Case Report on Fibrocalculous Pancreatic Diabetes in a Tertiary Hospital in Cumilla, Bangladesh

Majumder MI¹, Mahadi AR², Ahmed M³, Uddin MN⁴, Shihab HM⁵, Billal ASMS⁶

Abstract

Background: This case highlights the emergence of diabetes as a significant global health concern and discusses Fibrocalculous Pancreatic Diabetes, a distinct condition characterized by pancreatic calcification and chronic inflammation resulting in pancreatic dysfunction. While typically seen in young males in tropical regions with malnutrition, this case presents a rare instance of late-onset disease in a female, a reported case in Bangladesh. The case involves a 53-year-old non-alcoholic healthy female with a history of chronic pancreatitis leading to severe Insulin Dependent Diabetes Mellitus. Abdominal imaging revealed typical pancreatic calcifications consistent with Fibrocalculous pancreatic diabetes. This report underscores the importance of maintaining a high index of suspicion for atypical presentations, even when diagnostic criteria suggest childhood onset of the disease.

Keywords: Pancreatitis, Calcification, Hospital, case report

Introduction: Fibrocalculous pancreatic diabetes mellitus (FCDP) is a distinct type of diabetes mellitus that occurs due to chronic calcification of the pancreas in young, non-alcoholic individuals, primarily found in tropical regions ¹. The hallmark features of FCDP include the onset of abdominal pain during childhood and the presence of pancreatic calculi, which are associated with dilation of the pancreatic duct and fibrosis of the gland during adolescence ². Over the years, various terms have been used to describe this form of diabetes, such as Pancreatic Diabetes, Pancreatogenous Diabetes, and Tropical Pancreatic Diabetes. However, the World Health Organization (WHO) Study Group Report on Diabetes has recently adopted the term Fibrocalculous Pancreatic Diabetes to specifically describe this condition ²,³.

In this case report, we present the case of a woman with non-alcoholic chronic pancreatic calcification, highlighting the unique characteristics and clinical challenges of managing FCDP. This report aims to contribute to the understanding of FCDP by detailing the patient’s symptoms, diagnostic process, and management strategies in a tertiary hospital setting in Cumilla, Bangladesh.

Case Presentation

A 53-year-old Bangladeshi woman, with no prior significant medical history, was referred to the Internal Medicine department at Central Medical College Hospital. The patient presented with a nine-year history of recurrent epigastric discomfort, initially misdiagnosed and treated as upper symptoms. The abdominal pain was characterized as intermittent and burning in nature, with radiation to the back. Additionally, the patient reported concomitant steatorrhea and subjective weight loss. There was no reported history of alcohol or cassava consumption.

References

1. Prof. Dr. Md. Mahabubul Islam Majumder, Professor & Head, Department of Medicine, Central Medical College, Cumilla, Bangladesh.
2. Dr. Ashrafur Rahaman Mahadi, Indoor Medical Officer, Department of Medicine, Central Medical College, Cumilla, Bangladesh.
3. Dr. Mostaque Ahmed, Associate Professor, Department of Medicine, Central Medical College, Cumilla, Bangladesh.
4. Dr. Mohammad Nazim Uddin, Assistant Professor, Department of Medicine, Central Medical College, Cumilla, Bangladesh.
5. Dr. Hossain Mohammed Shihab, Registrar, Department of Medicine, Central Medical College, Cumilla, Bangladesh.
6. Dr. Abu Sayed Md Shahed Billal, Registrar, Department of Medicine, Central Medical College, Cumilla, Bangladesh.

Correspondence: Prof. Dr. Md. Mahabubul Islam Majumder, Professor & Head, Department of Medicine, Central Medical College, Cumilla, Bangladesh. Mobile: 01713-459545, E-mail: m.i.majumder@gmail.com

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gallstone disease, or hepatitis. Furthermore, the patient’s family history was negative for pancreatic disease or diabetes mellitus.

Fig 1: The abdominal X-ray reveals widespread pancreatic calcification affecting the head, body, and tail of the pancreas, with a notable concentration to the right of the twelfth thoracic and first lumbar vertebrae.

