CASE REPORT

Celiac Disease Associated with Recurrent Guillain Barré Syndrome

VIJAY GUPTA AND A KOHLI

From the Department of Pediatrics and Neonatology, Dr Ram Manohar Lohia Hospital, New Delhi , India.

Correspondence to:
Dr Vijay Gupta, C/o Chaudhary Traders, Dal Bazaar, Lashkar, Gwalior 474 009, MP, India.
yzaygupta@gmail.com
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Celiac disease is associated with multiple extraintestinal presentations, including bone disease, endocrine disorders and neurological deficits. We report a 9 year old girl with celiac disease presenting with recurrent Guillain Barre syndrome (third episode). There was no other clinical manifestation except for refractory iron deficiency anemia. Molecular mimicry explaining the association between these two disorders, is far more interesting.

Key words: *Celiac disease, Recurrent, Guillain Barre Syndrome*.

The mode of presentation of celiac disease can be quite variable(1). We present a case of recurrent Guillain Barré syndrome (GBS) with meningismus which is rare in children. Adding to its rarity is its association with celiac disease with no other manifestation except for refractory iron deficiency anemia.

CASE REPORT

A nine year old female child was admitted with acute flaccid paralysis of all four limbs. She also had headache and pain in the neck and thigh muscles. She had two such episodes of similar illness in the past, first at the age of 6 yr and another at the age of 8 y. Previous two episodes were diagnosed Guillain Barré Syndrome (on the basis of clinical, laboratory and nerve conduction studies). She had near about complete recovery from previous two episodes in 6-10 weeks period. This time also child was diagnosed as case of recurrent Guillain Barré syndrome (third episode) on the basis of clinical picture, CSF examination (3 cells mainly lymphocytes, protein 92 mg%, sugar 59 mg%), and nerve conduction studies (axonal and demyelinating neuropathy, predominantly demyelinating). She also had pallor with signs of meningeal irritation. Intravenous immunoglobulin was given for 5 days. Child started showing signs of recovery and power improved in the 2nd week of illness. The child was having concomitant iron deficiency anemia, despite having received several hematinics for last several months (Hb 8.2g/dL, total iron 10.0µg/dL, TIBC 516 µg/dL, transferrin saturation 3%) .

Child was investigated for celiac disease. Her tissue transglutaminase level was 97.55 U/mL (normal <4.00). Duodenal biopsy revealed villous atrophy, crypt elongation, increased intraepithelial lymphocytes and the girl was diagnosed as having celiac disease type 3b(2). Stool culture, serology and PCR was negative for *Campylobacter*. Anti-ganglioside antibody could not be done for financial constraints.

Child was started on strict gluten free diet along with hematinics and other multivitamins. Followed up at around three months, she had almost complete recovery from the weakness and also had normal iron stores (Hb 10.9 g/dL, total iron 56.0 µg/dL, TIBC 310 µg/dL, transferrin saturation 22 %).

DISCUSSION

Celiac disease is associated with multiple extraintestinal presentations, including bone disease,

endocrine disorders and neurological deficits(1). Neurological disorders include cerebellar ataxia, peripheral neuropathy, epilepsy, dementia, migraine, encephalopathy and Guillain-Barré like syndrome(3).

Recurrent GBS is a rare condition characterized by 2 or more attacks of acute inflammatory demyelinating neuropathy with an onset to peak time of 4 weeks or less having complete or near complete recovery(4,5). Acute onset, frequent facial involvement, brief clinical course, near complete recovery and very long asymptomatic periods may distinguish these patients of acute relapsing demyelinating polyneuropathy (ARDP) from chronic relapsing demyelinating poly-neuropathy(6).

Celiac disease presenting as acute flaccid paralysis is rare. Both have autoimmune background, which may explain their linkage. Recent studies have shown a significant correlation between anti-ganglioside antibodies, GBS, and neurological disorders in patients with underlying celiac disease. Gangliosides are abundant in the nervous system and in gastrointestinal tract(7). It is not known what triggers the release of anti-ganglioside antibodies in people with gluten sensitivity. But, the mechanism is likely to involve the intestinal immune system response to ingested gliadin, a component of wheat gluten. Two mechanisms have been postulated for the release of anti-ganglioside antibodies: one is the presence of ganglioside-like epitopes in gliadin and the other is the potential for complex formation between gliadin and GM1 ganglioside(3).

Certain gliadin spices are reported to be glycosylated. but they do not appear to carry GM1-like carbohydrate moieties(8). In contrast, *in vivo* formation of gliadin-GM1 complexes is probably feasible, since abundant GM1 is found in gut epithelial cells(8). Anti-ganglioside antibody formation in celiac disease may play a role not only in developing neurological complications of celiac patients, but also in developing celiac disease itself(3).

About 25% of patients with GBS have a recent *Campylobacter jejuni* infection. The lipooligosacharide located in the wall of *Campylobacter jejuni* cross-reacts with ganglioside in axonal membrane of neurons. Gangliosides-like epitopes common to both lipopolysacharide coats of certain strains of *C. jejuni* and gangliosides in cell structure of gastrointestinal mucosa may cause an autoimmune response. This may lead to atrophy and degeneration of mucosa possibly by apoptosis in a manner similar to nerve tissue injury in GBS. The proposed mechanism can also explain the presence of neurological manifestations of celiac disease(3). However, molecular studies need to be conducted to evaluate further the association between celiac disease and GBS.

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