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Macroamylasemia in a Patient with Acute Appendicitis : A Case Report

Macroamylasemia is a condition of persistent, elevated serum amylase activity with no apparent clinical symptoms of a pancreatic disorder. In Korea, however, no such case has been reported to date. We report a case of a 17-year-old female diagnosed with macroamylasemia and acute appendicitis. One day earlier, she developed epigastric and right lower quadrant abdominal pain. She was characterized by high level of serum amylase, but normal lipase. Amylase isoenzyme analysis demonstrated increased fraction of salivary type and follow-up amylase level was persistently increased. Immunofixation disclosed the macroamylase binding with an immunoglobulin, consisting of IgA and kappa chain. The patient was treated by appendectomy, and the abdominal pain subsided.

Key Words: *Amylases; Appendicitis*

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INTRODUCTION

Macroamylasemia is a condition of persistently elevated serum amylase activity with no apparent clinical symptoms of a pancreatic disorder (1-2). It was attributed to the presence of a large-sized amylase-immunoglobulin complex which precluded its excretion into urine even though renal function was unimpaired (1). Macroamylasemia is found in approximately 0.4% of the general population in Western countries (3, 4). To the best of our knowledge, however, this is the first report of macroamylasemia in Korea.

We recently experienced a case of macroamylasemia in a patient with acute appendicitis. After treatment with appendectomy, abdominal pain subsided, but serum amylase activity was persistently elevated. There was no reasonable explanation for the cause expect for macroamylasemia.

CASE REPORT

A 17-year-old female was admitted with epigastric and right lower quadrant abdominal pain for one day. The abdominal pain was relatively localized in right lower quadrant (RLQ) abdomen and did not radiate into the flank or right inguinal area. She was free from febrile sense, chills, or diarrhea, but three bouts of nausea developed during abdominal pain. She did not have any history of hepatitis, diabetes, tuberculosis or alcohol intake.

On admission, she was acutely ill-looking. Her blood pressure was 130/90 mmHg, respiration rate 21 per min, and body temperature 36.4℃. Physical examination revealed clear lung and heart sounds. Abdominal physical examination showed marked decreased bowel sounds, soft abdomen, localized abdominal tenderness with rebound tenderness in RLQ abdomen, but neither liver or spleen was palpated.

Laboratory evaluation revealed a white blood cell count of 7600/μL, hemoglobin 13.5 g/dL, hematocrit 38.7%, aspartate aminotransferase 14 IU/L, alanine aminotransferase 5 IU/L, total protein 6.4 g/dL, albumin 4.2 g/dL, alkaline phosphatase 49 IU/L, total bilirubin 0.6 mg/dL, total cholesterol 148 mg/dL, triglyceride 58 mg/dL, blood glucose 109 mg/dL, creatinine 0.8 mg/dL, amylase 639 U/L, and lipase 36 U/L. Amylse/creatinine clearance ratio was 0.5% and salivary type isoamylse was predominantly increased in fraction of 86.0% by two consecutive electrophoretic isoenzyme analysis (Fig. 1). Immunofixation disclosed the macroamylase binding with an immu-

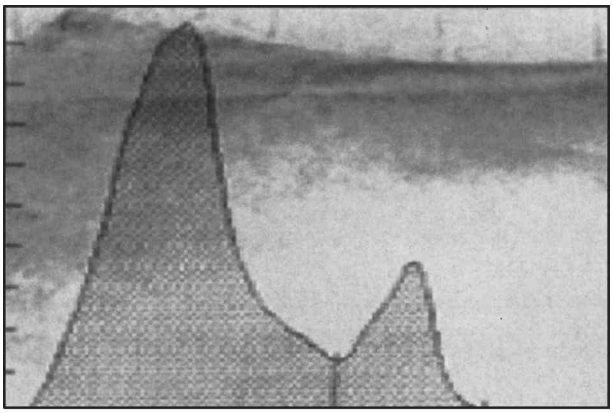

Fig. 1. Serum amylase isoenzyme electrophoresis showed increased total amylase activity. S-type isoamylase was 84.0% and P-type isoamylase was 16.0%.

