

Myositis in a 22-year-old girl suffering from Celiac Disease: A Case report

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ABSTRACT:

Celiac disease is a chronic intestinal disorder with many extra manifestations including bone disease, muscular disorders, endocrine disorder, and neurological deficits. We report a 22 years old girl who presented with diarrhea and symmetrical lower limb muscular weakness. Patient had short stature as well as refractory iron deficiency anemia. Her reports showed a positive anti transglutaminase IgA, and duodenal mucosal villous atrophy thereby establishing the diagnosis of celiac disease. Moreover, her muscle enzymes were also raised along with abnormal electromyography contributing to diagnosis of myositis secondary to celiac disease. Coexistence of both disorders in a patient provided evidence to the fact that both share an autoimmune etiology.

Keywords: Celiac disease, Myositis, Short stature.

INTRODUCTION:

Celiac disease is a systemic inflammatory disorder having an autoimmune basis, triggered in genetically susceptible individuals with the ingestion of gluten¹. It is an underdiagnosed condition in adults presenting with intestinal as well as multiple extra intestinal symptoms including bone disease, muscular disorders, endocrine disorders, and neurological deficits². Various autoimmune disorders are also associated with celiac disease such as insulin dependent diabetes, dermatitis herpetiformis, autoimmune thyroiditis, IgA nephropathy, primary biliary cirrhosis, autoimmune hepatitis, and sclerosing cholangitis³⁻⁵.

Myositis is the inflammatory disorder affecting the muscles of the body especially the skeletal muscles and the muscles of gastrointestinal tract⁶. It can be caused by an injury, infection or an autoimmune disease. Inflammatory myopathies are characterized by progressive symmetrical muscle weakness, elevated serum muscle enzyme levels, abnormal electromyography and inflammatory infiltrates on muscle biopsy⁷. We report a case of 22 years old female who developed inflammatory myopathy secondary to celiac disease.

CASE REPORT:

A 22 years old female was admitted with acute lower limb paralysis. It was preceded by diarrhea for 6 days and vomiting for 2 days. Treatment was done for acute gastroenteritis by a local practitioner with no recovery. Later the patient developed weakness of both her lower limbs. On examination patient had pallor and short stature. She was fully conscious and well oriented in time, place, and person. Her sensory system was intact whereas her motor system examination revealed normal power in both upper limbs (5/5) and decreased power in both lower limbs (4/5). Initially patient was suspected for Guillain-Barré syndrome (GBS) on the basis of symmetrical muscle involvement. Her work up showed raised muscle enzymes (CPK: 25630 u/L) but nerve conduction studies revealed no demyelinating changes thereby ruling out GBS. Her muscle biopsy showed inflammatory infiltrates pointing towards myositis. In order to confirm it electromyography was done which showed fibrillatory waves thereby confirming our diagnosis.

Patient was also investigated for celiac disease. Her anti transglutaminase IgG level was 160 U/ml (cut off 10 U/ml). Endoscopy showed scalloped duodenal folds and

grooves along with short villi. Duodenal biopsy revealed villous atrophy, crypt elongation, increased intraepithelial lymphocytes, and the patient was diagnosed as having celiac disease type 3b. Anti-ganglioside antibody could not be done for financial constraints.

Short stature of patient can be explained by the malnutrition secondary to celiac disease as thyroid profile of the patient was within normal range (TSH: 2.0 uIU/mL). Patient was having concomitant iron deficiency anemia despite having received blood transfusion (Hb 4.4 g/dl, MCV 72.5 fl, MCH 16.1 pg, MCHC 22.2 g/dl).

Patient was started on low dose prednisone as initial therapy which resulted in recovery. She was also started on gluten free diet due to coexisting celiac disease. Hematinic were given to improve pallor and short stature. Follow up was advised after 3 months.

DISCUSSION:

Celiac disease is a chronic inflammatory disease that is characterized by mucosal villous atrophy. The most common presentation of celiac disease is diarrhea with bulky fatty stools, fatigue and weight loss. However, it can also manifest with other clinical presentations involving multiple organs that makes its diagnosis difficult. Its prevalence is estimated to be 1:300 in the general population⁶. It is an autoimmune disorder, and has association with multiple autoimmune disorders.

