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A Rare Case of Fibrocalculous Pancreatic Diabetes in a Young Sub-Saharan African Migrant

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Abstract

Fibrocalculous Pancreatic Diabetes (FCPD) is a rare form of secondary Diabetes Mellitus (DM) mainly describes in tropical regions, and presenting as the ultimate stage of the Tropical Chronic Pancreatitis (TCP). We present a case of FCPD in a young male recently immigrated to Tunisia from a Sub-Saharan African country, with no medical history. On clinical examination he had low body mass index of 18.6 kg/m2, ascites, upper abdominal pain on palpation and bilateral erysipelas. Laboratory tests revealed malnutrition with low albumin, cholesterol and folate levels as well as anemia. DM was particularly ketosis-resistant and difficult to manage because of glycemic variability. The exact etiopathogenesis is still unclear, but a genetic susceptibility is incriminated, and malnutrition seems to be the consequence rather than the cause. Alongside with insulin therapy, nutritional intervention includes pancreatic enzyme and nutrients supplementation with low-fat high-protein diet. To our knowledge, this is the first reported case of FCPD in Tunisia in decades, and it is a concerning problem with the rise of immigration.

Keywords: Fibrocalculous pancreatic diabetes; Tropical chronic pancreatitis; Diabetes mellitus; Tunisia

Introduction

Fibrocalculous Pancreatic Diabetes (FCPD), defined as idiopathic non-alcoholic Chronic Pancreatitis (CP), found particularly in tropical regions and presenting with abdominal pain and pancreatic calcification, is a unique and rare form of secondary Diabetes Mellitus (DM) with an estimated prevalence of 0.2% to 1.6% among all types of DM. The entity preceding the onset of FCPD is called Tropical CP (TCP) and typically occurs in younger subjects [1]. To our knowledge, TCP has not been recently reported in Tunisia. We present a case of FCPD in a young male recently emigrated from a Sub-Saharan West African country.

Case Presentation

A 17-years-old male, illegally immigrated three months ago to Tunisia from a Sub-Saharan African country and a former rice farmer with no medical history, was admitted to the gastroenterology department in September 2023 for a severe malnutrition and non-cirrhotic ascites, then transferred to our department of endocrinology for management of newly diagnosed DM.

He had no history of chronic abdominal pain or steatorrhea. Physical examination revealed extracellular dehydration, low Blood Pressure (BP) of 110/70 mmHg, distended abdomen and epigastric pain on palpation (Figure 1). He weighed 41 kg with a Body Mass Index (BMI) of 18.6 kg/m^2. The diagnosis of erysipelas has been made in the presence of an erythematous, edematous and painful rash on the left leg.

Laboratory findings revealed random serum glucose of 27 mmol/L, significant glycosuria (3+) without ketonuria and HbA1c of 16%. Serum calcium level, liver function and creatinine clearance tests were normal. Also, there was no hypertriglyceridemia.

Serum tests exploring malnutrition showed low albumin level (25.6 g/L), normochromic anemia with a hemoglobin (8.4 g/dL), low serum cholesterol (115 mg/dL) and low folate level (2.57 ng/mL).

For ascites investigation, serology of viral hepatitis A, B, C and Human Immunodeficiency Virus (HIV) were negative.

Type 1 DM related autoantibodies (IA-2A, GAD-65, and ICA) were negative.

On the second day of admission, he suffered from abdominal pain and lipase level was 37 IU/L

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Figure 1: Patient phenotype.

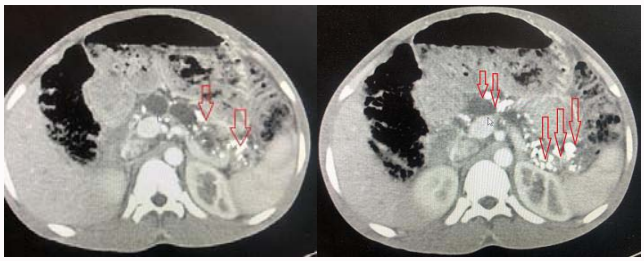

Figure 2: Abdominal CT scan.

