Case report

Hyperamylasemia as manifestation of gastrointestinal involvement in adult type Henoch-Schönlein purpura

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SUMMARY

Henoch-Schönlein purpura is a generalized vasculitis presenting with purpura, glomerulonephritis, cryoglobulinemia, arthralgias and acute abdominal pain. It occurs mainly in childhood and serum IgA is increased in half of the patients. A 54-year-old male patient was admitted to the Department of Nephrology because of lower extremities purpura, mild abdominal pain and acute renal failure (creatinine clearance at 60 ml/min). Physical examination showed both lower extremities purpura and mild epigastric tenderness. Abdominal imaging did not show anything remarkable. Further laboratory investigation showed serum urea 75 mg/dl, serum creatinine 1.9 mg/dl, amylase 600 UI/ml (normal values up to 90 UI/ml) and albumin 3.2 gr/dl. Serum isoamylase analysis was compatible with that of pancreatic origin. Urine amylase was 138 UI/ml, while the examination of the urinary sediment showed microscopic hematuria and proteinuria. In the 24-hour urine collection albuminuria reached nephrotic syndrome levels (4.8gr/24h). There was no evidence of elevated IgA levels, of cryoglobulinemia or of circulating immune complexes. Skin and kidney biopsies confirmed Henoch-Schönlein purpura diagnosis. In conclusion, we report on an adult patient with hyperamylasemia during Henöch-Schonlein purpura initial diagnosis.

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INTRODUCTION

Henoch-Schönlein purpura is a generalized vasculitis that is characterized by a combination of purpuric rash, arthralgias, glomerulonephritis and gastrointestinal symptoms.[1-3]

Gastrointestinal involvement occurs in up to 60% of patients and manifests as abdominal pain with diffuse tenderness, nausea and vomiting.[4-5] Abdominal pain is thought to be due to edema and hemorrhage into the small bowel wall, secondary to a small vessel vasculitis.[6] Pancreatitis secondary to Henoch-Schönlein purpura is rare.[7-15] Hyperamylasemia with or without clinical evidence of an acute abdominal episode is extremely rare in Henoch-Schonlein purpura cases. In addition, elevation of serum amylase levels is self-limiting when Henoch-Schönlein purpura is properly treated.

Herein we report on an adult patient with hyperamylasemia during Henöch-Schonlein purpura initial diagnosis.

CASE REPORT

A 54-year-old patient was admitted to the Department of Nephrology because of lower extremities purpura, mild abdominal pain and acute renal failure. The patient had a negative history of previous hospital admissions or any other chronic illness.

Physical examination showed bilateral lower extremities purpura and mild epigastric tenderness. Blood pressure was 150/80 mmHg, pulse rate was 78/min and body temperature was 36.8° C. Blood testing showed Ht

42%, Hb 13.2g/dl, white cell count $15.4x10^9$/L. Serum testing showed glucose 98 mg/dl, urea 75mg/dl, creatinine 1.9 mg/dl, albumine 3.2g/dl and amylase 600 UI/ml (normal values up to 90 UI/ml). Ratio of amylase clearance/creatinine clearance was 8%. Serum isoamylases analysis was compatible with that type of pancreatic origin. Liver function tests were within normal limits and no acid-base balance or electrolyte deterioration was evident. Urine amylase was 338 UI /ml and urinary sediment examination showed microscopic hematuria and proteinuria. In the 24-hour urine collection albuminuria reached the levels of nephrotic syndrome (4.8gr/24h). There was no evidence of elevated IgA levels or cryoglobulinemia. Rheumatoid factor and CRP were within normal range, complement (C3, C4) was normal and anti-DNA, ANA and cardiolipins were negative. Abdominal ultrasound and computed tomography showed no pancreatic abnormality. Kidney biopsy showed rapidly progressive glomerulonephritis and skin biopsy confirmed Henoch-Schönlein purpura diagnosis.

The patient was started on therapy- as rapidly progressive glomerulonephritis-with 1g bolus methylprednizolone i.v for 3 days and after that methylprednizolone orally (32mg/day) and cyclophosphamide (150 mg/day) per-os. Amylase levels turned were normalized at the 5^{th} day of treatment. In the two-year follow-up the patient is clinical status is excellent with no medication and renal function is normal with serum creatinine at 0.8 mg/dl and proteinuria below 250 mg/day

DISCUSSION

In this particular case of Henoch-Schonlein purpura with renal and gastrointestinal involvement this moderate hyperamylasemia could be the result of either pancreatic or extra-pancreatic cause.

Pancreatic involvement represents a rare and generally benign complication of Henoch-Schönlein purpura.[7-12] The clinical presentation in such cases of pancreatic involvement is usually abdominal pain. This kind of pain can be expressed either as pancreatitis or as a generalized digestive system involvement including the pancreas.[6]

The exact pathophysiologic mechanism of pancreatic inflammation is not yet well recognized. A small vessel vasculitis, - especially of vessels of pancreatic origin - leading to inflammation may explain this pancreatic involvement occurring in Henoch-Schönlein purpura cases.[6] This pancreatic involvement ranges from mild serum amylase elevation to the clinical presentation of a severe pancreatitis episode.

Hyperamylasemia and long-standing protein-losing enteropathy, during the course of Henoch-Schönlein purpura, may imply pathophysiological mechanisms, which should be further investigated, particularly relating to the kidneys and the gastrointestinal tract.[16]

In this case hyperamylasemia due to pancreatic isoenzyme elevation was the unique laboratory finding of gastrointestinal involvement in Henoch-Schonlein purpura, as no radiological evidence of pancreatic damage was found.

All other cases reported in the literature clearly showed evidence of pancreatic damage.[7-12]

Increased amylase production may be the result of several extra-pancreatic diseases, mainly of the abdominal area. It must also be noted here that renal failure may also cause hyperamylasemia due to reduced amylase clearance.

In addition, amylase levels in blood and urine were moderately increased while inflammation markers ESR and CRP were within normal limits. Furthermore, amylase levels normalized during administration of high doses of corticosteroids.

In conclusion, we diagnosed a patient with Henoch-Schonlein purpura with gastrointestinal involvement and hyperamylasemia without any evidence of pancreatitis.

This case emphasizes again that pancreatic imaging as well as hyperamylasemia should be further investigated and followed up in any patient with Henoch-Schönlein purpura and abdominal pain in order to early recognize and diagnose a subclinical pancreatic involvement.

Hyperamylasemia in such cases does not always imply pancreatic involvement as other extra-pancreatic causes may result in this amylase elevation. However, the mechanism of hyperamylasemia still remains unknown.

Furthermore it seems that early recognition of pancreatic involvement is very critical during Henoch-Schönlein purpura clinical course, as optimal treatment may prevent further complications and avoid prolonged hospitalization.

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