Case Report

Fibrocalculous pancreatic diabetes in adult

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Abstract

Fibrocalculous pancreatic diabetes has distinctive features like younger age at onset, presence of large intraductal calculi, aggressive course of the disease, and proneness for pancreatic cancer. Pancreatic calculi are the hallmark for the diagnosis. We report a 32-year-old male patient, a known case of diabetes since 2 years, presented with recurrent pain abdomen, malabsorption, and neuropathic symptoms. The diagnosis was established on the basis of clinical examination, biochemical and radiological investigations. He was prescribed two doses of premix insulin and pancreatic enzyme supplements for relief of abdominal pain and steatorrhea.

Key Words: Fibrocalculous pancreatic diabetes, insulin, pancreatic diabetes

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Received: 26.07.2013, **Accepted:** 06.04.2015

INTRODUCTION

Fibrocalculous pancreatic diabetes (FCPD) is seen mainly in young and malnourished individuals belonging to tropical and developing countries. It is associated with an increased risk of pancreatic carcinoma. The association of malnutrition and diabetes was first documented by Zuidema in 1959 from Indonesia.[1] Shaper in 1969 reported similar association from Uganda.[2] The first case of pancreatic calculi from India was reported by Kini in 1937[3] and this was followed by reports of pancreatic calculi observed at postmortem from Vellore in Southern India.[4] However, it was after documentation by Geevarghese one of the largest series in the world from Kerala in Southern India that FCPD attracted international attention.[5] In FCPD the pancreas undergoes extensive fibrosis, the ducts are dilated and characteristically show large intraductal calculi.[6]

Access this article online	
Quick Response Code:	Website: www.advbiores.net
	DOI: 10.4103/2277-9175.164008

CASE REPORTS

Case 1

A 32-year-old male patient came for the management of diabetes mellitus to the Diabetes Centre KLES Dr. Prabhakar Kore Hospital and Medical Research Centre, Belgaum, Karnataka, South India. He was diagnosed to have diabetes mellitus since 2 years. He had a history of missing insulin doses frequently for extended periods and was never hospitalized after the initial diagnosis. There was no documented episode of diabetic ketoacidosis. He presented with complaints of frequent hypoglycemia, tiredness, loss of bodyweight and appetite and neuropathic symptoms. There was no history hypertension, dyslipidemia, tuberculosis and any other medical illnesses such as alcoholic pancreatitis and hyperparathyroidism. His diet was irregular and erratic. He was nonsmoker and nonalcoholic.

How to cite this article: Jali MV, Kambar S, Hiremath MB, Wasedar J, Jali SM. Fibrocalculous pancreatic diabetes in adult. Adv Biomed Res 2015;4:184.

General physical examination showed body mass index (BMI) - 14.72, pulse rate (PR) - 80b/min, blood pressure (BP) - 140/100 mmHg, relative risk - 20 cyc/min. No pallor, icterus, clubbing, lymphadenopathy, edema, and thyromegaly. Systemic examination (respiratory, cardiovascular, and central nervous system) was within normal limits. Per abdomen, examination showed hepatomegaly.

The patient was hospitalized for further investigations and was started with premix insulin oral hypoglycemic agents (OHA) and pancreatic enzyme supplements. Patient tolerated the combination well. On the day admission fasting plasma glucose was 159 mg/dl and 2 h postprandial plasma glucose was 200 mg/dl and HbA1c - 6.7%. During hospitalization his blood sugars were monitored 5 times in a day (fasting blood sugar [FBS], postprandial blood sugar [PPBS], prelunch, predinner, and bedtime random blood sugar [RBS]) and there were no episodes of hypoglycemia. Laboratory investigations revealed that hemogram, liver function tests (LFT), kidney function tests, thyroid and lipid profile were within normal limits. Chest X-ray and electrocardiogram (ECG) findings were normal. Serum amylase (69 U/L) and serum lipase (142 U/L) were within normal limits. Fundus examination showed normal study.

By ultrasonography of the abdomen [Figure 1] it was possible to evaluate the size of the pancreas and also confirm the intraductal location of calculi and the degree of fibrous.

