A CASE OF CELIAC DISEASE PRESENTING WITH RECURRENT ATTACKS OF QUADRIPARESIS

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Abstract

Celiac disease, once considered a rare disease in Bangladeshi population, have recently been reported more frequently. Recognition of celiac disease is not easy and a high degree of suspicion in required. Although the disease presents usually with malabsorption syndromes, atypical presentations also may occur which may cause confusion in making a prompt diagnosis. We intend to report a case who presented primarily with flaccid type of quadriparesis who ultimately was proven to be a case of celiac disease. Although neurological symptoms have been associated with celiac disease, motor weakness is very uncommon. We have proposed that in our case electrolyte imbalance have caused or contributed in causing this motor weakness.

Keywords: *Celiac disease, Recurrent weakness*

J Dhaka Med Coll. 2016; 25(2) : 138-141

Introduction

Celiac disease, once considered a rare disease in Bangladeshi population, have recently been reported more frequently.[1] Recognition of celiac disease is not easy and a high degree of suspicion in required.[2] Although the disease presents usually with malabsorption syndromes, atypical presentations also may occur which may cause confusion in making a prompt diagnosis. We intend to report a case who presented primarily with flaccid type of quadriparesis who ultimately was proven to be a case of celiac disease.

Case report

A Bangladeshi Muslim male, aged 22 years, non-diabetic, normotensive patient was admitted in the hospital on 16 March 2016 with the complaints of chronic diarrhea for 2 months, weakness of all four limbs and muscle cramp for same duration. he had been previously admitted on 01 March 2016 in a clinic with above complaints along with flaccid quadriparesis. There his serum electrolyte profile revealed severe hypokalemia (his electrolytes level are given in the Table 1). They attributed his weakness to hypokalemic periodic paralysis as the patient gave history of recurrent episodes of concomitant diarrhea and such quadriparesis and treated him. After some recovery of limb functions, they referred the patient to DMCH for further management and to search for specific cause of hypokalemia because of the severity and persistence of symptoms. The patient also reported about loss of 10 Kg weight within 2 years.

The patient had no relevant family history. He is non-smoker, non-alcoholic and used to live for 2 years in Malaysia 1.5 year back as an immigrant student. His HIV p24 Ag and HIV Ab tests were done which were negative. He had consulted his gastroenterologist 7 months back and had been suspected and treated for tropical sprue, but no significant improvement was seen. His thyroid hormone profile had been done which had been within normal range excluding hyperthyroidism induced diarrhea.

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Received: *01 July 2016* ***Accepted:*** *01 September 2016*

Table 1
Changes in serum electrolytes over time.

Serum electrolytes	12/03/16	13/03/16	14/03/16	15/03/16	16/03/16	27/03/16
Sodium (mmol/L)	133	136	133			138
Potassium (mmol/L)	1.3	1.5	1.6			3.8
Calcium (mg/dL)					7.3	
Magnesium (mg/dL)	1.1			1.5	1.6	
Chloride (mmol/L)	97	100	100			104

On admission, his general examination revealed: pulse-88/min, blood pressure-100/60 mm-Hg, temperature- 98.2º F, respiratory rate- 13 breaths/min, mildly anemic, non-icteric, ill-looking appearance. His nervous system examination revealed- diminished limb jerks along with muscle power (3/5) on all four limbs. Other systemic examinations were non-remarkable.

His investigation findings were, Hb- 11.8mg/dL, WBC- 12,600/mm^3, ESR - 25mm, USG of whole abdomen- gall bladder sludge, chest x-ray P/A view- normal study, urine R/E and urine culture- normal, fasting blood glucose - 4.9 mmol/L and 2 hours after 75 g glucose intake level- 5.7 mmol/L. Serum albumin 2.6 g/dL, serum creatinine-0.84, SGPT – 65 u/L. His investigation profile suggested, he was found with persistent low level of several electrolytes such as sodium and potassium, (shown in Table 1), but his urinary electrolyte levels were all lower than the normal range. Which signifies that the loss of electrolytes was not due to urinary loss, but considered together with his history of recurrent diarrhea, they were due to gastrointestinal loss. To assess the possibility of several diseases causing different malabsorption syndromes, stool R/E, endoscopy and colonoscopy etc. were done. Stool R/E showed only mucus (+). Although he gave no history of TB, to exclude the possibility of intestinal TB, both Quantiferon TB gold test and Mantoux test were also done and found negative. At this point celiac disease was considered and anti-TTG Ab titer was found-52.3 u/mL (cut off value 50 u/mL). Colonoscopy did not reveal any abnormality. Biopsy taken by upper GI endoscopy, showed total villous atrophy with increased number of goblet cells in the lining mucosa and dense infiltration of lymphoid cells in the lamina propria, forming follicles in some areas. The lymphoid cells have infiltrated the mucosa in some areas. Thus the endoscopic findings were suggestive of celiac disease.

