

Celiac Disease Presenting as Cardiomyopathy - A Rare Extra Intestinal Manifestation

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Received May 03, 2020; Revised June 07, 2020; Accepted June 25, 2020

Abstract Cardiac manifestations of celiac disease has been poorly described in literature though some studies have emphasized on correlation of ischemic heart disease, atrial fibrillation and dilated cardiomyopathy with celiac disease. We describe a patient with celiac disease associated cardiomyopathy whose cardiac function improved substantially after treatment with a gluten-free diet. A young lady of 35 years age was admitted with complaints of chronic diarrhea, vomiting, significant weight loss, dizziness and chronic iron deficiency anemia since last 3 months. Physical examination revealed tachycardia, tachypnoea and hypotension requiring multiple high dose inotropic support. She had pallor, skin changes and pedal edema suggestive of malabsorption. Investigations revealed iron deficiency anemia, high bilirubin and transaminases, prothrombin time, low albumin and elevated tissue transglutaminase antibody. Echocardiography showed global hypokinesia with a left ventricular ejection fraction of 20%. Diagnosis of celiac disease made on clinic features, serology and biopsy finding of subtotal villous atrophy, crypt hyperplasia, and increased intraepithelial lymphocytes. She was kept on gluten free diet and on intravenous steroids for acute celiac crises and celiac related cardiomyopathy as patient had severe diarrhea, hypocalcaemia, weight loss, hypoproteinemia and hypotension. Patient improved within 48 hours of steroid with discontinuation of inotropes. She was discharged with gluten free diet and follow up after 6 months showed near normalization of all biochemical abnormalities and ejection fraction of 55%. Cardiomyopathy associated with celiac disease and celiac crisis is a serious and potentially lethal condition. However, with early diagnosis and treatment with a gluten free diet, and steroids cardiomyopathy in patients with celiac disease may be completely reversible.

Keywords: *celiac disease, dilated cardiomyopathy, celiac crisis, malabsorption, gluten-free diet*

Cite This Article: Shravan Bohra, and Apurva Shah, "Celiac Disease Presenting as Cardiomyopathy - A Rare Extra Intestinal Manifestation." *International Journal of Celiac Disease*, vol. 8, no. 2 (2020): 55-56. doi: 10.12691/ijcd-8-2-4.

1. Introduction

Celiac disease (CD) is a chronic small intestinal immune-mediated enteropathy precipitated by exposure to dietary gluten in genetically predisposed individuals. [1] CD is now considered a relatively common disease affecting about 0.6-1% of the world's population. [2] Its frequency in India seems to be higher in the Northern part of the country, a finding that is at least partially explained by the wheat-rice shift from the North to the South. [3] CD characterized by clinically evident gastrointestinal and/or extra intestinal symptoms attributable to gluten intake. The diagnosis of celiac disease is made on the basis of the modified European Society of Pediatric Gastroenterology, Hepatology and Nutrition (ESPGHAN) criteria, including clinical manifestations, serology, histological features suggestive of celiac disease and an unequivocal response to gluten-free diet (GFD). The main treatment for CD is a gluten-free diet (GFD), which typically results in the restoration of the small intestinal villi and restoration of other affected organ systems, to their normal functioning.

Extra intestinal manifestations of celiac disease are well described and often prove more distressing to the patient than do the gastrointestinal symptoms. Cardiomyopathy is the most frequently documented cardiovascular condition observed in conjunction with CD, and seems to mostly or completely resolve with appropriate treatment, including a GFD. Celiac crisis, a life threatening syndrome, was defined as acute onset or rapid progression of gastrointestinal symptoms that could be attributed to celiac disease and required hospitalization and/or parenteral nutrition, along with signs or symptoms of dehydration or malnutrition. [4] Systemic steroids with nutritional support for short term and then gluten free diet is cornerstone for treatment of celiac crisis. [4] We describe a patient with celiac disease associated cardiomyopathy and celiac crisis whose cardiac function improved substantially after treatment with steroids and a gluten-free diet.

2. Case Description

A 35 years lady admitted with complaints of chronic diarrhea, vomiting, significant weight loss since 3 months with history of full term normal delivery 2 months ago.

