SPECIAL ARTICLE

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Fibrocalculous Pancreatic Diabetes (FCPD) : A Rare Type of Pancreatogenic Diabetes

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ABSTRACT

Background. FCPD is a rare form of secondary diabetes. FCPD is mainly found in the tropical region; it is characterized by diabetes, chronic abdominal pain, calcification of the pancreas, and steatorrhea. The Incidence of FCPD is often misdiagnosed with type 2 or type 1 diabetes mellitus.

Case Illustration. A 46-year-old man came with chief complaints of chronic abdominal pain. Abdominal radiography showed calcification in the pancreas. The patient was malnourished. Abdominal X-ray revealed pancreatic calcification, which was confirmed by an abdominal MSCT scan. C-Peptide decreased with an intermediate degree. The patient was given supportive therapy, and insulin was given to control his diabetes.

Discussion. FCPD is a morphological pancreas change caused by chronic tropical pancreatitis. The aetiology of chronic tropical pancreatitis is unknown. FCPD can be diagnosed by history taking and supporting examinations such as abdominal X-ray examination, ultrasound and abdominal CT Scan. C-peptide was examined to assess the function of pancreatic beta cells. The primary treatment for FCPD is insulin therapy; metformin or Sulfonylureas can be used in the early phase of diabetes. In reducing pain, non-steroidal analgesics are used as an option. The use of pancreatic enzyme supplementation can improve the nutritional status of patients.

Conclusion. FCPD is a rare case, occurring mainly in tropical countries and in people who are malnourished. The primary treatment for FCPD is insulin therapy.

Key words : Fibrocalculous Pancreatic Diabetes, Diabetes Mellitus, Chronic Pancreatitis

INTRODUCTION

The highest prevalence of diabetes is type 2 diabetes mellitus and type 1 diabetes mellitus. About 1-5% of diabetes is caused by other causes, such as endocrinopathy and pancreatic disorders.[1,2] Fibrocalculous Pancreatic Diabetes (FCPD) is common in countries with tropical climates. The study conducted by Geeverghese is the first to describe cases of FCPD on a large scale in the Indian state of Kerala, with a prevalence of 29.3% among total diabetes registered in the medical college in 1960. However, the prevalence decreased drastically, with a prevalence of 0.36% in 2001-2003 and 0.2% in 2001 - 2006.[3] There are no studies in Indonesia that explain the prevalence of FCPD.

The cause of FCPD is unknown. Several hypotheses regarding its aetiology exist, including the malnutrition hypothesis, the casava hypothesis, familial and genetic factors, deficiency of essential elements, oxidative stress, and autoimmunity.[4–6] FCPD is an often overlooked diagnosis. Most of the patients with FCPD were diagnosed as type 1 or type 2 diabetes mellitus before the exact diagnosis was known. FCPD has a variety of symptoms, and the disease is not widely recognized by medical personnel.

CASE ILLUSTRATION

A 46-year-old man came to Dr Moewardi Hospital with chief complaints of left abdominal pain 2 weeks ago. Abdominal pain was intermittent and accompanied by nausea and vomiting. He had lost 12 kilograms of weight in the last 2 months. One month earlier, he had been hospitalized in a regional hospital, and he was diagnosed with chronic pancreatitis and type 2 diabetes mellitus. He received oral therapy with metformin to control his blood sugar.

The average blood pressure during the physical examination was above normal. The patient appeared malnourished with a BMI of 16.5 kg/m2. Daily fasting blood glucose monitoring was 98-130 mg/dl, while postprandial blood glucose examination was 160-180 mg/dl. Lipid profile, liver and kidney function within normal limit. C-peptide level decreased to 0.5 ng/ml. The abdominal ultrasound examination was normal. A plain photo examination of the abdomen showed multiple opacities in the left hypochondriac region to the epigastrium as high as corpus vertebrae thoracal 12 mid clavicula sinistra to corpus vertebrae lumbal para vertebrae dextra, which leads to pancreatic calcification (Figure 1). The patient's abdominal computer tomography (CT) scan supported the diagnosis of chronic pancreatitis with multiple calcifications in the pancreas (Figure 2). Based on the criteria, this patient was diagnosed with FCPD.

