Case Report

FCPD: An unusual cause of abdominal pain; reminder of important clinical lesson

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Abstract

Fibrocalculous pancreatic diabetes (FCPD) is a type of chronic calcific non-alcoholic pancreatitis. This type is reported mostly in the developing tropical and subtropical countries. Definite diagnosis of FCPD requires younger age of onset, history of malnutrition, and presence of diabetes mellitus along with extensive pancreatic calcification and ductal calculi. Pain abdomen is one of the classical clinical triad of the disease with steatorrhoea and diabetes. In this case report we are presenting a case of 17 years old girl with past history of recurrent pain abdomen for last two years. She came to casualty with complaints of generalized weakness along with pain abdomen. She was diagnosed as case of FCPD with uncontrolled hyperglycemia on subsequent investigations. Importance of this case is that patient has multiple episode of abdominal pain over two years and she consulted several practitioners for the same and she was treated symptomatically without proper work-up. However, FCPD is a rare entity but should be considered in case of recurrent abdominal pain in young patient.

Key words

Fibrocalculous pancreatic diabetes, FCPD, Recurrent abdominal pain, Chronic pancreatitis, Abdominal pain, Tropical chronic pancreatitis.

Introduction

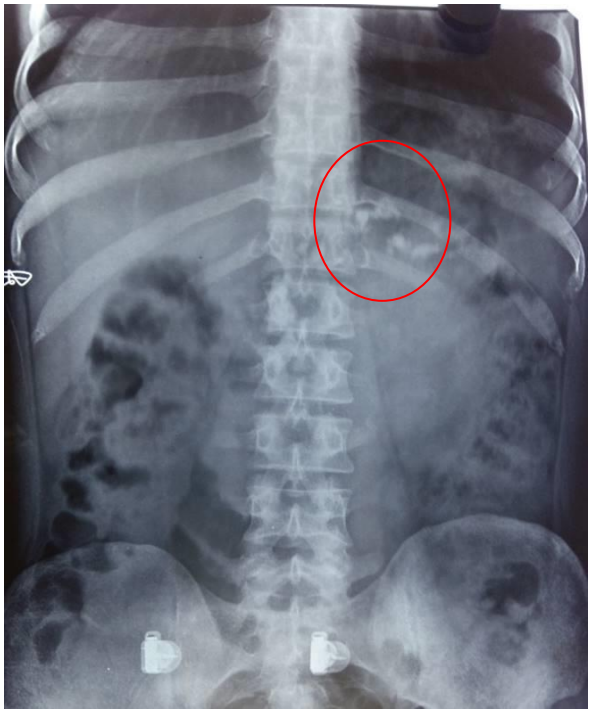
The fibrocalculous pancreatic diabetes (FCPD) is a diabetes mellitus secondary to tropical chronic pancreatitis (TCP) which is considered as a form of chronic calcific non-alcoholic pancreatitis. It generally presents in the second and third decades of life. It is associated with the formation of multiple pancreatic calculi and a high probability of developing insulin dependent diabetes mellitus. This type of DM is almost exclusively present in the developing countries of the tropical world including India [1]. Recurrent abdominal pain in young patient has

limited differential diagnosis. Fibrocalculous pancreatic diabetes (FCPD) or chronic pancreatitis is uncommon but important cause of it.

Case report

A 17 year old girl admitted to our hospital with history of pain abdomen and generalized weakness for last 2-3 days. Pain was mild and present over epigastrium without any radiation. Pain was not associated with nausea; vomiting, or loose stool. There was no history of fever, burning micturition or decrease urine output. She also had a history of nocturia for last few months. She had history of pain abdominal off and on for last two years for which she visited several practitioners and get treated. All her family members were healthy. Nobody had diabetes or hypertension. She does not have any addiction. She belongs to low socio economic status. On examination she was conscious, oriented, and afebrile. She was poorly nourished. There was mild pallor but no icterus, cyanosis or clubbing was noted. There was no lymphadenopathy or parotid land enlargement. Her body mass index was low (15.2 kg/m^2). Abdomen was soft. There was no organomegaly. There was mild tenderness over epigastrium. Neurological examination was normal. At the time of presentation random blood sugar was 375 mg/dl. The complete blood picture revealed mild leucocytosis of 13600/mm^3 (Normal 4000-10000). Her hemoglobin was 10.2 gm. /dl; platelet count was 296000/ mm^3. Her HbA1C level was 8.5; total serum protein level was 7.92 mg/dl. Her kidney and liver function tests were normal. Serum lipid levels were very low (Serum cholesterol-125.0, LDL-79.76, HDL- 34.8, VLDL-10.44, serum triglyceride was 52.2 mg/dl) Urine analysis showed glycosuria, without proteinuria. Urine for ketones was negative and ABG was also normal. The stool analysis revealed no fat globules, ova or cysts. Examination of fundus was normal. ECG showed normal sinus rhythm. Chest x-ray was normal. The plain X-Ray of abdomen revealed pancreatic calcifications (**Figure - 1**). Ultrasonography of abdomen showed diffuse calcifications in the head, body and tail of the pancreas. CT abdomen was done which confirmed the findings of Plain X-Ray abdomen and USG abdomen (**Figure - 2**). She was diagnosed as a case of FCPD with uncontrolled diabetes based on above clinical features and investigations. She was managed with intravenous fluid, insulin, and antibiotics along with another supportive measure. She was discharge after one week of admission on insulin and pancreatic enzyme supplement. She is doing well on follow-up.

