Case Report

Association of Celiac Disease with Cardiomyopathy and Pulmonary Hemosiderosis

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Summary

Celiac disease is predominantly a disease of the small intestine characterized by chronic malabsorption in genetically susceptible individuals who ingest grains containing gluten, such as wheat, barley and rye. Although previously believed to be uncommon, celiac disease may be present in up to 1% of the general population. Celiac disease is frequently associated with many extraintestinal disorders, but rarely with cardiomyopathy. We describe a patient with celiac disease associated with cardiomyopathy and pulmonary hemosiderosis.

Introduction

Celiac disease is characterized by chronic malabsorption in susceptible individuals who ingest grains containing gluten, such as wheat, barley and rye. Until recently, celiac disease was believed to be relatively uncommon. However, a recent report from Finland estimates its prevalence at 1% in the general population [1]. Celiac disease is associated frequently with iron deficiency anemia, dermatitis herpetiformis, selective IgA deficiency [2], thyroid disorders [3], diabetes mellitus [4, 5] and various connective tissue disorders [6–8]. Cardiomyopathy associated with celiac disease is reported infrequently. We describe a case of cardiomyopathy in a patient with celiac disease who had neither gastrointestinal symptoms nor manifestations of malabsorption.

Case

A 13-year-old boy presented to us with the complaints of recurrent hemoptysis for 3 years. Blood amount was about 50–100 ml each time. There was no history of prolonged cough, breathing difficulty, weight loss or bleeding from any other site. There was New York Heart Association (NYHA) class I dyspnea on exertion, from past 6 months. In the past, he was evaluated elsewhere for hemoptysis, but cause could not be determined. He has also received two blood transfusions for increasing pallor. On examination, he was undernourished, underweight and had short stature. He had severe pallor, no cyanosis, clubbing or lymphadenopathy. Respiratory system and cardiovascular system examinations were normal. There was no hepatosplenomegaly on abdominal examination. Ear, nose and throat examination was also normal. Laboratory investigations revealed that hemoglobin was 8.4 g/dl, along with normal leukocytes, differential leukocytes and platelet counts. Peripheral blood picture showed microcytic and hypochromic anemia. Liver function tests, prothrombin time and activated partial thromboplastin time were within normal range. HIV serology and cANCA were negative. Three sputum examinations were negative for acid fast bacilli. Chest X-ray revealed cardiomegaly with cardiothoracic ratio of 60%. Ultrasound abdomen was noncontributory. Bronchoscopy was suggestive of erythema at entry point in bronchus and broncho alveolar lavage was done which revealed hemosiderin laden macrophages suggestive of pulmonary hemosiderosis. Patient underwent echocardiography with the background of dyspnea and cardiomegaly. Echocardiography showed dilated left ventricle and global hypokinesia, with left ventricular dimensions of 54 mm and 62 mm in end systole and end diastole, respectively. Ejection fraction was 26% and fractional shortening was 12%, suggestive of severe left ventricle systolic dysfunction. He was diagnosed with dilated cardiomyopathy.
In view of malnutrition, short stature and anemia, tissue transglutaminase antibody (tTG IgA) levels were done. Tissue transglutaminase antibody levels were 67 U/ml (reference range of <20 U/ml). Upper gastrointestinal endoscopy revealed scoloped duodenal folds, and results of a histopathologic examination revealed partial villous atrophy, crypt hyperplasia and increased intraepithelial lymphocytes. Celiac disease with cardiomyopathy along with pulmonary hemosiderosis was diagnosed, and a strict gluten-free diet was initiated. Patient was also started on diuretics, digoxin and enalapril. He responded to the treatment with improvement in clinical features and gain in height and weight on follow-up.

**Discussion**

Celiac disease is associated frequently with iron deficiency anemia, dermatitis herpetiformis, selective IgA deficiency [2], thyroid disorders [3], diabetes mellitus [4, 5] and various connective tissue disorders [6–8]. Association of celiac disease with pulmonary hemosiderosis is known. Cardiomyopathy associated with celiac disease is reported infrequently. An association between celiac disease and cardiomyopathy has been reported recently. Curione et al. [9] studied 52 patients and reported an increased prevalence of celiac disease (5.8%) in patients with dilated cardiomyopathy. The prevalence of celiac disease in patients with dilated cardiomyopathy was 2.2% compared with 0.35% in controls in the study by Prati et al. [10]. Polat et al. [11] also detected subclinical systolic dysfunction of the left ventricle in children of celiac disease. No such case has been reported from India as of now.

Several mechanisms have been proposed for the development of cardiomyopathy in celiac sprue. Chronic malabsorption, common in celiac disease, may lead to cardiomyopathy secondary to nutritional deficiencies [12]. Another proposed mechanism in patients with celiac disease is abnormalities of intestinal permeability may lead to increased systemic absorption of various luminal antigens or infectious agents that may cause myocardial damage through immune-mediated mechanisms [13–15]. Finally, myocardial injury may be secondary to an immune response directed against an antigen present in both the myocardium and the small intestine [12, 16–18]. A recent study by Not [19] suggested that celiac disease is associated with cardiomyopathy rather than being a cause of cardiomyopathy because celiac disease did not co-segregate with cardiomyopathy on human leukocyte antigen (HLA) testing [19].

Our case highlights several important points about celiac disease and associated conditions. Celiac disease is often asymptomatic or presents with extremely few symptoms. A high degree of clinical suspicion is required to make a prompt and correct diagnosis. Cardiomyopathy associated with celiac disease is a serious and potentially lethal condition. A careful medical history should be obtained to elicit gastrointestinal symptoms, and the presence of iron deficiency anemia should be investigated in patients who present with dilated cardiomyopathy in the absence of known etiologies. Such patients also should be screened for celiac disease with serologic tests such as tissue transglutaminase antibody.

**References**


