Case Report

# A case of fibro calculous pancreatic diabetes (FCPD)

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**Abstract:**

**Background:** Diabetes has now globally burst into an epidemic form. As the days passing we are learning more about this disease. Now diabetes mellitus is just about type 1 & type 2 diabetes, there are other entities of diabetes which need to be managed in distinguished ways. As in fibro calculous pancreatic diabetes (FCPD) which is a unique entity of diabetes often confused with type 1 diabetes as it is commonly associated early age onset, severe emaciation and negative family history of diabetes mellitus. FCPD is a non-alchoholicpancreatopathy where pancreatic calcification and chronic inflammation leads to exocrine and endocrine defects. It is a disease with male preponderance and mostly prevalent in the tropical region where malnutrition and poverty go hand in hand.

**Case presentation:** A 22 years old woman previously diagnosed a case of type 1 diabetes presented with the complaints of weakness, polyuria, polydipsia, weight loss, occasional abdominal pain and steatorrhoea. Surpringly despite not taking insulin for 7 months she didn't develop diabetic ketoacidosis. As theimaging of abdomen showed presence of pancreatic calculi, the diagnosis was changed to fibrocalculous pancreatic diabetes.

**Conclusion:** This is a classical case of FCPD. Diagnostic dilemma may arise at times. A good notion & discreetness are required for the early diagnosis & management to prevent the chronic complications, so that the patient can lead a better life.

**Key words:** Fibro calculous pancreatic diabetes (FCPD)

## Introduction:

FCPD is a form of diabetes which results due to exocrine defect of pancreas. It is one of the varieties of "diabetes due to other specific causes". It is secondary to "tropical calcific pancreatitis". This entity was 1st described by Zuidema in 1959 in patients from Indonesia[1]. FCPD is more prevalent among young non – alcoholic persons in tropical countries who belong to poor society and often malnourished[1].The cardinal features of FCPD are frequentabdominal pain since childhood and pancreatic calculi resulting dilatation of pancreatic duct as well as fibrosis of the pancreas in adolescence[2]. The onset of diabetes usually occurs in early adulthood in more than 90% of cases which is severe and requires insulin for management[3]. Although ketosis is rare.Various terminologies have been proposed for this kind of diabetes including pancreatogenous diabetes, pancreatic diabetes, and tropical pancreatic diabetes. Later World Health Organization (WHO) Study Group Report On Diabetes introduced the term Fibrocalculous Pancreatic Diabetes (FCPD) for this variety of diabetes[2,4].

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In spite of its high prevalence in tropical regions FCPD is still a rare form of diabetes consisting of less than 1% of total cases of diabetes in those regions[5]. A recent study in urban southern India reported a prevalence of 0.36% among subjects with self reported diabetes & 0.019% among general population[6].

## Case presentation:

A 22 years old lady, known case of type1 diabetes mellitus got admitted with the complaints of weakness, weight loss,polyuria & polydipsia for last 6 months. She has been a known case of type 1 DM for last 4 years and used totake insulin but for last 7 months she didn't any insulin and surprisingly DKA was not developed. She has also stated that she has been suffering from occasionalupper abdominal pain for last 10 years. She has been treated for PUD over last 10 years. Her abdominal pain was intermittent and burning in nature, radiated to back. She has also complained of steatorrhoea which aggravates after taking oily foods. She doesn't has any complaint of chronic cough,evening rise of temperature, dysuria or abnormal per vaginal discharge. She has no history of eating cassava, drinking alcohol, gall stone disease or hepatitis and no family history of diabetes mellitus or any pancreatic disease.

On examination she was emaciated, weight-44.24 kg,height-1.45 m, BMI-15.3 kg/m2, waist-70 cm, hip-84 cm, W/H ratio-.83, mildly pale, nonicteric, acanthosis nigricans absent, no lymphadenopathy, absence of any features of haemochromatosis or chronic liver disease.

On admission her random blood sugar was 27.3 mmol/l, bedside urinary albumin and acetone test revealed no abnormality.

X-ray abdomen showed dilated main pancreatic duct with pancreatic calculi. MRCP and ultrasonography also showed the same finding of x-ray abdomen.

Complete blood count revealed mcirocytic hypochromic anaemia with a Haemoglobin of 10.5 g/dl, MCH – 26.9 pg, MCHC – 31.2 g/dl. Serum Lipase was 85 U/L, Alanine aminotransferase was 45 U/L.

