

A Rare Cause of Hyperamylasemia: Macroamylasemia

Hiperamilazeminin Nadir Bir Nedeni: Makroamilazemi

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Abstract

Elevated serum amylase activity is commonly seen in acute pancreatitis and salivary gland inflammation, but also rarely seen in a benign phenomenon, macroamylasemia, that does not require treatment. Hyperamylasemia is a laboratory finding that may cause diagnostic failure occasionally in patients presenting with abdominal pain. Elevated serum amylase levels and abdominal pain may lead to misdiagnosis as acute pancreatitis in macroamylasemic patients. Macroamylasemia should be considered in patients with abdominal pain and hyperamylasemia but without imaging findings of pancreatitis. Here, we present a patient with macroamylasemia who had abdominal pain and persistent hyperamylasemia. The patient had no complaints but persisting hyperamylasemia (450 U/L) and normal lipase activity at the follow-up outpatient visit three months later.

Öz

Artmış serum amilaz aktivitesi akut pankreatit ve tükürük bezi enflamasyonunda sıklıkla görülmekle birlikte nadir olarak, tedavi gerektirmeyen benign bir fenomen olan makroamilazemide de görülmektedir. Hiperamilazemi karın ağrısı ile başvuran hastalarda zaman zaman tanısız karışıklığa yol açabilen bir laboratuvar bulgusudur. Serum amilaz düzeyi yüksekliği ve karın ağrısı, makroamilazemik hastaların yanlış akut pankreatit tanısı almalarına yol açabilir. Karın ağrısı ve hiperamilazemisi olan ancak pankreatiti destekleyen görüntüleme bulgusu olmayan hastalarda makroamilazemi de akılda bulundurulmalıdır. Burada karın ağrısı ve kalıcı amilaz yüksekliği ile başvuran makroamilazemili bir olguyu sunmaktayız. Hastanın şikayeti olmadan, üç ay sonra poliklinik takibinde lipaz değeri normal iken amilaz yüksekliği (450 U/L) devam etmekteydi.

Keywords

Abdominal pain, hyperamylasemia, macroamylasemia

Anahtar Kelimeler

Karın ağrısı, hiperamilazemi, makroamilazemi

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Introduction

Elevated serum amylase activity is commonly seen in acute pancreatitis and salivary gland inflammation, but also rarely seen in a benign phenomenon, macroamylasemia, that does not require treatment. Every abdominal pain and amylase increase may not indicate pancreatitis (1,2). Hyperamylasemia is a laboratory finding that may cause diagnostic failure occasionally in patients presenting with abdominal pain. Here, we present a patient with macroamylasemia who had abdominal pain and persistent hyperamylasemia.

Case Report

A 63-year-old female patient was admitted to our gastroenterology department with intermittent abdominal pain in the umbilical region radiating to the back, which was unrelated to nutrition, lasting for one year. Her medical history and family history were unremarkable. There was not a history of drug or herbal treatment. Physical examination findings were normal. Complete blood count, erythrocyte sedimentation rate and C-reactive protein levels were normal. Serum amylase level was 517 U/L (reference range: 25-125 U/L). The other biochemical laboratory results including serum lipase were normal. Abdominal ultrasound and computed tomography revealed bilateral, multiple renal parapelvic cysts. The pancreatobiliary system was normal. Ultrasound imaging of the parotid and submandibular salivary glands, performed in accordance with the recommendations of the otorhinolaryngology consultant physician, was normal. Thus, we excluded salivary gland pathology. During follow-up, it was observed that elevation of serum amylase levels persisted. Serum amylase levels were 480 U/L and 493 U/L, respectively in measurements performed at one-month intervals. Amylase and creatinine levels were measured simultaneously in serum and spot urine samples in order to determine the amylase creatinine clearance ratio (ACCR). ACCR was determined using the formula $[\text{urine amylase (U/L)} \times \text{serum creatinine (mg/dL)}] / [\text{serum amylase (U/L)} \times \text{urine creatinine (mg/dL)}] \times 100$. The test results were as follows: urine amylase: 134 U/L, serum amylase: 493 U/L, urine creatinine: 63.5 mg/dL, and serum creatinine: 0.52 mg/dL. ACCR was calculated to be 0.22% (reference range: 1.8%-3.2%). Macroamylasemia was diagnosed on the basis of these findings. Hyoscine-N-butylbromide + medazepam and trimebutine maleate treatment was started. The patient had no complaints but persisting hyperamylasemia (450 U/L) and normal lipase activity at the follow-up outpatient visit three months later. We did not study pancreatic isoenzymes.

Discussion

Hyperamylasemia is not always seen in pathological situations. Macroenzymes are serum enzymes having a higher molecular mass than the corresponding enzyme under physiological or pathophysiological

situations (3). Macroenzymes have a slower clearance rate and thus cause an increase in the activity of the corresponding enzyme in serum samples (4). Most commonly, macroenzymes are composed of an enzyme-immunoglobulin complex (5). Macroamylase was the first macroenzyme described by Wilding et al. (6) in 1964. The prevalence of macroamylasemia has been estimated at 1% in normal amylasemic patients and 2.5% in hyperamylasemic patients (3). The most useful diagnostic method for identification of macroamylasemia is the evaluation of amylase levels in serum and urine together, due to the technical difficulties in performing specific agarose-gel electrophoresis (1,5). Coexistence of macroamylasemia and various diseases such as celiac disease (7), ulcerative colitis (8), Crohn's disease (9), haematological malignancies (10,11), systemic lupus erythematosus (12), and rheumatoid arthritis (13) has been reported in the literature. Although macroamylasemia is a benign phenomenon and does not require a specific therapy, these associations make it more intriguing as a possible diagnostic marker. We could not find a concomitant disease in our patient.

Elevated serum amylase levels and abdominal pain may lead to misdiagnosis as acute pancreatitis in macroamylasemic patients, albeit Cho et al. (14) reported a case of macroamylasemia that occurred immediately after acute pancreatitis. It would be important to clinicians to be aware of the presence of macroamylasemia as one of possible causes of increased serum amylase activity in order to avoid unnecessary, expensive, invasive diagnostic procedures. Macroamylasemia should be considered in patients with abdominal pain and hyperamylasemia who have no imaging findings related to pancreatitis.

Ethics

Informed Consent: Consent form was filled out by all participants.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: D.Ç., Concept: A.C., Design: A.C., Data Collection or Processing: A.C., Analysis or Interpretation: A.C., A.Ö.K., M.H.Y., Literature Search: A.C., Writing: A.C.

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