The patient’s initial blood glucose levels were within normal limits; however, during routine follow-up assessments, she was found to have poor glycemic control, as evidenced by an elevated fasting plasma glucose level of 122 mg/dL and a significantly high HbA1c of 13.8%. Initially managed with oral hypoglycemic agents, she was transitioned to biphasic insulin therapy four years prior due to inadequate glycemic control. Her current insulin regimen consists of 16 units in the morning and 10 units in the evening.

Despite the persistent poor glycemic control, the patient has not experienced any episodes of ketoacidosis. Moreover, she does not report any osmotic symptoms, and there is no clinical evidence of macrovascular or microvascular complications commonly associated with diabetes. However, it is noteworthy that she has not yet been evaluated by nephrology or ophthalmology specialists to definitively rule out such complications.

The patient has been receiving ongoing management for chronic pancreatitis. Her treatment regimen includes medical therapy with 30,000 IU of pancreatic enzyme supplements daily, along with other essential micronutrients. Additionally, she has undergone surgical intervention for her condition.

Fig 2: Magnified view of the X-ray abdomen showing generalized pancreatic calcification

The patient has been undergoing annual Endoscopic Retrograde Cholangiopancreatography (ERCP) procedures combined with stent placement. This recurring interventional approach has proven to be highly effective in mitigating her chronic abdominal pain, providing significant symptomatic relief.

Upon physical examination, the patient presented as emaciated, with a Body Mass Index (BMI) of 19.2 kg/m², indicating underweight status. She exhibited mild pallor without icterus, and there were no peripheral stigmata indicative of chronic liver disease or any clinical features suggestive of hemochromatosis.

Radiographic imaging, including abdominal X-rays (Fig1, Fig 2), revealed diffuse pancreatic calcification involving the head, body, and tail, consistent with chronic pancreatitis.
Ultrasonography (USS) and Computed Tomography (CT) scans of the abdomen further corroborated this diagnosis, demonstrating pancreatic atrophy and the pathognomonic “bag of stones” appearance.

Hematological investigations revealed hypochromic microcytic anemia, with a hemoglobin level of 8.5 g/dL. Iron studies confirmed iron deficiency anemia, with a transferrin saturation of 13% (normal range: 20-50%). Biochemical analysis showed marginally elevated alkaline phosphatase at 317 IU/L and hypoproteinemia. Thyroid function tests were within normal limits, with a TSH level of 4.43 µIU/mL. The lipid profile indicated low levels across parameters. Urinalysis demonstrated marginal microalbuminuria, with a urine albumin:creatinine ratio of 30.39.

Tumor marker Carbohydrate Antigen (CA19-9) was 13 U/mL, and the C-peptide level was at the lower end of the normal range at 0.5 nmol/L (normal range: 0.5-1.15). Based on this comprehensive clinical, radiological, and laboratory evaluation, a diagnosis of fibrocalculous pancreatic diabetes (FCDP) was established.

The patient’s treatment regimen focused on three key aspects: glycemic control, pain management, and maintenance of both exocrine and endocrine pancreatic functions. Upon follow-up assessment, the patient demonstrated clinical improvement, characterized by a modest increase in BMI to 19.9 kg/m², achieved good glycemic control, and is currently experiencing pain-free status.

Discussion
Fibrocalculous pancreatic diabetes (FCPD) is a distinct type of diabetes that occurs due to pancreatic calcification in individuals who do not consume alcohol, typically affecting young people 1. Most FCPD patients are diagnosed with diabetes between ages 10 and 40, and many have a history of severe, recurring epigastric pain 4. While cases of FCPD beginning in childhood have been reported in Africa and Asia, this particular patient’s case is unusual. They developed FCPD symptoms at age 49, which is considered late-onset, and had no history of such symptoms during their youth. A previous case report shows two cases of FCPD in elderly which is very rare similar to this case 5.

Several factors contribute to the development of fibrocalculous pancreatic diabetes (FCPD). These include genetic predisposition, poor nutrition, and the harmful effects of cyanide, which is commonly ingested through frequent consumption of cassava. Additionally, deficiencies in vitamins C and A lead to increased oxidative stress in the body, further contributing to the disease’s onset 6.