Further, the patient was evaluated with computed tomography (CT) scan of abdomen [Figure 2] it showed chronic calcific pancreatitis with dilated pancreatic duct with intramural calculi with hepatomegaly.

During hospital stay the patient improved clinically and glycemically. On discharge patient was advised to take (i) Two doses of premix insulin, (ii) OHA once a daily and (iii) pancreatic enzyme supplements twice daily. Nutrition counseling and diabetes education (insulin injection technique and self-monitoring of blood glucose) was given to the patient. Later, the patient was followed up at 1st month, 3rd month, and 6th month. He was regular in taking treatment and follow diet and exercise.

Case 2

A 43-year-old male patient came for the management of diabetes mellitus to the Diabetes Centre KLES Dr. Prabhakar Kore Hospital and Medical Research Centre, Belgaum, Karnataka, South India. He was diagnosed to have diabetes mellitus since 2 years. He presented with body pain, tiredness, and loss of bodyweight. There was no history hypertension, dyslipidemia, asthma, tuberculosis and any other medical illnesses such as alcoholic pancreatitis and hyperparathyroidism. His diet was irregular and erratic. He is a smoker and smokes 2–3 cigarettes a day. He has a history of having alcoholic drinks and beverages regularly, since 8 months he has stopped drinking.

General physical examination showed BMI - 20.56, PR - 80b/min, BP - 110/70 mmHg HbA1c - 15.4. Per abdomen, examination showed hepatomegaly.

Patient on further investigations and was started with premix insulin OHA and pancreatic enzyme supplements. The patient tolerated the combination well. On the day admission fasting plasma glucose was 339 mg/dl and 2 h postprandial plasma glucose was 503 mg/dl and HbA1c - 15.4%. After the following above administrations, his blood sugars were monitored 5 times in a day (FBS, PPBS, prelunch,

