

Macroamylasemia: A Benign Cause for High Serum Amylase

Hyperamylasemia is the most common biochemical marker for the diagnosis of acute pancreatitis, but has low specificity. A 9-year-old girl presented to our hospital with complaints of pain abdomen, fever and vomiting for a week. Her serum amylase done elsewhere was 3500 U/L, and with suspicion for acute pancreatitis, she was referred to our center. On examination, her abdomen was soft with mild tenderness in right iliac area. Her blood investigations indicated a neutrophilia (white blood cell count $13 \times 10^9/L$), high serum amylase (4310 U/L), normal serum lipase (32 U/L), and normal serum creatinine. Ultrasonography of abdomen showed mild peri-appendicular edema and normal pancreas. In view of above findings and non-fulfillment of diagnostic criteria for acute pancreatitis [1], patient was managed conservatively as a case of acute appendicitis. At follow-up after 4 weeks, she was asymptomatic, but her serum amylase continued to be abnormally high (2588 U/L), with normal lipase. Ultrasound and contrast-enhanced computed tomography of abdomen were normal. With no obvious intrabdominal or salivary pathology to explain such high amylase values in absence of renal failure, possibility of macroamylasemia was considered. Amylase creatinine clearance ratio (ACCR) [(urine amylase/serum amylase) \times (serum creatinine/urine creatinine) $\times 100$] estimation was done which was 0.36 (normal 1-4%), confirming the diagnosis of macroamylasemia.

Apart from pancreatitis, serum amylase is increased in intestinal obstruction, perforated peptic ulcer, biliary tract disease, tubo-ovarian pathology, diabetic ketoacidosis, salivary gland lesions, and renal failure. Macroamylasemia, a rare but benign condition, results due to circulating macroamylase complexes, which cannot be cleared by the renal glomeruli. Amylase

aggregation with immunoglobulins, usually IgA (92%), increases their mass to 10^5 Daltons, and thus these macromolecules are not filtered [2]. In the absence of renal failure, a low ACCR is diagnostic of macroamylasemia [3]. In acute pancreatitis, amylase clearance increases significantly, and thus ACCR is increased.

In adults, macroamylasemia is reported in 2.5% of all hyperamylasemic patients. However, it is rarely reported in children. There are few pediatric case reports of macroamylasemia with Celiac disease and Crohn's disease [4,5].

Failure to immediately identify macroamylasemia as the cause for persistent hyperamylasemia can lead to extensive investigations and confusion; hence, early suspicion and identification is crucial to eliminate diagnostic perplexity.

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