The three key features of fibrocalculous pancreatic diabetes (FCPD) are abdominal pain, steatorrhea, and diabetes. Abdominal pain is the most common symptom, characterized by severe, radiating discomfort in the epigastric region, with periods of improvement and worsening 7. About one-third of patients report passing large, oily stools, although our patient’s stool test showed no fat globules, likely due to a low-fat, high-carbohydrate diet 8.

Pancreatic calculi are often visible on abdominal X-rays. Ultrasound and CT scans help locate these stones and show other signs like atrophy, fibrosis, duct dilation, and in extreme cases, a ‘bag of stones’ appearance 8.

Despite high blood sugar levels, ketosis is usually absent, possibly due to some remaining beta-cell function, as indicated by moderate C-peptide levels. Most patients need insulin to manage their high blood sugar 9.

In patients who have negligible C-peptide, it was proposed by Yajnik that the mechanisms other than beta-cell function are also involved 8. The key factors are as follows:

1. Residual B-cell function adequate to prevent ketosis
2. Concomitant destruction of A-cells and thus loss of glucagon, a major ketogenic hormone
3. Subcutaneous fat loss and therefore reduced supply of non-esterified fatty acids (NEFAs) – the ‘fuel’ for ketogenesis
4. The resistance of subcutaneous adipose tissue lipolysis to adrenaline
5. Carnitine deficiency affecting the transfer of NEFA across the mitochondrial membrane.
Diagnostic criteria for FCPD [10]:

1. Occurrence in a ‘tropical’ country
2. Diabetes by the WHO study group criteria [11]
3. Evidence of chronic pancreatitis: pancreatic calculi on X-ray or at least three of the following:
   a. Abnormal pancreatic morphology by USG
   b. Chronic abdominal pain since childhood
   c. Steatorrhoea
   d. Abnormal pancreatic function test.
4. The absence of other causes of chronic pancreatitis i.e., alcoholism, hepatobiliary disease or primary hyperparathyroidism, etc.

In this patient’s case, the diagnosis of fibrocalculous pancreatic diabetes (FCPD) was made based on recurring abdominal pain, diabetes without ketosis, and evidence of pancreatic calculi seen on ultrasound and x-ray scans.

FCPD patients may develop small blood vessel (microvascular) complications such as retinopathy, neuropathy, and nephropathy. Large blood vessel (macrovascular) issues are less common, likely due to their young age, lean body type, and low cholesterol levels 12-15.

However, one case reported severe peripheral vascular disease with gangrene without other risk factors for atherosclerosis, suggesting the need for thorough foot examinations at least annually. A study of 30 FCPD patients found cardiac autonomic neuropathy in 63.3%, indicating a need for frequent autonomic dysfunction screenings 16.

Although rare, a high level of suspicion is needed when managing diabetes. The main treatment involves good blood sugar control, pancreatic enzyme supplements for malabsorption, and nutritional support to prevent complications. Misdiagnosed or delayed diagnoses can lead to life-threatening issues like pancreatic cancer, so regular screenings are also essential.

Conclusion

While fibrocalculous pancreatic diabetes (FCPD) is typically seen in young people, this rare case demonstrates that it can also affect middle-aged individuals, even without a history of abdominal pain in childhood. This underscores the importance of maintaining a high level of suspicion for FCPD to ensure early diagnosis and prompt treatment.

To enhance the quality of life for FCPD patients, several key factors are crucial: achieving and maintaining good blood sugar control with regular monitoring, providing effective pain relief, addressing deficiencies in both macro and micronutrients, and managing pancreatic failure. These comprehensive care strategies are essential in improving outcomes and well-being for those living with FCPD.

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Conflict of interest statement

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Data availability statement

All the required information is available in the manuscript itself.

Ethics statement

The Institutional Review Board of Central Medical College in Cumilla, Bangladesh, approved this study. A formal letter was provided as clarification for conducting this Case review. For publishing of the clinical details and photographs in this article, signed informed consent from the Patient was acquired.

Consent

Written informed consent was obtained from the patient To publish this report in accordance with the journal’s patient consent policy.

References