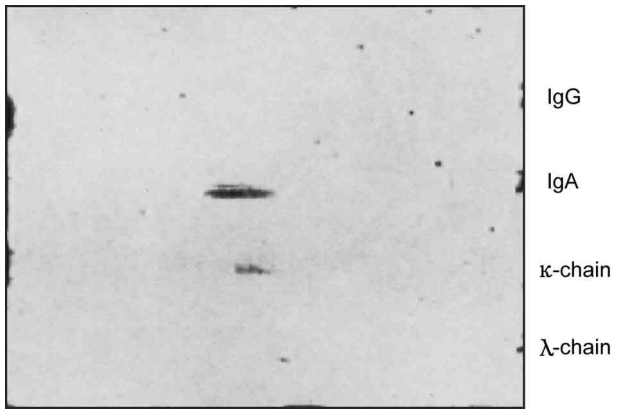

Fig. 2. Serum amylase immunofixation showed the macroamylase binding with an immunoglobulin which consists of IgA and kappa chain.

noglobulin, consisting of IgA and kappa chain (Fig. 2). Enzyme-linked immunosorbent assay (ELISA) for human immune deficiency virus was negative. Serological tests for hepatitis B and C were all negative. The urinalysis result was normal. Chest radiography showed no abnormal infiltration or mass in both lung fields and abdominal computed tomography revealed no significant abnormalities, including in the pancreas. Diagnosis of acute appendicitis was made from symptoms and physical findings, but routine preoperative laboratory finding in serum amylase was high up to 639 U/L. Appendectomy was performed and abdominal pain subsided soon. However, follow-up of serum amylase activity, two days after operation, showed 552 U/L and persistently increased values between 500 to 600 U/L in three months post-operative period (Table 1).

DISCUSSION

Amylase are enzymes that catalyze the hydrolysis of amylopectin, amylose, glycogen, and their partially hydrolyzed products (1). α-amylase occurs in animal tissue and fluid, β-amylse found in both animals and plants, and γ-amylase is found in numerous fungi (1). Of these three amylase, only α-amylase is of clinical interest and is predominantly of pancreatic and salivary origin. α-amylase of pancreatic and salivary origin is abbreviated to P-type and S-type isoamylase, respectively. These two types of amylase are closely related enzymes but also have organ-specific variations. P-type is synthesized by the acinar cells of the pancreas and account for 40% in all measurable serum amylase (5, 6). On the other hand, S-type is present in parotid, sweat gland, lung, and fallopian tube, and represents about 60% in origin (1, 5, 6).

Table 1. Serum amylase and lipase levels of the patient

Post-Op. days	Amylase (U/L)	Lipase (U/L)
-1	639	36
0	514	21
2	468	47
6	500	-
10	552	38
26	534	-
60	654	49
90	557	6

Serum amylase is increased in acute pancreatitis on the initial day of symptoms and remains elevated in most patients (7). Increases in total serum amylase reflect leakage of P-type isoamylase from the inflamed pancreas with subsequent reabsorption into the systemic circulation from venous pathways. However, in intra-abdominal conditions such as intestinal perforation, disease of salivary glands, macroamylasemia, or tumors, total serum amylase activity is also increased (7, 8). Among these numerous clinical situations unrelated to the pancreas, macroamylasemia is a condition of persistently elevated serum amylase activity with no apparent clinical symptoms of a pancreatic disorder (1, 2, 9) and is found in approximately 0.4% of the general population (3, 4). In macroamylasemia, a macromolecular complex consisting of amylase linked to immunoglobulins circulates in the plasma and usually causes benign hyperamylasemia with low or normal amylasuria. In this case, immunofixation consisting of an macromolecular complex between amylase and IgA-kappa chain immunoglobulin disclosed the macroamylase. A variety of conditions such as malabsorption, cancer, liver disease, and diabetes has been reported to be associated with macroamylasemia (2, 10, 11). Thus, it

is clinically important to differentiate macroamylasemia from other conditions associated with hyperamylasemia, especially in patients with abdominal pain.

Macroamylasemia per se is not a disease entity because no clinical symptoms is consistently accompanied. It is an benign condition that may occur in apparently healthy individuals and is found more frequently in males than in females (2, 6). Any patient with hyperamylasemia, a very low amylase/creatinine clearance ratio, and normal renal function should be considered for the possibility of having macroamylasemia (1, 8). The biochemical characteristics of clinical implications are unknown, requiring different methods for its detection that will give a better efficacy and reproducibility. In this case, initially there was clinical problem in diagnosing macroamylasemia because acute appendicitis was known to increase serum activity of amylase (8). However, serum amylase activity was persistently increased for three months during follow up after treatment with appendectomy in our patient. Serum creatinine level, amylase/creatinine clearance ratio, and serum lipase levels were all within normal limits. In our case, direct demonstration of the existence of macroamylase molecule, persistently increased serum amylase activities combined with normal serum lipase, normal creatinine and low amylase/creatinine clearance ratio were diagnostic. In addition, acute pancreatitis could be excluded because serum P-type isoamylase was not increased and the pancreas in the abdominal CT scan was normal. The patient had no classical symptoms and signs of acute pancreatitis. Because the occurrence of macroamylasemia may be an early sign of disease (2), however, it is needed to monitor clinical symptoms and signs in the patient.

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