Our study is unique in the sense that it describes a case of celiac disease which presented with symmetrical lower limb weakness establishing the diagnosis of polymyositis that is very rare in adult patients with celiac disease. The occurrence of juvenile dermatomyositis has been studied in patients with celiac disease but only a few cases have been studied in adults⁸⁻¹⁰. In a study by I. Marie, patient was diagnosed with dermatomyositis initially and later on as diagnosed with celiac disease⁶. In our case signs of malnutrition were already evident in the form of anemia and short stature. Initially the disease was latent in the patient for many years but her presentation in the hospital led to her diagnosis on the basis of her endoscopy findings revealing duodenal folds and grooves, and positive anti-transglutaminase IgG levels. Patient was diagnosed to have myositis due to raised muscle enzyme levels, electromyography findings and muscle biopsy.

Both celiac disease and myositis have an autoimmune basis. The association between both disorders can be explained on the basis of presence of HLA genes. Unfortunately, in our setting the test for presence of HLA genes could not be performed due to financial constraints and non-availability of test in the hospital. Relation between both disorders can also be explained by the fact that gluten free diet resulted in remission of

symptoms of both diseases. After taking gluten free diet power of lower limbs was also restored to normal.

CONCLUSION:

Our study describes a unique scenario of myositis in a suspected patient of celiac disease, which is overall rare among celiac patients. Thus, A complete workup should be done including muscle enzyme levels, electromyography, and muscle biopsy to establish the diagnosis. Furthermore, other neurological causes should be excluded by nerve conduction studies. This would lead to the early diagnosis, and definitive management of the patients reducing their morbidity. However further studies should be conducted to check the frequency of myositis in celiac disease patients.

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ZA, BA and MZ have given substantial contributions to the conception or the design of the manuscript, MSK and AK to acquisition, analysis and interpretation of the data. All authors have participated to drafting the manuscript, TH revised it critically. All authors read and approved the final version of the manuscript.

REFERENCES:

1. Alessandra Pacitto, A. Paglino, Lorenza Di Genova, A. Leonardi, et al; Celiac disease presenting with peripheral neuropathy in children; Int J environ res public health 2017; 14(7): 785.<https://doi.org/10.3390/ijerph14070785>
2. Vijay gupta, A. kohli; Celiac disease associated with recurrent Guillain barre-syndrome; Indian pediatrics 2010; 47: 797-798.
<https://doi.org/10.1007/s13312-010-0105-3>
3. Maki M, Collin P; Coeliac disease; Lancet 1997; 349: 1755-9.[https://doi.org/10.1016/s0140-6736\(96\)70237-4](https://doi.org/10.1016/s0140-6736(96)70237-4)

4. Schuppan D; Current concepts of celiac disease pathogenesis; *Gastroenterology* 2000; 119: 234-42.
<https://doi.org/10.1053/gast.2000.8521>
5. Trier JS; Celiac sprue; *N Engl J Med* 1991; 325:1709-
19.
<https://doi.org/10.1056/nejm199112123252406>
6. Marie I, Lecomte F, Hachulla E, et al; An uncommon association: Celiac disease and dermatomyositis in adults: Clinical and experimental rheumatology 2001; 19: 201-203.
<https://europepmc.org/article/med/11326486>
7. Albert SC, Francesc C, Ines DT, et al; Celiac disease and antibodies associated with celiac disease in patients with inflammatory myopathy; *Muscle and nerve* 2007; 35 (1); 49-54.
<https://doi.org/10.1002/mus.20652>
8. Buderus S, Wagner N, Lentze MJ: Concurrence of celiac disease and juvenile dermatomyositis; Result of a specific immunogenetic susceptibility; *J Pediatr gastroenterol Nutr* 1997; 25: 101-3.
<https://doi.org/10.1097/00005176-199707000-00018>
9. Evron E, Abarbanel JM, Branski D, Sthoeger ZM; Polymyositis, arthritis, and proteinuria in a patient with adult celiac disease; *J Rheumatol* 1996; 23: 782-3.
<https://pubmed.ncbi.nlm.nih.gov/8730148/>
10. Falcini F, Porfirio B, Lionetti P; Juvenile dermatomyositis and celiac disease; *J Rheumatol* 1999; 26: 1419-20.
<https://pubmed.ncbi.nlm.nih.gov/10381073/>