Discussion

There are several different approaches for diagnosing celiac disease, but usually all of them require presence of serologic evidence e.g. anti TTG antibody, positive biopsy findings and change in clinical features and serology with exposure to or withdrawal of gluten.[3] Leffler et.al. has proposed a similar approach, in their "Proposed Modified Gluten Challenge Algorithm" where positivity of baseline serology indicates probable celiac disease, and then subsequent positive duodenal biopsy confirms the diagnosis.[4] But, according to proposition of Leffler et.al., negative baseline serology does not exclude the diagnosis of celiac disease, but rather it demands HLA DQ2/DQ8 testing, and if the patient has susceptible HLA DQ2/DQ8 pattern, a gluten challenge test is done with three gram of gluten per day for 2 to 6 weeks, which may bring out characteristic serological changes and histological changes in biopsy.[4] Multiple biopsies should be obtained from the duodenal bulb and the second and third portion of the duodenum.[5,6] There are differences of opinions as for to the number of biopsies to make a certain diagnosis of celiac disease but, 4 to 6 biopsies are recommended by some.[7,8] Pais et.al. have suggested that only 2 biopsy specimens will lead to a confirmed diagnosis of CD in 90%, and a suspected diagnosis in all, but for 100% confidence in diagnosis of CD, 4

duodenal biopsy specimens should be taken.[9] Here it can be noted that antibodies have a great role in diagnosis of celiac disease. IgA tissue transglutaminase antibodies has a sensitivity of 90 to 98 percent and a specificity of 95 to 97 percent.[10] Nonetheless, a positive antibody test suggests that a person might have celiac disease, but to confirm, a biopsy will be needed.[11] Peripheral neuropathies maybe found in up to 50 percent of patients with celiac disease and may precede its diagnosis. Ludvigsson et.al. reported in a large population-based study from Sweden that included 14,000 celiac patients and 70,000 controls, that celiac disease was associated with an increased risk of polyneuropathy (hazard ratio 3.4), but not with other neurologic outcomes.[12] They also recommended that individuals with polyneuropathy should routinely undergo screening for CD.[12] In our case, the patient presented with recurrent attack of quadriparesis. Instances of such manifestation of celiac disease is truly rare, although have been reported in recently published medical literature a very few times.[13–15] Celiac disease is a rare cause of neuropathy that most commonly presents with symmetric distal sensory disturbances.[15,16] Neurologic manifestations, such as ataxia and peripheral neuropathy, are increasingly recognized to be the presenting features of CD.[17] Hadjivassiliou et.al. described 9 patients with neuromuscular disorders with CD, 3 patients had sensorimotor axonal peripheral neuropathy, 1 had axonal motor peripheral neuropathy, 1 had probable inclusion body myositis and axonal motor peripheral neuropathy, 1 had polymyositis and sensorimotor peripheral neuropathy, 1 had mononeuropathy multiplex, 1 had neuromyotonia, and 1 had polyneuropathy.[18] Hernandez and Green reported neurologic and psychiatric disorders associated with celiac disease include cerebral ataxia, peripheral neuropathy, epilepsy, dementia, and depression.[19] As for the mechanism of neurological manifestations of CD, current data point to molecular mimicry and intermolecular help as two possible mechanisms.[17] Rigamonti et.al. described two patients in whom celiac disease was diagnosed after the onset of peripheral neuropathy characterized by predominant motor nerve involvement in whom, after a gluten-free diet, significant improvement of symptoms and neurophysiological findings occurred.[15] In our case, changes in serum electrolytes may reflect the reason why our patient suffered from quadriparesis due to hypokalemic periodic paralysis. The only known instance of such phenomenon was found in India.[14]

Conclusion

Although neurological symptoms have been associated with celiac disease, motor weakness is very uncommon. We have proposed that in our case electrolyte imbalance have caused or contributed in causing this motor weakness.

Conflict of interest: None

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