The patient was educated on medical nutrition therapy. The patient's blood glucose monitoring was still above average, so it was decided to use basal-bolus insulin to control the patient's blood sugar. He also received pancreatic enzyme supplementation to prevent absorption disorders in the patient. NSAIDs were given to reduce abdominal pain experienced by the patient. After 2 months of evaluation, HbA1c level decreased to 7.0%, fasting blood sugar 119 mg/dl and postprandial blood sugar 149 mg/dl. The patient's symptoms have decreased, but he lost follow-up and returned to his home area for economic reasons.

DISCUSSION

FCPD is a form of diabetes that is rarely encountered in clinical practice. FCPD is generally found at a young age. In men, the diagnosis of FCPD occurs in the range of 10 years to 40 years old. Whereas in women, the average diagnosis occurs at the age of 17 years old.[7] Most patients with FCPD are malnourished and typically occur in tropical countries. The classic symptoms of FCPD are diabetic symptoms accompanied by chronic abdominal pain, stones or calculi in the pancreas, and the emergence of steatorrhoea.[2,8] The diagnostic criteria for FCPD include : [1,9]

1. It happens in developing countries
2. Diabetes diagnosis based on WHO Study group criteria
3. There is evidence of chronic pancreatitis, which includes clinical and radiological examinations that include the following three criteria:
   a. Abnormal pancreatic morphology according to radiological examination
   b. Chronic abdominal pain
   c. Abnormal pancreatic function
   d. Steatorrhea

4. There were no other causes of chronic pancreatitis, such as alcoholism, hepatobiliary disorders, and primary hyperthyroidism

Abdominal pain is the main symptom that often occurs since childhood. Abdominal pain is usually episodic and improves with sitting and bending. However, the frequency of abdominal pain that occurs gradually decreases with the development of diabetes. In our case, the criteria for the diagnosis of FCPD have also been met with the incident taking place in a tropical country (Indonesia), the diagnosis of diabetes according to the WHO study group criteria, evidence of chronic pancreatitis through clinical and laboratory studies, and no other causes of chronic pancreatitis that occur such as alcohol use and hepatobiliary disease.[10]

The physical examination often found in patients with FCPD is known as the classic triad of FCPD, including parotid gland enlargement, abdominal distension, and cyanosis of the lips. However, the physical examination results with the triad are rarely found to have more heterogeneous properties. Malnutrition with a BMI below the average value is often seen in FCPD patients.[11] Examination of the abdomen shows various symptoms; it can be normal, or there is pain when pressed in the epigastric region and right hypochondriac.

Radiological examination plays a vital role in diagnosis. Some radiological examinations used are plain abdominal radiographs, ultrasound, CT scans, endoscopic retrograde cholangiopancreatography (ERCP), endoscopic ultrasound, and magnetic cholangiopancreatography. Ultrasound shows dilatation of the pancreatic duct and increased echogenicity secondary to fibrosis of the gland. Abdomen X-ray examination showed pancreatic calcification.[1,12,13] CT scan is an ideal examination to diagnose chronic pancreatitis.

Increased blood sugar levels, characterized by increased levels of HbA1c and an increase in postprandial blood glucose, are caused by a decreasing number of residual pancreatic beta-cells.[8] The examination used to assess pancreatic beta-cell function is done using a C-peptide assay. C-Peptide levels in FCPD patients tend to decrease to an intermediate degree.[4]

Non-opioid analgesics are used to reduce the pain experienced. The primary choice in the management of FCPD is the administration of insulin. Insulin is given in a combination of rapid-acting and long-acting insulin, adjusted to the patient's needs. [1,9] The diabetic oral drug can be given, especially in the early phase of diabetes. A long-acting insulin secretagogue is not recommended because it increases the risk of hypoglycemia. Metformin can be given but needs to be considered for gastrointestinal side effects. Incretin-based drugs are not recommended. Only a few literatures have discussed the effect of thiazolidinediones and SGLT-2 administration in FCPD patients. Giving patients pancreatic enzyme supplementation can improve their nutritional status.[1,14]

CONCLUSION

FCPD is a rare case, often occurring in young adults, characterized by malnutrition, diabetes and chronic pancreatitis. Initial management can be given oral antidiabetic drugs, but at an advanced stage, insulin is used to control blood glucose. Adequate glycemic control, reduced pain, and improved pancreatic function are the main treatments that improve the patient's quality of life.

