Case Report

Coeliac Disease: Unusual Cause of Iron Deficiency Anaemia in Adults

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Introduction

Coeliac disease, long considered to be a disease of post weaning period, is increasingly being diagnosed in adults with atypical presentations. We report a case of adult coeliac disease with one such atypical presentation.

Case History

NA, 42 years old, male was seen with complaints of loose bowel movements and weakness for two years. Stools were large volume without mucus or blood with a frequency of 4 - 5 per 24 hours. He gave history of significant weight loss. Past medical and surgical history was unremarkable. He had received various medications without any significant improvement.

Physical examination revealed pallor with thin built. There were no other significant physical findings.

Initial laboratory investigations showed haemoglobin of 8.6 G/dl, MCV 63, MCHC 23. Stool for occult blood was negative on two occasions. Screening test for Hepatitis B & C were negative. Upper gastrointestinal endoscopy was normal except for granular appearance of descending duodenum and biopsies were taken. Histopathological examination showed flattening of surface due to loss of villi, elongation of crypts and marked infiltration of lamina propria with lymphocytes and plasma cells. Giardia, PAS positive macrophages or granuloma were not seen. Impression was of total villous atrophy consistent with diagnosis of coeliac disease (Fig. 1).

Later, result of antigliadin and antiendomysium antibodies were available and turned out positive.

He was advised gluten free diet and given iron and folic acid supplements. On follow up visit 6 months later, he showed clinical improvement. Diarrhoea had settled and he had gained weight. Haemoglobin was now 11.6 G/dl.

Repeat endoscopy and biopsy from the distal duodenum showed that villi had not appeared, cellular infiltrate had decreased and crypts appeared normal (Fig. 2).

Discussion

Coeliac disease commonly manifests between 6-24 months after introduction of weaning food. Up to half of the cases present in childhood or adults. The disease was considered to be rare. However, serologic screening shows world-wide prevalence of 1:266.

Coeliac disease is a disease of proximal small intestine that can involve the entire small intestine in some individuals. Proximal segment involvement results in malabsorption of iron, folic acid, calcium and fat soluble vitamins. Diarrhoea and weight loss, the cardinal symptoms are mostly due to progression of disease to distal small bowel. Our patient had features of proximal as well as distal segment involvement. He gave history of diarrhea, weight loss and was found to have iron deficiency anaemia. No obvious source of gastrointestinal bleeding could be identified. It is reported that 10 % of adults with iron deficiency anaemia not due to gastrointestinal bleeding have undiagnosed coeliac disease.

An accurate diagnosis of coeliac disease is mandatory as it requires life long dietary restrictions,
special attention to complications and associated conditions.

Fig. 1: Before Treatment: Duodenal Biopsy from 2nd Part showing Complete Villous Atrophy, Mono Nuclear Inflammatory Cells Infiltrate in the Lamina Propria. (H & E X 100)
Diagnosis of coeliac disease was based on strict criteria first proposed at ESPGAN meeting in 1969. These include the finding on endoscopic small intestinal biopsy sample of a structurally abnormal jejunal mucosa when the patient is taking gluten containing diet, a clear improvement of villous structure on a gluten free diet and finally the deterioration of mucosa during gluten challenge. Utility of gluten challenge has been questioned and the criteria was revised in 1990 by a working group of ESPGAN.

The disease may be suspected if endoscopic signs of villous atrophy are present. These include reduction in the number of circular folds, mucosal fissure and nodular appearances. Our patient had granular appearances in the 2nd part of duodenum. These endoscopic appearances are not specific to coeliac disease and may occur in several other disorders. Adequate number of biopsies should be taken because the disease may be patchy.

Major histopathological features for diagnosis are villous atrophy, crypt hyperplasia and intra-epithelial lymphocytosis. In adults histological abnormalities may improve with gluten free diet but they often do not return to normal. Our patient illustrates this observation. His histopathological appearances improved, there was decrease in cellular infiltrate but villi did not reappear.

Serological tests have been employed for screening patients for endoscopic biopsy. Antigliadin antibodies have been assessed in several studies resulting in heterogenous expressions of sensitivity and specificity. Presence of antibodies against endomysium by indirect immunofluorescence is almost 100% specific in the diagnosis of coeliac disease. Anti tissue transglutaminase antibodies uses the same antigen as for endomysial antibody and has the same significance. Positive serological tests lend support to diagnosis but are not essential. They may be absent in patients with IgA deficiency, diabetes mellitus and chronic liver disease.

Cellobiose mannitol test has also been extensively used in the screening for coeliac disease and has shown good sensitivity but poor specificity.

In equivocal biopsy or negative serological test, HLA typing for histocompatibility complex II class HLA DQ-2 and HLA DQ-8 is helpful as it is present in 98% of patients with coeliac disease. Strict gluten free diet remains the cornerstone in the treatment of coeliac disease. Trial of gluten restriction without firm diagnosis should be avoided. Most patient show good response. Iron and folic acid supplement can be used if deficiency is documented. Patients with poor clinical response need re-evaluation of diagnosis after exclusion of unintentional gluten ingestion. Patients with refractory disease can be given steroids, immunosuppressive agents or infliximab after exclusion of malignant transformation into T-cell lymphoma.
References