(normal range: <65). An abdominal CT scan revealed practically destroyed atrophic pancreas with dilation of the Wirsung duct and macrocalcifications of almost the entire parenchyma (Figure 2).

He was diagnosed with FCPD following these criteria: younger age of onset, no alcohol consumption, DM, malnutrition extensive pancreatic calcification and ductal calculi.

For etiological investigation there were no similar cases in his family and no Cassava consumption. Nutrient profile revealed deficit in protein intake (protein: 11.85 %, carbohydrate: 62.36% and fat: 25.77%).

The DM was difficult to equilibrate and the patient was treated symptomatically with intravenous fluids, basal-bolus insulin therapy with a daily dose of 0.98 units per day (insulin analogues), analgesics and pancreatic enzyme supplements while avoiding refeeding syndrome. He was put on Cefazolin and rest for erysipelas.

Discussion

TCP is defined as a juvenile form of non-alcoholic CP, characterized by younger age of onset, large pancreatic calculi, accelerated evolution towards DM and/or steatorrhea, and susceptibility to cancer development [2]. While FCPD is defined as unique form of secondary DM associated to TCP [1]. This latter has been described for the first time in Indonesia in 1959 [3] and since then in many other developing tropical countries and it seems to be endemic in some regions of the Indian sub-continent [4].

In Tunisia, TCP is uncommon, except for a small series of ten patients in the early 1990's by Chatti et al. and four of them had FCPD [5] and to our knowledge, this is the first described case in decades, but involving a migrant from a West African country.

The exact etiopathogenesis of FCPD is still unclear. For decades, malnutrition has been considered the main cause, following the epidemiological features of the disease, but it seems to be the consequence of pancreatic damage rather than the cause. Plus, cassava prolonged consumption, as a source of carbohydrates, was particularly considered a major factor leading to pancreatic cyanide-mediated damage. Another possible nutritional theory is high-carbohydrate, independently of the source, and low-protein diet, that may predispose to CP [1,6]. And for this patient, rice was indeed the main nutritional source, which is in favor of the latter theory. Genetic susceptibility is another possible theory for TCP and FCPD in the presence of familial clustering in some tropical countries. Candidate genes are Serine Protease Inhibitor Kazal (SPINK) type 1, cathepsin B and cationic trypsinogen gene (PRSS1), and anionic trypsinogen (PRSS2). Possibly, defect in SPINK1 leads to deficiency of pancreatic secretory trypsin inhibitor, a protein that protects the pancreas from prematurely activated trypsinogen damage, and as a consequence recurrent acute pancreatitis destroying the gland [1,6]. The patient did not mention any similar case in his family, but we assumed that the condition is certainly underdiagnosed because of lack of healthcare accessibility.

The clinical presentation of FCPD is typically recurrent abdominal pain, steatorrhea and ultimately DM [1]. In this case, it was difficult to establish this triad due to some communication difficulties and language barriers and the patient may have had recurrent episodes of acute pancreatitis since his childhood, and since he is actually in third phase of the natural history of the disease, abdominal pain is usually absent or less important. And for pancreatic exocrine insufficiency, it was obvious to confirm in the presence of clinical and biological stigmata of fat and vitamin malabsorption. The nutritional profile of patients with FCPD is typically low vitamin D and high triglycerides level, and low energy, carbohydrates, protein, fiber, thiamine, niacin intake. Besides the low socioeconomic status, other factors may contribute to malnutrition: Fear of pain and steatorrhea and the increased basal metabolism [7]. Endocrine pancreatic insufficiency occurs approximately after 5 years of evolution and in about 90% of TCP cases, and typically before the age of thirty years. FCPD is insulin-dependent, ketosis-resistant and brittle DM with manifest glycemic variability and frequent hypoglycemia [1,8]. For this patient, DM was in deed very difficult to manage because of glycemic variability and required an intensified insulin protocol. FCPD is certainly ketosis-resistant in relation to several pathophysiological mechanisms including sufficient residual insulin secretion to prevent ketogenesis, but not to overcome hyperglycemia, deficient substrates for ketogenesis especially fatty acids, glucagon deficiency secondary to pancreatic damage and possible decreased insulin sensitivity [1]. On imaging study, large ductal calcifications, ductal dilatation evolving mainly the Wirsung duct and pancreatic atrophy are very suggestive of FCPD [6,8]. Recent criteria of TCP have changed recently and include, in the absence of other cause of CP, younger age of onset, history of malnutrition, and ultimately DM in presence of extensive pancreatic calcification and ductal calculi. Another non necessary criterion for TCP is low BMI<18.5 kg/m^2, because cases with normal weight and even obesity have been described. Idiopathic Chronic Pancreatitis (ICP) is diagnosed when the criteria of TCP are not complete [8]. In India, where CP is frequent, some authors consider TCP as a special form of aggressive ICP, and that the term