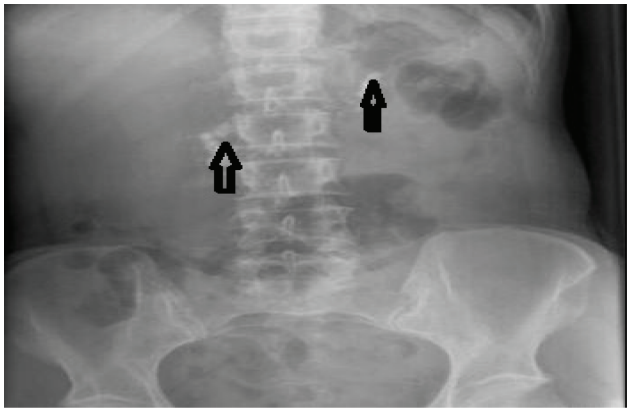

**Fig.-1:** X-ray abdomen showing presence of pancreatic calculi at the level of lumbar 1 (left Side)& 2 (right side) vertebrae

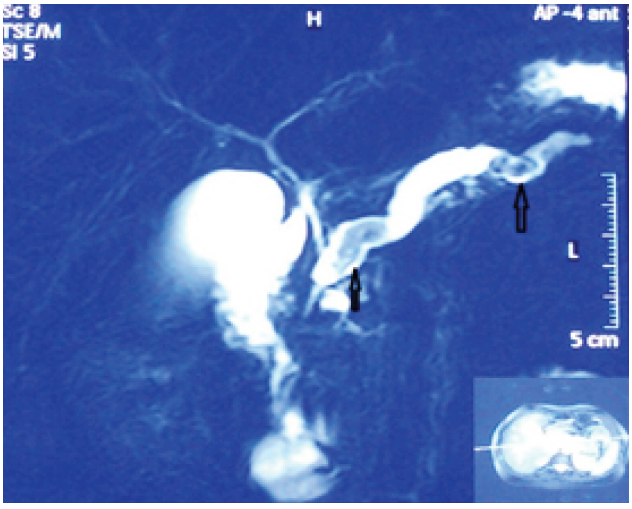


**Fig.-2:** MRCP showing dilated main pancreatic duct with calculi.

Based on her history & investigation findings the diagnosis of fibrocalculous pancreatic diabetes (FCPD) was made. Treatment was based on glycaemic control with insulin, management of pain and pancreatic function were done by endoscopic removal of pancreatic stone as well as supplementation of pancreatic enzyme.

On follow up visit, the patient showed significant improvement-her blood sugar was controlled, BMI was 18.6 and she was free of pain.

**Discussion:**

FCPD is a unique form of diabetes mellitus secondary to pancreatic calcification in non-alcoholic persons. Predominantly seen in young[1] – usually diagnosed between ages of 10 years to 40 years. There is a marked male predominance[7,4,8]. People are mostly diagnosed having previous histories of severe episodic epigastric pain. The pain usually abates by the time as the diabetes sets in. About one third of the patients complain of passing bulky or oily stool following taking oily foods. Patients also present with some other features, like: extreme emaciation (70%), a peculiar cyanotic hue of the lips, bilateral enlargement of parotid glands, distension of abdomen, etc[7, 4, 8, 9.] Previously it was believed that low socio-economic status has some contributions in developing FCPD as most the patients are from that background but nowadays patients are also seen from middle and upper strata of the society[9,10]. Although most of the cases are of juvenile onset but there are some classical cases too, where a patient has been reported to have presented with the features of FCPD at the age of 49 years with no previous histories[11].

Several aetiologicalfactors are thought to be responsible for pathogenesis of FCPD.Malnutrition - it is believed micronutrient deficiency contribute to tissue damage. Malnutrition in early life is associated with beta cell dysfunction and glucose intolerance in later life[26,27,28]. A follow up study done in Pune[29] showed a low BMI in 72% of insulin requiring diabetes patientsimplying that diabetes related malnutrition is a significant factor. Cassava consumption - cassava is a tuber which containcyanogenic glycosides linamarin and lotaustralinleds to transient hyperglycemia after ingestion. It is yet to be proved that cassava consumption lead to permanent diabetes but may explain the prevalence of FCPD where tuber is consumed[12]. Other dietary factors - low fat intake may be responsible for occurrence of TCP[13,14]. Familial and genetic factors - familial occurrence is not uncommon in

FCPD[15]. Recent studies have showed 10% of cases of FCPD have familial aggregation[14, 16, 17]. Another study has supported genetic predisposition of FCPD where it was found that FCPD shares susceptibility genes in common with type 1 and type 2 diabetes mellitus[18]. Oxidant stress - studies have suggested that low intake of antioxidants like beta carotene, vitamin C, vitamin E may predispose to topical pancreatitis through free radical injury [19, 20, 21]. Evidence confirms a link between the serine protease inhibitor, KAZAL type-1 (SPINK1) gene and tropical calcific pancreatitis[22,23]. It is a vital protease inhibitor that preventsinappropriate activation of the pancreatic enzyme cascade by inhibiting trypsin activity.

