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Macroamylasemia in pediatrics: Case report and literature review

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Introduction: Macroamylasemia is a benign condition reported in 2.5% of adult patients with hyperamylasemia; however, only few cases have been reported in children.

Case Report: A 5-year old boy was presented to a tertiary-care hospital with periumbilical pain, vomiting and fever for one day. Patient had history of recurrent abdominal pain over the prior year. His mother had history of pancreatitis. On examination his abdomen was soft with mild diffuse tenderness, but no peritoneal signs. On admission his serum amylase was 1540 IU/L and lipase was 158 IU/L. An abdominal ultrasound and contrast-enhanced computed tomography were normal. Patient was initially managed conservatively as a case of acute pancreatitis. By his second day of admission he was already asymptomatic. His serum amylase remained persistently elevated during his admission and at 1-month follow up. Serum lipase remained normal throughout. A serum isoamylase analysis revealed pancreatic isoenzyme predominance. He had a normal renal function. His amylase/creatinine clearance ratio (ACCR) was 0.03; confirming the diagnosis of macroamylasemia.

Discussion: Macroamylasemia results from circulating large-sized amylase complexes that cannot be excreted in the urine. An abnormally low ACCR reflects the retention of these large complexes by the kidney. Complex formation with IgA immunoglobulin has been reported in 92% of adult patients with macroamylasemia. The etiology and natural history of macroamylasemia remains unclear. Based on a case review of 16 pediatric patients, abdominal pain (64%) is the most common presenting symptom, but 14% of patients can be asymptomatic. ACCR was obtained in 88% of patients with a value of less than 1 in all of them. Persistent amylase elevation was noted on subsequent testing for every reported case. There were no associated conditions in 50% of pediatric cases, but it has been described in patients with celiac disease, IgA deficiency, malignancies, inflammatory bowel disease, hepatitis and autoimmune hypothyroidism. Only one patient had family history of macroamylasemia, but 2 patients had relatives with elevated amylase of unknown etiology.

Conclusion: Macroamylasemia should be considered in the differential diagnosis of patients who have persistently elevated serum amylase, but do not exhibit any features of pancreatitis or salivary gland inflammation. The ACCR may be a rapid and inexpensive screening test to preclude unnecessary tests and therapies.

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