

Images of the month 1: Fibrocalculous pancreatic diabetes (FCPD): a rare form of secondary diabetes

Authors: Matiullah Kamin,^A Sajjad A Khan^B and Tejhmah Rehman^C

KEYWORDS: fibrocalculous pancreatic diabetes, exocrine pancreatic insufficiency, tropical diabetes, FCPD, pancreatic calcifications

DOI: 10.7861/clinmed.2021-0041

Case presentation

A 33-year-old woman of Pakistani descent presented with a 3-month history of significant weight loss (over 10 kg), fatigue, anorexia and new diagnosis of diabetes mellitus. She gave a long history of post-prandial abdominal bloating and discomfort, and oily stools. There was no associated dyspepsia, nausea, vomiting, melaena or erratic bowel habits. There was no past personal or family history of note. There was no history of smoking or alcohol intake. On examination, the patient appeared emaciated with a body mass index of just 17 kg/m². There was no postural hypotension and no clinical evidence of jaundice, pallor, hyperpigmentation, lymphadenopathy or clubbing. Systemic examination including gastrointestinal system was unremarkable.

Investigations

Laboratory tests are shown in Table 1. Glycated haemoglobin (HbA_{1c}) reflected poor glycaemic control with low normal C-peptide suggestive of reduced insulin secretory capacity.

Contrast enhanced computed tomography (CT) of the abdomen revealed an atrophied pancreas with multiple large intraductal and parenchymal calcifications (Fig 1). Magnetic resonance cholangiopancreatography (MRCP) confirmed extensive calcifications and dilated pancreatic duct (PD) with multiple filling defects (Fig 2). Endoscopic retrograde cholangiopancreatography (ERCP) was performed at another centre. It proved challenging with difficult PD access necessitating pancreatic sphincterotomy.

Diagnosis

On the basis of clinical and radiological findings, a diagnosis of fibrocalculous pancreatic diabetes (FCPD) was made.

Authors: ^Aendocrine fellow, Shifa International Hospital, Islamabad, Pakistan; ^Bendocrine fellow, Agha Khan University Hospital, Karachi, Pakistan; ^Cconsultant in endocrinology and assistant professor of endocrinology, Shifa International Hospital, Islamabad, Pakistan

Table 1. Laboratory results

Test	Result	Normal range
Haemoglobin, g/dL	13.4	12.0–15.5
WCC, /μL	6,500	4,500–11,000
Platelets, /μL	297,000	150,000–450,000
HbA _{1c} , % (mmol/mol)	12.5 (113)	<5.6 (<38)
C-peptide, ng/mL	1.4	0.8–3.85
ESR, mm/hour	22	0–20
Bilirubin, mg/dL	0.4	0.2–1.2
ALT, IU/L	30	19–25
ALP, IU/L	186	44–147
Hepatitis B surface antigen	Negative	-
Anti-HCV antibody	Negative	-
Serum calcium, mg/dL	9.2	8.6–10.3
Coeliac screen, tTG-IgA	Negative	-

ALP = alkaline phosphatase; ALT = alanine aminotransferase; ESR = erythrocyte sedimentation rate; HbA_{1c} = glycated haemoglobin; HCV = hepatitis C virus; tTG-IgA = tissue transglutaminase antibody – immunoglobulin A; WCC = white cell count.

Management

The patient was started on a basal bolus insulin regimen with comprehensive counselling. Dietician review was arranged to optimise nutritional intake. A faecal elastase test was not available. Pancreatic enzyme replacement was initiated for presumed diagnosis of exocrine pancreatic insufficiency (EPI). At 6-week review, she improved considerably with weight gain of 3 kg. Her glycaemic control improved, and she reported reduced gastrointestinal symptoms. Periodic follow-up was arranged with radiological surveillance.

Discussion

FCPD is rare and unique form of diabetes characterised by chronic calcifications of pancreas in the absence of alcoholism. It is mainly reported in tropical countries with majority in patient age group of 10 to 40 years. Aetiopathogenesis remains unclear. Multiple factors including malnutrition, familial aggregation,

