

# Macroamylasemia as the First Manifestation of Celiac Disease

Roman Depsames Zvi Fireman Eva Niv Yael Kopelman

Department of Gastroenterology, Hillel-Yaffe Medical Center, Hadera, Israel,  
affiliated to the Bruce Rappaport Faculty of Medicine, The Technion, Haifa, Israel

## Key Words

Macroamylasemia · Celiac disease · Gluten-free diet · Maculopapular rash · Anemia

## Abstract

Macroamylasemia is a biochemical disorder characterized by an elevated serum amylase activity resulting from the circulation of a macromolecular complex of amylase with a serum component, often an immunoglobulin. The increased molecular weight of this complex prevents the normal renal excretion of the enzyme. A few cases of celiac patients with macroamylasemia have been published in whom the biochemical disorder disappeared after treatment with a gluten-free diet.

## Background

Macroamylasemia occurs in approximately 0.4% of the general population, and about 2.5–5.9% of these patients have hyperamylasemia. Hyperamylasemia may be either an isolated, benign laboratory finding without pathological significance or it may be associated with underlying diseases, such as lymphoma, AIDS, carcinoma, liver disease and autoimmune disorders. The circulating macroamylase complexes of pancreatic or salivary amylase are bound to plasma proteins which cannot be cleared by the renal glomeruli. In most cases, the macromolecular amylase represents a complex of normal amylase and either immunoglobulin A or G and may be a specific antigen-antibody complex [1, 2]. Celiac disease is a permanent intolerance to ingested gluten that results in immunologically mediated inflammatory damage of the small intestinal mucosa. Several recent population-based serologic surveys have shown celiac disease to be a common disorder, possibly affecting 1 in 200–250 individuals in most countries studied. Only few cases of celiac disease associated with macroamylasemia have been described, and the macroamylasemia disappeared once the patients followed a gluten-free diet [3–9]. We describe a patient in whom celiac disease was the cause of persistent hyperamylasemia.

## Case Report

A 52-year-old woman was referred to our outpatient clinic with a 6-month history of weakness, weight loss of 6 kg, maculopapular rash on her legs and persistent hyperamylasemia for 3 months (amylase >1,400 U [normal 35–120 U] and very low 24-hour urine amylase and amylase clearance/creatinine clearance ratio (1.9% [normal  $3.0 \pm 1.1\%$ ]), consistent with macroamylasemia. Her medical history was remarkable only for several years of chronic normocytic normochromic anemia (Hb 9.9 g%) and osteoporosis. She was not receiving any medications. Other than a sallow pallor and the maculopapular rash on both legs, the physical examination was completely negative. Laboratory results: thrombocytosis (510,000 platelets/ml), albumin 3.1 g, AST 60 U (normal 15–30), rheumatoid factor 81 (normal 0–15), anti-mitochondrial antibody (AMA) 1:80. The rest of the laboratory results were normal. An abdominal computerized tomogram with contrast yielded no pathological findings. Biopsy from the skin lesions on her legs revealed leukocytoclastic vasculitis.

Our patient had macroamylasemia and anemia, which can present as an autoimmune disease, such as celiac disease. Her serology for celiac disease was strongly positive, and she underwent esophagogastroduodenoscopy which revealed scalloping of folds in the second part of the duodenum. A histological evaluation confirmed the diagnosis of celiac disease. The patient was started on a gluten-free diet which led to a significant clinical improvement two months later: she felt generally stronger, had gained weight, the rash had disappeared, the serum amylase values were normal, hemoglobin had increased to 11.7 g%, and there was a return of serum albumin to normal values.

## Conclusion

The case we describe demonstrates an association between celiac disease and other medical conditions that present as macroamylasemia. Celiac disease was the cause and first manifestation of her persistent hyperamylasemia. We recommend ruling out celiac disease in cases of unexplained hyperamylasemia.

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Conflict of interest: none whatsoever.