symptoms, need to be screened for autoimmune and paraneoplastic causes.

CASE REPORT

A 58-year-old male presented with 20 days history of vomiting, headache, vertigo and subacute onset ataxia, which progressed rapidly over next 5 days. A week later, he noticed some slurring of speech but his ataxia remained static after initial progression. He had no accompanying history of any drug intake, fever, weight loss or headache. He cleared his bowel twice everyday with no change in frequency or consistency of his stools. He did not have any significant past or family history.

His general examination was unremarkable. Positive findings in his neurological examination were those of asymmetrical cerebellar signs in the form of intention tremors, dysdiadokokinesia and mild dysmetria more prominent on right. His speech was cerebellar and gait ataxic. Ocular examination did not reveal any nystagmus. His higher mental functions, motor and sensory examination were unremarkable. Initially on the basis of his rapidly progressive symptoms and subsequent achievement of a static course, we thought it of as cerebellitis.

His routine laboratory investigations were normal. Thus, we conducted magnetic resonance imaging (MRI) brain and cerebrospinal fluid (CSF) examination, both of

*Senior Professor
†Senior Resident
Dept. of Neurology
SMS Medical College, Jaipur, Rajasthan
Address for correspondence
4-Ba-19, Jawahar Nagar, Jaipur, Rajasthan - 302 005
E-mail: drdivyagoel@hotmail.com

which proved out to be normal. Thus, we thought of ruling out autoimmune and paraneoplastic causes. He was started on injectable methylprednisolone in the meantime but without any improvement in his symptoms, leading us to stop it after 3 days. His antinuclear antibody and tumor markers were negative, thyroid profile including antithyroid peroxidase antibodies were normal. Anti-tissue transglutaminase (anti-tTG) titer came out to be positive, thus we subjected the patient to intestinal biopsy for confirmation. He was commenced on a gluten-free diet and was discharged in a stable condition with his biopsy report awaited. He came for follow-up after 10 days with improvement in ataxia and his histopathology report showed changes in favor of celiac disease type 3a, according to the modified Marsh classification.

DISCUSSION

Celiac disease is an autoimmune enteropathy due to gluten sensitivity manifesting as diarrhea, steatorrhea, weight loss,[1] and occasionally neurological symptoms such as cerebellar ataxia, peripheral neuropathy, myoclonus, chorea, palatal tremor and opsoclonus-myoclonus.[2] Cerebellar ataxia is one of the most common presentations of celiac disease as is shown in previous studies, but majority of patients have a long-standing celiac disease before the onset of neurological symptoms. This case has been reported owing to the subacute onset and rapid progression of ataxia in a patient of unsuspected celiac disease, though similar cases have been reported in literature.

A case presented with rapidly progressive ataxia, dysarthria and bilateral lateral rectus palsy in the presence of minimal abdominal symptoms. He was worked up for celiac disease and was found to be positive but had a fulminant course of his symptoms, not responding to gluten restriction, steroids or other immunosuppressants, culminating into death due to myocardial infarction.[1] The mean age of gluten ataxia is 53 years and has no gender predominance. There are many mimickers of gluten ataxia in this age group, including paraneoplastic cerebellar degeneration, anti-GAD ataxia and cerebellar variant of multiple system atrophy (MSA-C). The rare presentation of rapidly progressive ataxia, as is seen in the reported case, mimics paraneoplastic cerebellar degeneration and anti-GAD ataxia. The search for primary and tumor markers needs to be conducted. Lack of autonomic dysfunction differentiates it from MSA-C.[2]

The mechanism of neurological complications in celiac disease is yet uncertain. The proposed mechanisms are: (1) Malabsorption of various neuroprotective nutrients and (2) antigliadin antibody neurotoxicity.[3] There are experiments proposing that Purkinje cells in cerebellum share the same antigen epitope as the gluten peptides resulting in cerebellar involvement.[2] Thus, a gluten-free diet can actually benefit the patients of gluten ataxia.

This case study highlights the importance of rare and atypical presentation of celiac disease and early screening for antibodies followed by biopsy confirmation, which can lead to reversibility of the neurological symptoms of this disease.

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Nutrition

Rule of 7

Consume at least 7 serving of fruits and vegetables in a day incorporating all 7 colors.

Formula of 30

- Normal requirement of water is 30 mL/kg body weight/day.
- Normal caloric requirement is 30 Kcal/kg body weight/day.

Formula of 500

- You eat 500 calorie extra when you take an 11 inch plate instead of 8 inch plate.
- You eat 500 calorie extra when you prefer a buffet over *à la carte*.
- You eat 500 calories extra when you eat in a party or a restaurant compared to eating at home.

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