Figure – 1: Abdominal X-ray, showing multiple pancreatic calcifications (red circle).



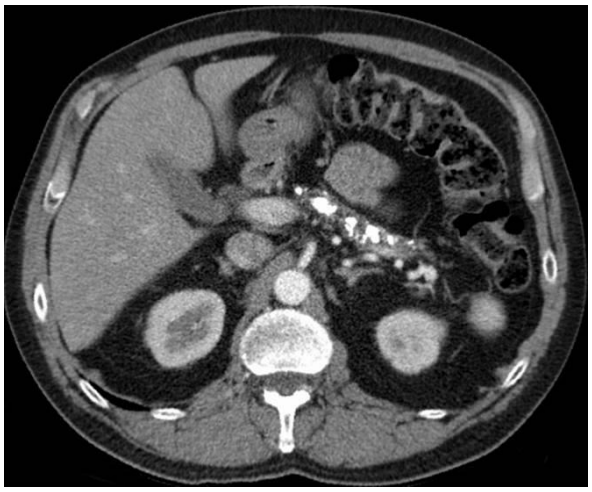
Discussion

FCPD is a juvenile form of chronic calcific, non-alcoholic pancreatitis, prevalent almost exclusively in the developing countries of the tropical world. In India, these cases have mostly been reported from south India, a few have also been reported from northern and eastern parts of India [2].

There are little data on the prevalence of FCPD in the general population. A study by Balaji, et al. found the prevalence of FCPD to be one in 793 individuals in Kerala. In a clinic based study

from Chennai, the prevalence of FCPD was found to have decreased from 1.6 % (for the years 1991–1995) to 0.2 % (during the years 2006–2010) [3].

Figure – 2: CECT scan of the abdomen showing multiple chunky ductal calcification in pancreas.



Tropical calcific pancreatitis (TCP) is a distinct variety of chronic pancreatitis, with a high prevalence being reported from southern India. The clinical presentation of TCP includes abdominal pain, steatorrhoea and weight loss, usually in early adulthood. The imaging classically shows multiple chunky calcifications in the pancreatic duct. Patients usually develop exocrine and endocrine dysfunction. When TCP progresses to diabetes, the condition is known as FCPD. FCPD has been classified later by the WHO as a form of diabetes secondary to the exocrine pancreas disease (Type 3c diabetes mellitus). Management of FCPD comprises of pancreatic enzyme supplementation and insulin therapy for diabetes [4].

Many patients give a past history of recurrent episode of abdominal pain in childhood or adolescence suggestive of relapsing pancreatitis. The pain is usually severe, epigastric in location and characterized by periods of remission and exacerbation. It radiates to the back on either side and is typically relieved by stooping forward or lying in a prone position. The pain usually abates by the time diabetes sets in. About one third of patients complain of passing bulky or oily stools. Stool was normal in our case. The low frequency of steatorrhoea has been attributed to low fat content of the diet. When the fat content of the diet was experimentally increased, steatorrhoea occurred in over 90% of patients [5].

Some of its distinctive features are younger onset, presence of large intra-ductal calculi, accelerated course of the disease, and high susceptibility to pancreatic cancer. These patients are at a higher risk of developing pancreatic cancer (about 100-fold) as compared to those with other forms of pancreatitis [6].

Multiple etiological factors have been thought to be associated with FCPD. These include malnutrition; toxic effects of cyanide derived from frequent cassava consumption, familial aggregation, genetic factors and increased oxidant stress from micronutrient deficiency (Vitamin C and A deficiencies).

Several studies have debated the theories that malnutrition have etiological role in TCP, they confirmed a link between the serine protease inhibitor, Kazak type 1 (SPINK 1) gene and TCP. It is a vital protease inhibitor that prevents unregulated or inappropriate activation of the pancreatic enzyme cascade by inhibiting trypsin activity [7, 8].

Demonstration of hyperglycemia and pancreatic calculi on plain abdominal X-Ray, abdominal computed tomography scan confirms the diagnosis as shown in the patient discussed. The diabetes is usually severe. Most patients require insulin for control of blood sugar. Interestingly despite requiring insulin for the control of diabetes, patients with FCPD rarely develop ketoacidosis even if insulin injections are withdrawn for prolonged periods. Similarly in our case she presented with hyperglycemia but no ketoacidosis. This is probably because of the residual pancreatic beta cell reserve, a low glucagon reserve and decreased adipose tissue mass. Other complications namely sight-threatening forms of retinopathy, maculopathy do develop in FCPD patients. In contrast macro

vascular complications are less common perhaps owing to the relative young age of the patients, their leanness and the low cholesterol levels. Neuropathy, nephropathy and left ventricular dysfunction also occur [9]. In our case patient doesn't have any micro or macro vascular complications.

Conclusion

Although fibrocalculous pancreatic diabetes remains a rare entity but it should be suspected in young patient with no family history of diabetes or autoimmune predisposition presented with recurrent abdominal pain. These patients need a regular monitoring to detect earlier multiple complications.

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