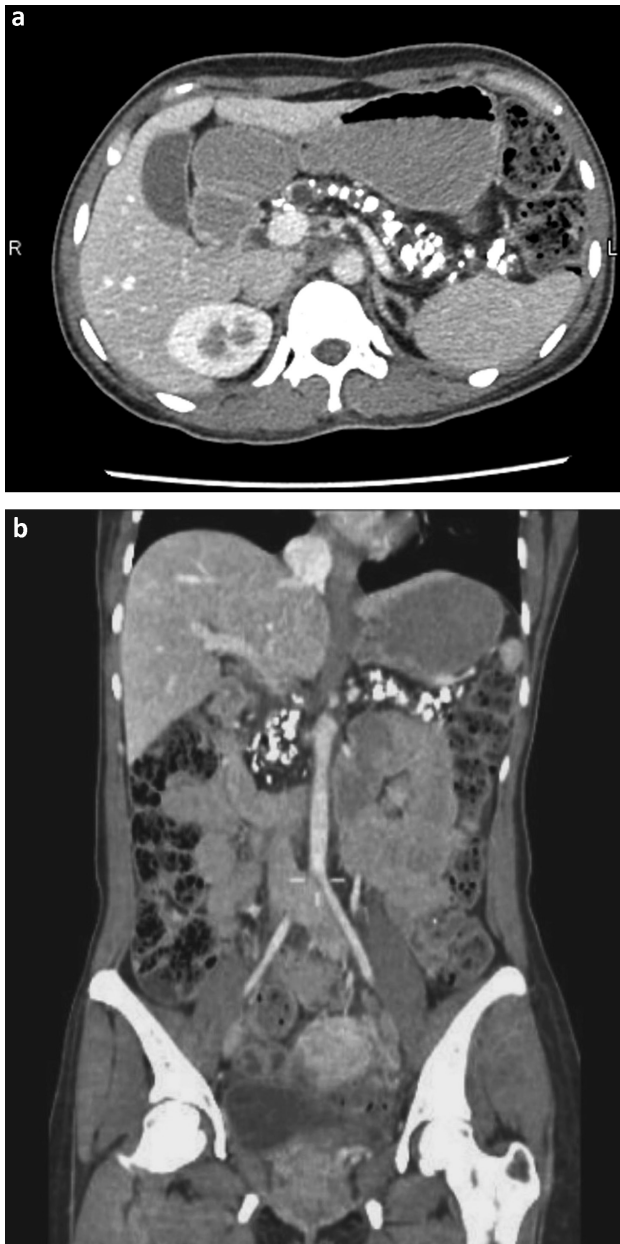


Fig 1. Contrast enhanced computed tomography of the abdomen revealed atrophied pancreas with multiple large intraductal and parenchymal calcifications. a) Axial plane. b) Coronal plane.

genetic factors, and vitamin A and C deficiencies are implicated.¹ Classical features include chronic abdominal pain, steatorrhoea, abnormal pancreatic morphology and calcification on imaging, with no history of alcoholism or hepatobiliary disease in someone of tropical origin. Pancreatic calcifications are radiological hallmark of this condition and can be seen on abdominal X-ray, ultrasound and computed tomography; the latter also illustrates extent of calculi in addition to identifying ductal dilatation and features of atrophy.^{2,3}

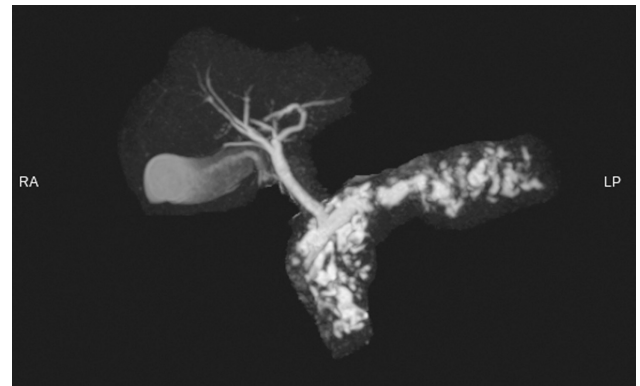


Fig 2. Magnetic resonance cholangiopancreatography confirmed extensive calcifications and dilated pancreatic duct with multiple filling defects.

Although diabetes is brittle and almost always requires insulin therapy, risk of ketosis is minimal, probably due to residual beta cell function and possible reduced non-esterified fatty acid (NEFA) availability due to subcutaneous fat loss.⁴ Holistic approach to management including controlling diabetes, addressing nutritional deficiencies and EPI, and avoidance of alcohol and smoking is essential for best outcome. FCPD is associated with increased risk of pancreatic malignancy and therefore warrants periodic radiological assessment.³

With increased global migration, physicians practising in non-tropics should be aware of and consider this rare form of diabetes in patients with suggestive features who originate from tropical countries.⁵ ■

References

- 1 Praveen G, Mohan V. Fibrocalculous pancreatic diabetes – current scenario in developing countries. *Int J Diabetes Dev Ctries* 2018;38: 131–2.
- 2 Dasgupta R, Naik D, Thomas N. Emerging concepts in the pathogenesis of diabetes in fibrocalculous pancreatic diabetes. *J Diabetes* 2015;7:754–61.
- 3 Bhat JA, Bhat MH, Misgar RA *et al*. The clinical spectrum of fibrocalculous pancreatic diabetes in Kashmir valley and comparative study of the clinical profile of fibrocalculous pancreatic diabetes and type 2 diabetes mellitus. *Indian J Endocrinol Metab* 2019;23:580–4.
- 4 Unnikrishnan R, Mohan V. Fibrocalculous pancreatic diabetes (FCPD). *Acta Diabetol* 2015;52:1–9.
- 5 Chong M, Hatley W, Hitman G. Fibrocalculous pancreatic diabetes detected in a British diabetic centre. *Prac Diab* 1990;7:82–3.

Address for correspondence: Dr Tejmal Rehman, Department of Endocrinology and Diabetes, Shifa International Hospital, Islamabad, 4 Pitrus Bukhari Road, H-8/4 Islamabad, Pakistan.
Email: tejmal@yahoo.com
Twitter: @tejmal