History of severe iron deficiency anemia (IDA) for which she required blood transfusions and iron therapy since last 3 months. On physical examination at admission she was alert had pallor, tachycardia, tachypnoea, dry scaly skin and bilateral pedal edema suggestive of malabsorption. She had hypotension refractory to fluid resuscitation - admitted in intensive care unit and required continuous multiple inotropic supports for days.

Investigations revealed iron deficiency anemia with hemoglobin 7.7 gm/dl, MCV-72 fl, deranged liver function tests in form of high total bilirubin-5.6 mg/dl, direct bilirubin- 3.2 mg/dl, transaminitis (2 times upper limit of normal), high prothrombin time- international normalized ratio-2.07, low albumin (2.4g/dl) and elevated tissue trasglutaminase and positive anti endomysial antibody. Her serum TSH, serum cortical levels were within normal limits. Ultrasound abdomen revealed mild hepatomegaly, mild ascites and proximal small bowel edema. Electrocardiogram showed sinus tachycardia. 2D Echocardiography showed global hypokinesia with LV ejection fraction of 20% and features of dilated cardiomyopathy. Coronary angiography revealed normal coronary arteries. Gastroscopy done for evaluation of IDA revealed scalloping of duodenal folds. Diagnosis of celiac disease was made on clinical features, serology and duodenal biopsy finding of subtotal villous atrophy, increased intraepithelial lymphocytes.

Other causes of dilated cardiomyopathy were ruled out; there was no history of any cardiac symptoms during previous pregnancy. She was monitored in the ICU for severe diarrhea, weight loss, hypocalcaemia, hypoproteinemia and persistent hypotension requiring multiple inotropic supports. She was started on gluten free diet (GFD) and on intravenous steroids for acute celiac crises and suspected celiac disease related cardiomyopathy. Patient improved within 48 hours of steroid treatment with discontinuation of inotropes over next 48 hours.

She was discharged on gluten free diet and at six months follow up she was asymptomatic, regained weight with near normalization of all biochemical abnormalities and cardiac ejection fraction of 55%.

3. Discussion

Extra intestinal manifestations of celiac disease like anemia, osteopenia, neurologic symptoms, cutaneous and menstrual abnormalities are well described in literature, but no significant literature on cardiac manifestations of celiac disease. It was determined that there has been a sharp increase in the number of CD - cardiovascular studies since 2000. Most of the publications are either of the type "article" or "case study". The largest number of documents published concerned CD in conjunction with cardiomyopathy (33 studies), and there have also been substantial numbers of studies published on CD and thrombosis (27), cardiovascular risk (17), atherosclerosis

(13), stroke (12), arterial function (11), and ischemic heart disease (11). [5] Though some studies have emphasized on correlation of ischemic heart disease, atrial fibrillation and dilated cardiomyopathy, only few case reported with cardiomyopathy related to celiac disease. Significant rapid improvement in cardiac status after initiation of steroid and gluten free diet points to cardiomyopathy associated with celiac disease in our patient.

Several mechanisms have been proposed for the development of cardiomyopathy in celiac disease. Firstly, malabsorption occurring in celiac disease may lead to cardiomyopathy caused by severe nutritional deficiencies. [6] Autoimmune response directed against antigens present in both the myocardium and small bowel may be responsible for myocardial injury. [6,7]

The mainstays of treatment of celiac crisis are initiation of a gluten free diet, parenteral fluid replacement and nutritional support, and in most cases corticosteroids as given in our case. [4] Patient came out of celiac crisis and steroids were stopped after short term administration.

In conclusion, dilated cardiomyopathy associated with celiac disease is a serious condition which requires multidisciplinary approach involving gastroenterologist and cardiologist. Early recognition of celiac crisis is important as it is associated with significant morbidity. Compliance with the GFD is mandatory if patients are to avoid progression of cardiomyopathy. Based on the published research, it can be concluded that many types of cardiovascular issues can occur in untreated CD patients, but that most tend to resolve on a GFD, often in conjunction with the healing of small intestinal villous atrophy. Screening of patients with idiopathic dilated cardiomyopathy for celiac disease should be considered and celiac crisis should be thought of if there is a rapid deterioration in cardiac function.

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