Calcific chronic pancreatitis in a 5 year old girl

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Introduction

Calcific chronic pancreatitis, sometimes referred to as Juvenile Tropical Pancreatitis Syndrome when it affects children, has been described in many parts of the world [1]. This Syndrome consists of pancreatic calcification associated with both exocrine and endocrine impairment singly or in combination and is commonly found in the tropical countries [2-4]. This condition is not common in Kenya. The importance of this case is its presentation in childhood and the immense suffering the girl had undergone before diagnosis.

Case report

A 5 year old girl presented with 18 month history of burning epigastric pain which initially was intermittent, non radiating with no associated vomiting or obvious precipitating or relieving factors lasting one to two days every month. After one year, the pain became more frequent, and this time aggravated by food intake, relieved by sitting up and leaning forwards and associated with occasional vomiting and diarrhoea. Two months before our review, she was seen in another hospital where all investigations done, including upper gastrointestinal endoscopy were reported as normal. She was subsequently put on antacids and advised to go back to school. However, her condition got worse with diarrhoea becoming more frequent associated with weight loss. She had no associated headaches, febrile episodes, cough or previous history of jaundice. She is the first born in a family of two siblings. The other sibling is alive and well at 3 years of age. Both parents are alive and well. Her paternal grandfather died in 1969. The rest of her grandparents are alive and well. No history of a similar illness has ever been reported in her family. She hails from Busia district, Western Kenya, but has lived in Nairobi almost all her life. However, when the illness started, she was on a visit to Busia where cassava is eaten occasionally. On examination, she was grossly wasted, unable to walk without support but had no pallor, jaundice, lymphadenopathy, cyanosis or peripheral oedema. Vital signs were normal and she weighed 16 kilogrammes. She had a non-tender splenomegaly of 4cm below the left costal margin, very tender epigastrium with no ascites. The rest of the physical examination was normal

Investigations

Laboratory

Full blood count: WBC 7.5 x 10^9/l (4-10), Hb 12.5g/dl (12-16), MCV 89.5 fl (81-99) and platelets were reported as adequate. WBC differential count was normal. Stool microscopy was normal and grew no pathogens on culture faecal fat 6.7g/day . Viral studies were not done. Liver function tests: Total bilirubin 13.4umol/l (1.71-20.5). Aspartate amino transferase 20 u/l (0- 33), Alamine amino transferase 22 u/l (0-43), Alkaline phosphatase 252 u/l (34-114), total protein 76.1g/l (60-80), Serum albumin 43.0 g/l (32-50); Blood chemistry: Blood urea nitrogen-1.6mmol/l (1.78-6.07), sodium-145mmol/l (130-149), potassium-3.9mmol/l (3.0-5.2), calcium 2.0 mmol/l (2.02-2.60), inorganic phosphate-1.74mmol/l, creatinine-56umols/l, Random blood sugar-4.3mmol/l, serum amylase-575Iu/l (25- 125) fasting triglycerides 0.6mmol/l (0.51-2.11). Glucose tolerance test 7.2 mmol/l (3.88-6.38) at 2 hours postprandial, Mantoux test: 4x5 mm.

*** In brackets are the normal ranges for the parameters.**

Radiology

Abdominal ultrasound showed moderate calcification within pancreatic tissue and distorted and headed pancreatic duct. Spiral CT Scan of abdomen showed similar results to ultrasound with tortuosity of the pancreatic duct. Chest radiograph was normal.

Management

The patient was started on oral pancreatic enzyme supplements taken after meals. The abdominal pain and diarrhoea subsided within two days. Serum amylase however, remained high. Patient gained 2kg within two weeks of treatment and went back to school. During subsequent follow up, she has remained largely asymptomatic and growth and development has been normal. At 10 years, 5 years later, she now weighs 30 kg and repeat abdominal ultrasound scan shows significant reduction in the pancreatic calcification.

Discussion

Pancreatic calcification is common in middle aged men with a positive history of alcohol abuse. Pancreatic calcification that affects young children who are born and reside in the tropics, notably Africa, Indian subcontinent, South-East Asia and Western pacific Islands is referred to as Tropical Pancreatic Pancreatitis [5-9]. Clinical presentation consists of abdominal pain, steatorrhoea, failure to thrive and diabetes mellitus in about one third of the patients. The hallmark of this disease is introductal pancreatic calculi and ductal dilatation [7]. The aetiology of this condition is unknown though it is thought to be associated with childhood protein calorie malnutrition, shortage of dieting lipids during childhood and ingestion of cassava [8-9]. Genetic predisposetion may also be important as has been noticed that mutation in series protease inhibitor, icaza type 1 (spink I) gene in north Indian patients with tropical calcific pancreatitis [8,9].

Management of these patients includes pancreatic supplements which are used for relief of abdominal pain and steatorrhoea, control of blood sugar using insulin in those who are diabetic and close surveillance to detect/prevent complications of diabetes.

Malabsorption surveillance would also allow early detection of pancreatic cancer as these patients tend to have increased susceptibility [8].

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