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

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**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, melena 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 (HbA₁c) 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.

**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|
| HbA₁c, % (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 | -         |

**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. Aetiology remains unclear. Multiple factors including malnutrition, familial aggregation,
genetic factors, and vitamin A and C deficiencies are implicated. 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.  

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. 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.  

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.

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