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AFFILIATIONS

All authors are staff of the Endocrine, Metabolic and Diabetes Division of Dr. Moewardi Hospital Surakarta / Faculty of Medicine, Sebelas Maret University.

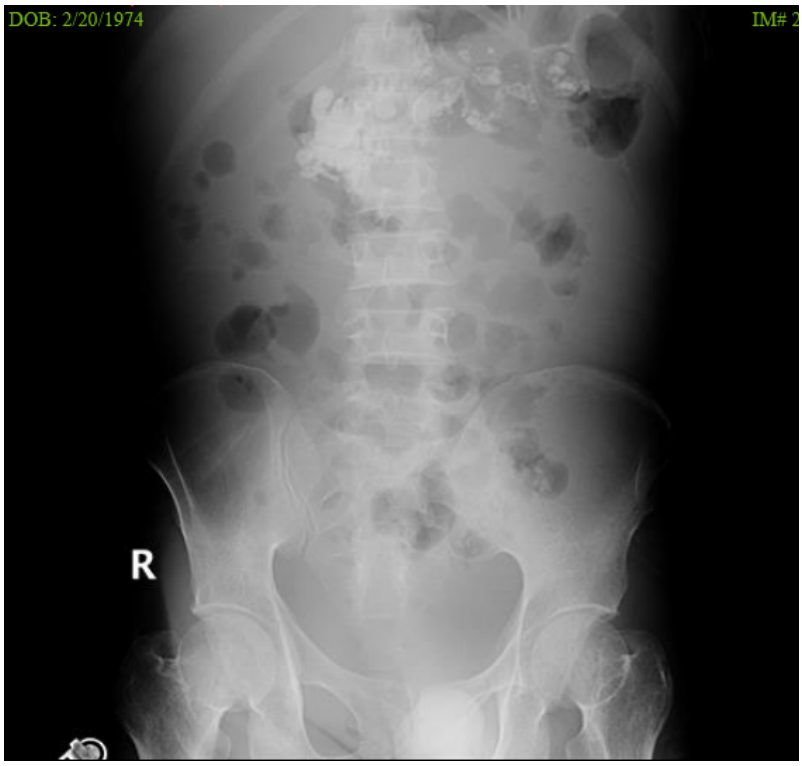

Figure 1. Calcification of pancreas appear in abdominal X-ray

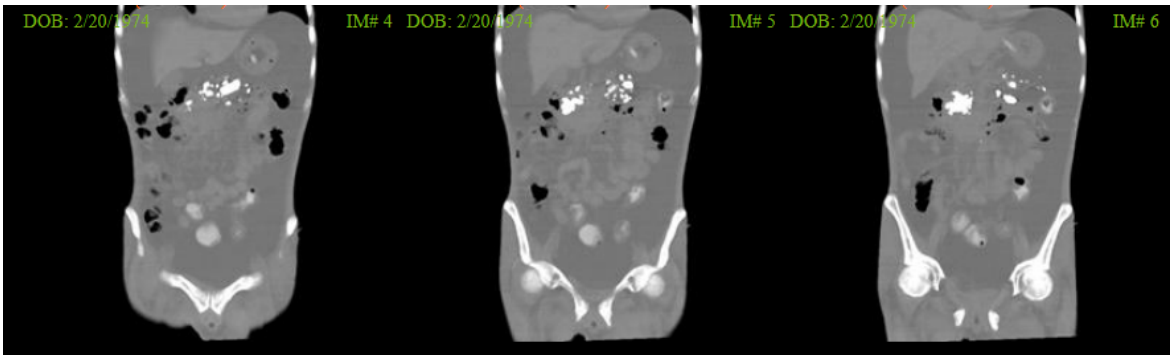

Figure 2. CT Scan Shows Multiple Calcification of pancreas

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