

Fibrocalculous Pancreatic Diabetes in a Bangladeshi Girl

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Abstract:

Fibrocalculous pancreatic diabetes (FCPD) is atypical but unique variety of diabetes, in which chronic inflammation and pancreatic calcification lead to exocrine and endocrine failure of the pancreas, primarily occurs in tropical countries. At an early age, these diabetic children are at risk of developing disabling and life-threatening complications, putting a major human and economic burden on families and societies. Here, authors report a case of young female patient presenting with the clinical features of FCPD.

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Introduction:

Diabetes is rising alarmingly in both developing and developed countries. Children with type 1 and type 2 diabetes are frequent, but diabetes due to other specific types are also reported in Bangladesh.^{1,2} Chronic pancreatitis is the most common pancreatic cause of diabetes. Chronic, calcific pancreatitis of unknown aetiology in children of tropical parts of the world is known as tropical chronic pancreatitis (TCP). The term FCPD is used when patients with TCP develop diabetes. FCPD was a subtype of Malnutrition related diabetes mellitus (MRDM). Later, MRDM was omitted and FCPD has been reclassified as a 'disease of exocrine pancreas' under the category of 'Other types of Diabetes'.^{3,4}

Zuidema described first case series of TCP from Indonesia in 1959. After that, similar cases were shared from many regions of tropical Asia and Africa, but mostly from the states of Kerala and Tamil Nadu of India.⁴ Both in Kerala and Chennai cases of FCPD decrease from 29.3% in 1964 to 0.03% in 1971 and 0.009% in 1980 and decreased from 1.6% in 1991-1995 to 0.2% in 2006–2010 respectively. The reasons for these downfall are not clear, but they may be connected to better nutrition as a result of rapid socio-economic growth.⁵ A recent study of North India, revealed FCPD was the cause of diabetes in 15%

cases⁶ and in Bangladesh, 25% of diabetic children had FCPD.² Rising trends of FCPD in these 2 hospitals are probably due to referral bias. The exact aetiology is not clear, but associated with genetic, dietary and inflammatory factors.⁷ Most Patients with FCPD are insulin deficient and require life-long insulin therapy.⁸

Case presentation: 'J', 15yr-old-girl, came from a rural area and belonged to low socio-economic background, presented with complaints of upper abdominal pain for 12 days, polyuria and polyphagia for last 2 months. She had total 4 episodes of severe abdominal pain in last 12 months, but this type of mild to moderately intense abdominal pain occurred off and on for last 2-3 years. She had no history of convulsion, loss of consciousness or oily greasy stool. She was treated with anti-spasmodic and anti-ulcerant drugs. Her height for age lies between 10th-25th centile, she was severely underweight and malnourished (WAZ -3.6; BMI Z score -3.8), CBG-13.8 mmol/l and abdominal examination revealed mild tenderness in epigastric region.

She was diagnosed FCPD based on above clinical features and investigations. The patient achieved good glycaemic control with basal-bolus regimen of insulin. Oral tiemonium methylsulphate and omeprazole alleviated her abdominal pain. She was also treated with pancreatic enzyme tablets with meals along with vitamins and calcium supplementation. She and her family were adequately counselled on the features of both hypo and hyperglycaemia, regarding diet and self-blood glucose monitoring. She was discharged in a stable condition.

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Table-I
Investigation reports

Tests	Result
CBC	Hb-10.9gm/dl RBC-3600/cumm WBC-7000/cumm N- 68%, L- 28%, M -3% Platelet -250000/cumm ESR- 20 mm in 1 st hr Hct-34%
Urine R/M/E	Sugar ++++ , Protein- nil, Ketone body - nil, Pus cell- 0-2/HPF, RBC-nil, Specific gravity-1.020
HbA1c	11.9%
RBS	14 mmol/L
Serum Lipase	68 U/L
Serum Amylase	125 U/L
Serum Calcium	9.3 mg/dl
Serum ALT	40 U/L
Serum Creatinine	0.6 mg/dL
X-ray of abdomen in erect posture	Pancreatic calculi
USG of whole abdomen with special attention to pancreas &HBS	Heterogeneous echotexture of pancreas. Pancreatic parenchyma is atrophied, multiple diffuse calcified bright echogenic structures noted in head, body and tail of pancrease. MPD is mildly dilated. Suggestive of chronic calcific pancreatitis with mild dilated main pancreatic duct (MPD).

Discussion:

The diagnosis of FCPD is clinical and based on the criteria listed below table ⁹

Table-II
Diagnostic criteria for FCPD

1. Patient should be from a tropical country.
2. Diabetes by WHO criteria should be present.
3. Evidence of chronic pancreatitis should be present- demonstration of pancreatic calculi on abdominal imaging or at least three of the following: 4. a. Abnormal pancreatic morphology on ultrasound abdomen or CT scan b. History of recurrent abdominal pain since childhood c. Steatorrhea d. Abnormal pancreatic function tests
5. Absence of other causes of chronic pancreatitis (alcohol abuse, hepatobiliary disease or primary hyperparathyroidism etc.)

Our patient fulfilled the criteria of FCPD. Triad of abdominal pain, steatorrhea and diabetes are common in FCPD. But in the natural history of FCPD, each of these symptoms usually predominates at different stages. In in many cases of India, steatorrhea may not be prominent as the diet is poor in fat like our

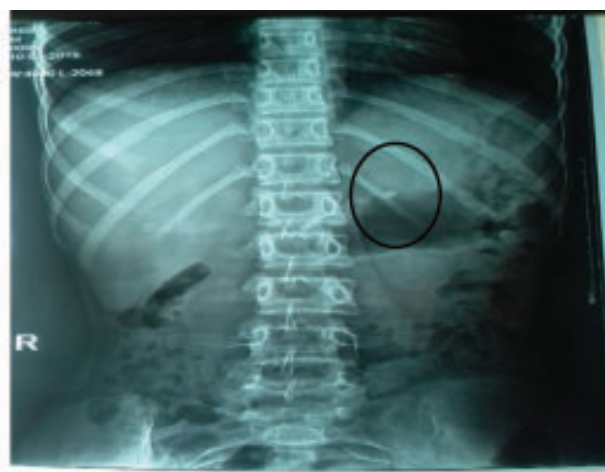


Fig. 2: Plain X-ray of abdomen showed pancreatic calculi

patient. Diabetes mellitus is developed between the ages of 10 - 40 years (15 years in our case) in the most of FCPD patients, usually.⁹ Absence of positive family history of diabetes is found in patients with FCPD usually, but reports of familial clustering has been observed.¹⁰ There is a marked male preponderance, but in a Bangladeshi study revealed that 81% of their FCPD patient were girl and 68% were from rural areas like our case.¹¹ The investigation of choice for the diagnosis is a plain X-ray of the abdomen, which will reveal the presence of the characteristic pancreatic calculi crossing the midline at the level of calculi are mostly situated to the right of the first or second lumbar vertebrae, but may rarely overlap the spine which are usually large in size and extend throughout the pancreas. Ultrasound examination of the abdomen is also useful and will pick up pancreatic duct dilatation in addition to calcification and calculi. Characteristics findings of both imaging were found in our case. Other imaging studies such as ERCP and CT or MRI scan are not indicated in routine practice.¹²

While the management of diabetes in this condition is challenging, a holistic approach including alleviation of symptoms and achievement of the best possible glycemic control so as to prevent the development of chronic diabetes complications.⁷ A high index of suspicion is necessary in early diagnosis and treatment of FCPD. Patients with FCPD should achieve better glycemic control with regular monitoring, pain alleviation, nutritional support and pancreatic enzyme supplements for improving the quality of life.

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