Celiac Disease

What's On Your Plate?

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Penny's case

Penny, 69, has a long history of mild aortic stenosis. She presents with progressive shortness of breath and increasing fatigue. She is currently taking digitalis and warfarin for atrial fibrillation, and bisphosphonates and calcium for osteoporosis. Recent cardiac investigations revealed a stable cardiac state.

On admission, Penny's symptoms and lab test results are all compatible with iron deficiency anemia (Table 1).

Recent air contrast barium enema, which was performed for abdominal pain, revealed left-sided diverticulosis. An upper gastrointestinal series revealed a large hiatus hernia with free reflux.

Penny is transfused with two units of red blood cells and undergoes colonoscopic exam. It confirms the presence of diverticular disease, but also reveals bleeding angiodysplasia in the cecum (Figure 1).

She undergoes successful coagulation of the lesion.

For more on Penny, go to page 70.

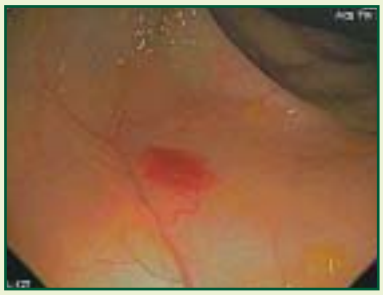

Figure 1. Endoscopic picture of cecal angiodysplasia.

Table 1

Penny's presentation

- Pale and tachypneic
- Chest: Clear to auscultation
- Hemoglobin: 68 g/L
- MCV: 72 fl
- Ferritin: 3 µg/L
- Stool: Occult blood negative

MCV: Mean cell volume

Celiac disease is a common, T-cell-mediated, autoimmune, intestinal disease. Its prevalence among North American Caucasians is one in 250.[1]

How does celiac disease present?

Celiac disease is characterized by the destruction of the small intestinal mucosa upon exposure to gluten in a genetically predisposed individual. The age of clinical onset varies, with 20% of cases occurring in patients older than 60.

Iron deficiency anemia appears to be the most common presentation in adults; however, patients may also present with:

- fatigue,
- abdominal bloating with diarrhea,
- inability to gain weight, or
- back pain secondary to osteoporosis.

The classic picture of weight loss, diarrhea, and steatorrhea may not be present.

Celiac disease appears to be associated with other autoimmune diseases, such as Type 1 diabetes and autoimmune thyroiditis. The coexistence of

A followup on Penny

Over the next six months, Penny does relatively well; however, despite therapeutic-dose iron supplementation, her hemoglobin does not go over 100 g/L. An antiendomysial antibody is positive, compatible with celiac disease.

A biopsy from the second part of the duodenum obtained at endoscopy confirms the diagnosis (Figure 2a) compared to a normal biopsy (Figure 2b).

Figure 2a. Subtotal villous atrophy with flattening of the villi, elongation of the crypts, and increased plasma cells.

Figure 2b. Normal duodenal biopsy.

The prevalence of celiac disease among North American Caucasians is one in 250.

Type 1 diabetes and celiac disease is reported to be between 6% and 8%.[2]

How is it investigated?

While small bowel biopsy used to be the main test used to diagnose celiac disease, serologic markers with high sensitivity and specificity have become available in the past 10 years. For a while, the antiendomysial antibody was the gold standard serologic marker, with a sensitivity of 85% to 98% and a specificity of 97% to 100%, but this test was cumbersome to perform and operator-dependent.

More recently, tissue transglutaminase was identified as the autoantigen recognized by the antiendomysial antibody. An immunoglobulin (Ig) A enzyme-linked immunosorbent assay (ELISA) that uses human tissue glutaminase has been found to be as effective as the antiendomysial antibody assay. Furthermore, it is easier to perform, cheaper, and can be used for population screening.

Patients who are suspected to have celiac disease, but whose tests are negative, should have their immunoglobulin electropheresis assessed, as an IgA antibody is used for the assay.

What is the treatment?

Treatment for this lifelong disease is a strict, gluten-free diet; all patients should be referred to a dietitian for ongoing counselling. Patients should avoid any food containing wheat, rye, and barley. Although oat used to be considered toxic, recent evidence has shown otherwise.

Certain drugs and foods may contain gluten as an additive, so all food labels should be monitored closely.

Frequently Asked Questions

1. Who should be screened for celiac disease?

First-degree relatives of patients with celiac disease, patients with Type I diabetes and anemia, and patients with unexplained osteoporosis.

2. What is the best test to screen for celiac disease?

Tissue transglutaminase (tTG) is the best test to perform in patients suspected of having celiac disease and those who are not deficient in IgA.

3. Should patients who are antibody-positive to tTG antibody and asymptomatic adhere to a strict diet?

Yes. Although they may be asymptomatic, these patients are still at risk of developing complications, including osteoporosis and malignancies.

4. How can we be sure patients are adhering to their diets?

In patients who are adherent to diet, anti-tissue transglutaminase levels disappear.

5. Is biopsy still needed to diagnose celiac disease?

Although serology is sensitive and specific, most gastroenterologists still recommend biopsy. This recommendation may change in the near future.

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Lactase deficiency is common secondary to villous atrophy; lactose should be avoided for the first six months after celiac is diagnosed and gradually re-introduced, providing the patient does not have an inherent lactose intolerance.

Multivitamins, including folate and iron, should be administered early, as should calcium and vitamin D.

Response to therapy usually occurs within the first two weeks, although the histologic response may take months.

The most common cause for poor response is lack of adherence to diet. In patients who adhere to a strict, gluten-free diet, serum levels of anti-tissue transglutaminase disappear.

Unresponsive patients may require immunosuppressive therapy.

What complications can arise?

Osteoporosis and osteopenia are the most common nonmalignant complications in celiac disease. Unless a bone densitometry is performed, these diagnoses can be easily missed until a fracture occurs.

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Take-home message

What is celiac disease?

- Celiac disease is a common, autoimmune, intestinal disease. In patients who have Type I diabetes and anemia, the possibility of celiac disease is increased. Similarly, patients with unexplained anemia, osteoporosis, or unexplained neurologic abnormalities should be screened for celiac disease.

How is celiac disease treated?

- Lifelong, strict treatment with a gluten-free diet is the main therapy and should be adhered to in order to prevent complications, such as osteoporosis and malignancies.

Neurologic complications have been recorded in celiac patients, including:

- ataxia,
- peripheral neuropathy, and
- other neurologic diseases.

Patients with celiac disease are at an increased risk of developing non-Hodgkin's lymphoma. Other malignancies that may be increased in these patients include small intestinal carcinoma, esophageal cancer, and melanoma.Patients who continue to have diarrhea, yet are antibody-negative, may be suffering from microscopic colitis or may have sufferd complications, such as lymphoma or microulcerative jejunoileitis. Dx

References

1. Fasano A, Berti I,Gerarduzzi T, et al: Prevalence of celiac disease in at-risk and not-at-risk groups in the United States: A large multicenter study. Arch Intern Med 2003; 163(3):286-92.
2. Cronin CC,Feighery A, Ferris JB, et al: High prevalence of celiac disease among patients with insulin-dependent (type I) diabetes mellitus. Am J Gastroenterol 1997; 92(12):2210-2.