

Case report

Fibrocalculous pancreatic diabetes in a patient residing in the Mediterranean region

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ABSTRACT

A 40-year old male residing in the Mediterranean region and afflicted with chronic pancreatitis and diabetes is presented. This is a case of chronic calcific non-alcoholic pancreatitis with characteristic intraductal calculi on abdominal x-ray. Five years subsequent to the first episode of pancreatitis, developed insulin-requiring diabetes mellitus. This case accords with the criteria set by Mohan et al for fibrocalculous pancreatic diabetes with the unique feature of the patient having been born in Greece and being a resident of Greece.

Key words: Diabetes mellitus, Fibrocalculous pancreatic diabetes

INTRODUCTION

Diabetes mellitus is a group of metabolic diseases characterized by hyperglycemia resulting from defects in insulin secretion, insulin action or both. Assigning a type of diabetes to an individual often depends on the circumstances present at the time of diagnosis, and many diabetic individuals do not easily fit into a single class. The American Diabetes Association in its recent annual report¹ presented the etiologic classification of diabetes mellitus. In the category IIIC under the title "diseases of the exocrine pancreas" fibrocalculous pancreatopathy is included. In this case presentation, we report on a Caucasian man who conforms to the criteria for fibrocalculous pancreatopathy.

CASE REPORT

This is a case of a 40-year old Greek male of Caucasian ancestry on both sides, who has been living in Greece. The patient, who has a 10-year history of insulin dependent diabetes, was seen at the diabetes clinic. He mentioned episodes of intermittent epigastric pain, radiating to the back and relieved by leaning forward or lying in a prone position. No history of gallstones, gastrointestinal bleeding, alcohol intake, cassava consumption or hepatitis was reported. Diabetes mellitus was detected for the first time five years after the initial episode of abdominal pain. The onset was gradual, the main symptoms being polyuria, polydipsia and steatorrhea. There was no family history of diabetes, past medical history of endocrinopathy or use of potentially diabetes inducing medications. On detection of diabetes mellitus, he was lean (BMI 19.3 kg/m²) undernourished weak and suffering from muscle cramps that forced him to seek medical attention.

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Received 08-09-05, Revised 28-11-05, Accepted 00-00-05

His glycosylated hemoglobin (HbA1c) at that time was 10.2%. He was started on insulin soon after the diagnosis of diabetes mellitus and his daily insulin requirements were 40 ± 8 units. The patient recalled one episode of mild ketosis after temporary withdrawal of insulin, during a severe respiratory infection. During his visit to our clinic, the physical examination was unremarkable except for his leanness (BMI 19 kg/m^2) and a few microaneurysms on fundoscopic examination. There was no evidence of clinical neuropathy or microalbuminuria. His routine blood tests were normal including hematology, thyroid function tests, liver function tests and serum calcium. Anti-TPO antibodies were negative. Markers of immune destruction of the beta cells (Insulin, Glutamic acid decarboxylase, and tyrosine phosphatase IA-2 antibodies) were negative. On abdominal x-ray, multiple large, rounded, dense calculi were detected along the pancreatic duct (Figure 1). Ultrasonography of his biliary tract did not show any stones. Ultrasonography of the pancreas confirmed dilated pancreatic ducts with intraductal calculi. Fasting C-peptide was 0.4 nmol/l (normal: $0.5\text{--}1.15 \text{ nmol/l}$) and fecal chymotrypsin was 3 units/g of fecal mass (normal: $>5.8 \text{ units/g}$ of fecal mass).^{2,3} Based on putative of fibrocalculous pancreatic diabetes, the treatment was mainly directed at the control of hyperglycemia and prevention of complica-



Figure 1. Plain radiograph of abdomen showing evidence of pancreatic calculi.

tions. He was instructed to follow a 2500-calorie, low-fat diet supplemented by pancreatic enzymes (30000 IU of pancreatic lipase with meals). On follow-up he had gained weight (BMI = 22), and there was significant improvement in his quality of life. His glycosylated hemoglobin was 8%.

DISCUSSION

Fibrocalculous pancreatic diabetes (FCPD) is a unique form of diabetes secondary to chronic calcific non-alcoholic pancreatitis and is observed almost exclusively in the developing countries of the tropical world. The classical triad consists of abdominal pain, steatorrhea and diabetes. FCPD is associated with overt protein-calorie malnutrition. FCPD affects young individuals who show characteristic large intraductal pancreatic calculi on abdominal x-ray.^{4,7} This form of pancreatitis is distinguished histologically from the other forms of chronic pancreatitis. Despite excellent clinical descriptions of the disease, up until recently no criteria have been established for the diagnosis of FCPD. Mohan and al were the first to propose a set of criteria⁶ and other workers in the field have generally accepted these criteria.⁸⁻¹⁰ The criteria for fibrocalculous pancreatic diabetes according to Mohan include: i) the patient should originate from a tropical country, ii) diabetes should be present, iii) evidence of chronic pancreatitis must be present (pancreatic calculi on abdominal X-ray or at least three of the following: a) abnormal pancreatic morphology on sonography/CT scan, b) recurrent abdominal pain since childhood, c) steatorrhoea, d) abnormal pancreatic function test), iv) absence of other causes of chronic pancreatitis. FCPD is now classified under the broad category of diseases of the exocrine pancreas, both by the American Diabetes Association and by WHO. Our patient, although not residing in a tropical country, fulfilled Mohan's criteria for FCPD.⁶ He had diabetes with partially preserved beta cell function, chronic calcific non-alcoholic pancreatitis with characteristic intraductal calculi on x-ray and absence of other causes of chronic pancreatitis. The occurrence of FCPD in a patient not residing in a tropical region is quite unique.

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