CASE REPORT

Gullo's Syndrome: A Case Report in a Pediatric Patient and Literature Review

Lotah Ali[1], Osama Hamdoun[1], Asia Al Mulla[1], Shamma Alzaabi[2], Amar Al Shibli[2]

[1]Department of Academic Affairs, Tawam Hospital, Al Ain, UAE, [2]Department of Pediatrics, Tawam Hospital, Al Ain, UAE

ABSTRACT

Benign pancreatic hyperenzymemia (Gullo's syndrome) is characterized by increase of the pancreatic enzymes lipase and amylase in the absence of a pancreatic disease over a period of time (more than 12 months) Gullo's syndrome is a diagnosis of exclusion of pathological causes of deranged pancreatic enzymes. There is no clinical or pathological evidence of pancreatic disease. The disease was described mostly in adults and only few cases where reported in children. Herein, we are reporting a 3-year-old male who was diagnosed as a case of Gullo's syndrome based on the clinical and laboratory data.

Key words: Benign pancreatic hyperenzymemia, Gullo's syndrome, pancreas, amylase, lipase

INTRODUCTION

Benign pancreatic hyperenzymemia (BPH) or Gullo's syndrome is a newly identified syndrome characterized by an abnormal increase in serum pancreatic enzymes (usually three-fold from the normal) in the absence of pancreatic disease.[1] In the majority of cases (95%), the hyperenzymemia concerns all pancreatic enzymes; in 5% of cases, it is possible to observe an increase of only the amylase and rarely of only the lipase.[2]

There is no clinical or pathological evidence of pancreatic disease. Gullo's syndrome is a diagnosis of exclusion of other causes of elevation of pancreatic enzymes.[3]

CASE PRESENTATION

A 3-year-old male with West syndrome and seizer disorder was admitted to the hospital because of increase in the seizer frequency. He was kept on antiepileptic medications. Pancreatic enzymes were done for the recurrent vomiting and they were high.

Laboratory tests were performed to try to diagnose the reason of the pancreatitis and all tests including viral studies and lipid profile and other tests were normal [Table 1].

Computed tomography and magnetic resonance cholangiopancreatography imaging were also done and there was no evidence of pancreatic inflammation, structural, or ductal anomalies.

The patient was known case of epilepsy and was getting Keppra that was stopped gradually with no improvements in the pancreatic enzymes.

Pancreatic enzymes remained high over a period of 14 months with high levels of both amylase and lipase with fluctuating level that was always above normal [Figure 1].

DISCUSSION

Pancreatitis is diagnosed based on the presence of two of the three following criteria: Typical abdominal pain, characteristic imaging findings, and amylase levels of three-fold or higher than the upper limit of normal values and elevated lipase.[4]

Address for correspondence:
Amar Al Shibli, Departmentof Pediatric, Tawam Hospital, Al Ain, Post Box: 15258, UAE.

Table 1: Demographic and laboratory data of the patient at the time of the presentation

Laboratory test	Result
Amylase	328 IU/L
Lipase	172 IU/L
Aspartate transaminase	25 IU/L
Alanine transaminase	<5 IU/L
Bilirubin total	5.7 micromol/L
Direct bilirubin	2.1 micromol/L
Albumin	35 g/L
Alkaline phosphatase	122 IU/L
Gamma-glutamyl transferase	9 IU/L
Cholesterol	3.51 mmol/L
High-density lipoprotein	0.60 mmol/L
Low-density lipoprotein	2.25 mmol/L
Triglyceride	1.24 mmol/L
Chol/high-density lipoprotein	5.8
White blood cell	4.9×10^{9}/L
Red blood cell	4.44×10^{12}/L
Hemoglobin	9.9 g/dl
Platelets	172×10^{9}/L
Red blood cell distribution width	16.7%
Phosphorous	1.29 mmol/L
Parathyroid hormone	2.36 pmol/L
Calcium level	2.53 mmol/L
Blood culture	No growth
Urine culture	No growth
Urine analysis	Not significant
Viral panel including respiratory syncytial virus, para-influenza 1, 2, and 3, adenovirus, and influenza virus A and B	Negative