'tropical' is a misnomer for the disease [9].

Chronic hyperglycemia in FCPD is associated with the similar risk of diabetic microvascular complications as in type 2 DM, but not for macrovascular disease. Besides that, malnutrition, a consequence of the disease rather than its cause, is secondary to fat and fat-soluble vitamins malabsorption. However, the most concerning matter is the markedly increased risk of pancreatic adenocarcinoma compared to other forms of CP, and for now there is no clear consensus on the surveillance modalities [10].

Finally, insulin analogues were preferred for this patient to better control the high glycemic variability, to prevent hypoglycemia and to achieve an acceptable level of HbA1C (<7%), alongside with pancreatic enzyme supplements and low-fat, high-protein diet and vitamin D supplementation. In fact, pancreatic enzyme therapy is important for digestion and nutrients absorption as well as for preserving incretin hormone secretion for better glycemic control. And it should be noted that pancreatic polypeptide is a promising treatment for FCPD by improving insulin sensitivity [10].

Conclusion

With the rise of Sub-Saharan African immigration to Tunisia, TCP may be observed more frequently in our environment. We report an interesting case of FCPD with a history of severe malnutrition. Early diagnosis and better management of the endocrine and exocrine dysfunction with an appropriate pancreatic enzyme replacement could help to ensure better survival and improve the prognosis and quality of life of these patients. Until now, there is no clear or a proven effective consensus for screening patients with CP for pancreatic cancer. Therefore, screening tests should be improved.

References

1. Unnikrishnan R, Mohan V. Fibrocalculous Pancreatic Diabetes (FCPD). Acta Diabetol. 2015;52(1):1-9.
2. Barman KK, Premalatha G, Mohan V. Tropical chronic pancreatitis. Postgrad Med J. 2003;79(937):606-15.
3. Zuidema PJ. Cirrhosis and disseminated calcification of the pancreas in patients with malnutrition. Trop Geogr Med. 1959;11(1):70-4.
4. Witt H, Bhatia E. Genetic aspects of tropical calcific pancreatitis. Rev Endocr Metab Disord. 2008;9(3):213-26.
5. Chatti N, Chaieb L, Jemni L, Letaief R, Tlili-Graies K, Hochlef S, et al. Juvenile idiopathic chronic calcifying pancreatitis: Report of 10 cases from central Tunisia. Pancreas. 1990;5(3):354-7.
6. Dasgupta R, Naik D, Thomas N. Emerging concepts in the pathogenesis of diabetes in fibrocalculous pancreatic diabetes. J Diabetes. 2015;7(6):754-61.
7. Joseph M, Dasgupta R, Ramachandran R, Anoop S, Ananad V, Devanithi N, et al. Nutritional intake in low Body Mass Index (BMI) males with type 1 diabetes and fibrocalcific pancreatic diabetes: What are the unmet needs? A cross-sectional study from a south Indian tertiary care hospital. J Clin Diagn Res. 2017;11.
8. Kangas-Dick A, Khan U, Awoniyi O, Waqar S, Tun NN, Viswanathan K, et al. A case of chronic calcific nonalcoholic pancreatitis. Case Rep Gastrointest Med. 2016;2016:2963681.
9. Garg PK, Narayana D. Changing phenotype and disease behavior of chronic pancreatitis in India: Evidence for gene–environment interactions. Glob Health Epidemiol Genomics. janv 2016;1:e17.
10. Makuc J. Management of pancreatogenic diabetes: Challenges and solutions. Diabetes Metab Syndr Obes. 2016;9:311-5.