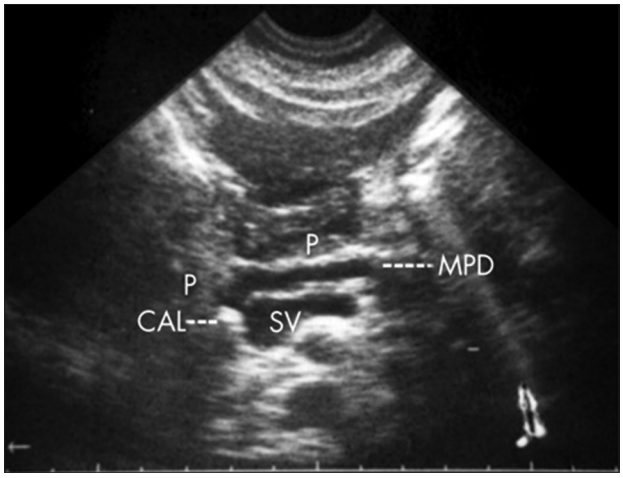

Figure 1: Ultrasonography abdomen

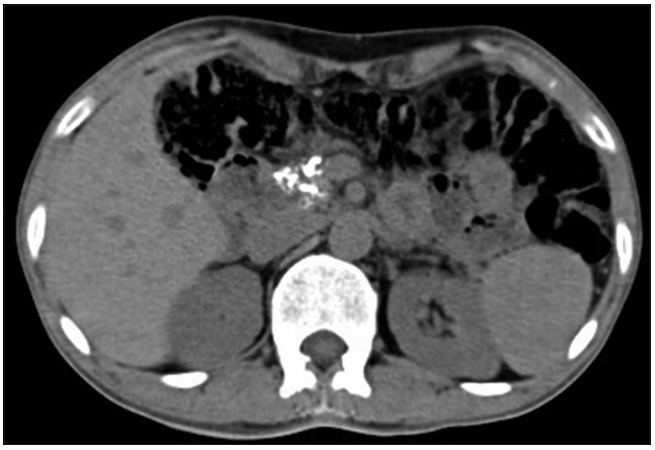

Figure 2: Computed tomography scan abdomen in case 1

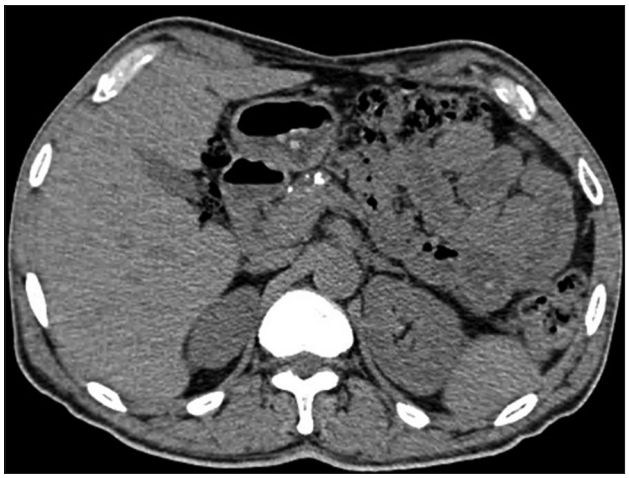

Figure 3: Computed tomography scan abdomen in case

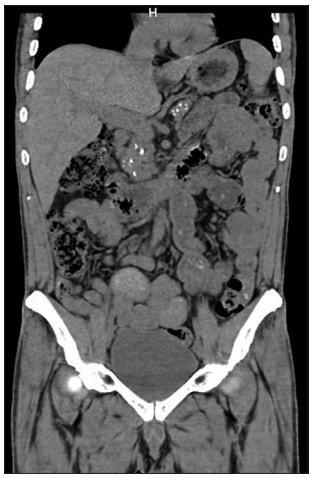

Figure 4: Computed tomography scan abdomen in case 2

predinner, and bedtime RBS) and there were no episodes of hypoglycemia. Laboratory investigations revealed that hemogram, LFT, kidney function tests, thyroid and lipid profile were within normal limits. Chest X-ray and ECG findings were normal.

Further, the patient was evaluated with CT scan of abdomen [Figures 3 and 4] it showed chronic calcific pancreatitis with dilated pancreatic duct with intramural calculi with hepatomegaly.

He is practicing storage of grains in the house with the enclosure of cow dung.

DISCUSSION

The classical symptoms of FCPD are abdominal pain and maldigestion leading to steatorrhea. It is usually diagnosed between the ages of 20 and 35 years. The exact pathogenetic mechanisms responsible for development of FCPD are not known; malnutrition, dietary toxins (e.g. cyanide present in cassava), genetic and immunological factors and lack of micronutrient antioxidants have all been proposed.[7,8]

The pathological process starts from the pancreatic head and subsequently extends to the body and tail regions. The pancreas in advanced FCPD is shrunken and on palpation appears as a bladder-like structure full of stones. Overt steatorrhea is reported in only up to 30% of patients.[8] However, on pancreatic function testing evidence of malabsorption is demonstrated in 90% of patients. Microvascular diabetic complications frequently occur in patients with FCPD.[9,10] The risk of developing pancreatic carcinoma is 100-fold higher in patients with FCPD than in the normal population.[10]

Most patients with FCPD require insulin for the control of symptoms and hyperglycemia. Generally, patients are resistant to ketoacidosis; the presence of some residual beta-cell function, low glucagon reserve, decreased body fat and carnitine deficiency are postulated to be the factors responsible for resistance to ketosis.[9,10] Patients with FCPD have sometimes been shown to be very sensitive to insulin due to low glucagon reserve and are prone to recurrent hypoglycemia.[11] Mohan *et al*. reported advanced retinopathy in FCPD patients.[12] Nephropathy was seen in 8.9% of our FCPD patients.[13] Renal failure due to diabetic nephropathy has also been reported in other forms of pancreatic diabetes.[14] Peripheral neuropathy[15] and autonomic neuropathy[16] have also been reported in those with FCPD. Macrovascular complications are, however, rare in FCPD. This is believed to be due to three reasons: The patients are young, lean and have low lipid levels.

Acknowledgment

The co-operation and support of the staff of Diabetes Centre, The Medical Records and Biostatistics Division, KLES Dr. Prabhakar Kore Hospital and MRC, Belgaum and KLES Belgaum Imaging and Diagnostics Pvt., Ltd. is greatly acknowledged.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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