Table 2: Causes of increase serum amylase and lipase. Modified from Wiederkehr *et al*.[6]

Amylase	Lipase
Acute pancreatitis	Acute pancreatitis
Pancreatic pseudocyst	Pancreatic pseudocyst
Chronic pancreatitis	Chronic pancreatitis
Biliary disease	Biliary disease
Structural pancreatic abnormalities	Structural pancreatic abnormalities
Intestinal obstruction	Intestinal obstruction
Intestinal ischemia	Intestinal ischemia
Appendicitis	Appendicitis
Renal insufficiency	Renal insufficiency
Parotitis	Anorexia/bulimia nervosa
Diabetic ketoacidosis	Hepatitis C infection
Lung carcinoma	Malignancy
Head trauma	
HIV infection	
Ovarian pathologies (cyst and neoplasm)	

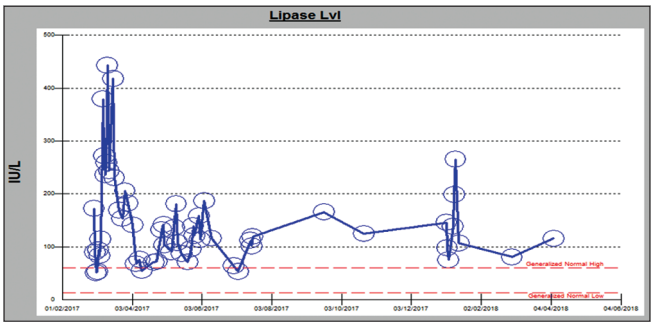

Figure 1: The trend of the lipase enzyme over the period of the follow-up

High serum amylase and lipase usually related to different pancreatic, intestinal, gallbladder, metabolic diseases, and extra pancreatic abnormalities of the pancreatic ducts such as recurrence of chronic pancreatitis, diabetic ketoacidosis, and chronic viral liver disease[5-14] [Table 2].

Gullo's syndrome was first described in 1996 by Gullo's with raised pancreatic enzymes in normal individuals without any pancreatic disease.[15] Gullo advises that the BPH diagnosis can only be given after an observation period of with no symptoms and/or signs of pancreatic diseases and no structural changes of the organ during the follow-up period.[1]

The diagnosis will be based on the elevations and significant undulations of pancreatic enzyme serum concentrations occur on a day-to-day basis for 5 consecutive days for a period of 1 year.[16]

The mechanism of increase enzymes is still not very clear. It may be a result from a defect in the intracellular transport of pancreatic enzymes in exocrine pancreatic cells, may be responsible for the increased passage of enzymes into the blood circulation,[17] or may also be caused by changes in the Wirsung conduct by secretin stimulation.[18] The fluctuating behavior could depend on the degree of the cellular defect, with the passage of enzymes being sporadic when the defect is mild and more frequent when it is more severe or extensive.[1,3] BPH found within family members has led to the hypothesis that there could be some genetic basis for Gullo's syndrome. There were no genes linked directly to the syndrome.[19,20]

Most of the reported cases affect males with males to females in ratio of 1.5:1.[16] Most of the cases were reported in adult patients; however, BPH has been documented in children also.[19-21]

Gullo's syndrome was previously considered to be a disease afflicting primarily Western populations and infrequently studied in Asian countries.[22]

Proper diagnosis of BPH is important to prevent multiple and expensive diagnostic tests or useless hospitalizations or therapies.[23]

CONCLUSION

The presence of persistent elevated pancreatic enzymes with the absence of definitive pancreatic disease should rise the possibly of Gullo's syndrome. Correct diagnosis of this disease is important to avoid costly test duplication, unfounded anxieties, and multiple consultations.

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