There are no definite criteria for diagnosis of FCPD in spite of having excellent descriptions of the disease by various authors. Mohan et al [24] have proposed the following criteria for the diagnosis of FCPD, based on their studies and extensive review of the literature.

Diagnostic criteria for fibro-calculous pancreatic diabetes [24] –

- Occurrence in a tropical country.
- Diabetes by WHO study group 4 criteria.
- Evidence of chronic pancreatitis : pancreatic calculi on X-ray or at least three of the followings:
  - Abnormal pancreatic morphology by ultrasonography.
  - Chronic abdominal pain since childhood.
  - Steatorrhoea.
  - Abnormal pancreatic function.
- Absence of other causes of chronic pancreatitis. i.e. alchoholism, hepatobiliary disease or primary hyperparathyroidism.

The classical traid of FCPD consists of abdominal pain, steatorrhoea& diabetes. Although features like young age onset, malnutrition and ketosis resistance are commonly found in FCPD paients's but these are not considered as diagnostic criteria [25].

Patients with FCPD usually require insulin for manangement of diabetes. But interestingly they rarely develop ketoacidosis despite not using insulin for prolong periods[3,30]. In this case we find the similar history. This feature helps to distinguish them from other entities of ketosis prone diabetes. Yajnik[31] has summarized the various hypotheses to explain the ketosis resistance in malnutrition related diabetes. They are as follows:

- Residual beta cell function adequate to prevent ketosis.
- Concomitant destruction of alpha cells & thus loss of glucagon, a major ketogenic hormone.
- Subcutaneous fat loss resulting decreased supply of Non Esterified Fatty Acids (NEFA) which is a fuel for ketogenesis.
- Resistance of subcutaneous adipose tissue lipolysis to adrenaline.
- Carnitine deficiency affecting transfer of NEFA across the mitochondrial membrane.

The classic radiological finding in FCPD is the presence of pancreatic calculi on a plain X-ray of abdomen[7,3.] The calculi are mostly found to the right of the first or second lumbar vertebrae, such as in this case but occasionally overlap the spine. In some cases the whole pancreas may be studded with calculi. It is extremely rare to find isolated calculi left to the vertebrae[16]. Calculi are usually large and rounded andinvariably intraductal in location. Ultrasonography also helps to localize calculi to the pancreas and document others feature of chronic pancreatitis, e.g. ductal dialation. CT scan studies done by Yajnik[31] showed that pancreatic mass was preserved in early stages with swelling of parenchyma followed by varying degrees of atrophy as the disease progresses and finally being replaced by the " bag of stones" appearance in extreme cases. Endoscopic retrograde cholangio-pancreatography (ERCP) shows marked ductal changes in presence of pancreatic calculi[32].

No microvascular and macrovascular complications were found in this case. But microvascular complications are commonly found in FCPD patients in long term cases, e.g.retinopathy,nephropathy,neuropathy[33,34,9].

In contrast marcovascular complications are less common, perhaps due to usual young age of onset, lennness and low cholesterol levels [35].

FCPD patients usually require low dose of insulin, possibly due to presence of residual beta cell function[36]. Pain is a major problem in FCPD. Surgical interventions are often needed if there is recurrent and severely intractable pain following annual ERCP & stenting. Surgical options include drainage procedures, shincterotoplasty, pancreatic necrostomy and celiac plexus ablation. Early surgery has been found to prevent the development of diabetes in early stage of disease[37]. The challenges in management are recurrent intractable pain, malnutrition, recurrent hypoglycemia, poor drug compliance, misdiagnosis & late diagnosis.

**Conclusion:**

FCPD is classically found in young. Early diagnosis and treatment can prevent the chronic complications which require a good knowledge on FCPD as it is not uncommon in this subcontinent. Adequate nutrition, good glycemic control, proper pain management, regular monitoring can ensure an improved quality of life for FCPD patient.

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