Ketoacidosis a Rare Complication of Fibrocalculous Pancreatic Diabetes

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Abstract
In a tropical country a case of ketosis resistant insulin dependent diabetes mellitus occurred in young lean patient who also shows evidence of malnutrition and pancreatic calculi with or without fibrosis and intermittent abdominal pain could be regarded as Fibrocalculous Pancreatic Diabetes (FCPD). We reported of a 24 years old female patient with shortness of breath since one day before she was admitted to the hospital. She also had a fever with sore throat and recurrent abdominal pain. Patient had history of abdominal pain, at that time she also developed white stools. She had a two years history of generalized body weakness associated with progressive weight lost, polydipsia and polyuria.Physical examination revealed a young lady with a low Body Mass Index (BMI) and tonsilopharyngitis. The haematological examination shown hyperglycemia, ketoacidosis, and leukocytosis. The abdominal x ray and ultrasound scan showed pancreatic calcification. The classical clinical picture of FCPD is primarily of a young diabetic patient presenting with recurrent epigastric pain, steatorrhoea, signs of malnutrition and micronutrient deficiencies. Although this case caused by ketoacidosis resistant, under special circumstances particularly after stress for example infection, trauma, etc ketosis could also be occurred.

Key word: ketoacidosis, complication, fibrocalculous pancreatic diabetes

Abstrak
Di negara tropis, kasus resistensi terhadap ketosis yang terjadi pada pasien diabetes mellitus tipe 1 (DMT1) berusia muda, bertubuh kurus, dan disertai penyulit berupa malnutrisi dan batu pankreas dengan atau tanpa fibrosis dan nyeri abdomen intermiten, dapat disebut sebagai Diabetes Melitus Terkait Malnutrisi (DMTM). Dilaporkan kasus seorang perempuan berusia 24 tahun dengan keluhan utama sesak nafas sejak 1 hari sebelum masuk RS. Keluhan juga disertai demam, sakit tenggorokan dan nyeri abdomen berulang disertai steatorroae. Sejak 2 tahun ini ia merasa badan lemas dengan penurunan berat badan progresif, polidipsi dan poliuri. Pemeriksaan fisik didapatkan Indeks Massa Tubuh (IMT) rendah dan tonsilofaringitis. Hasil laboratorium menunjukkan hiperglikemia, ketoasidosis, dan leukositosis. Foto polos dan USG abdomen menunjukan adanya kalsifikasi pankreas. Gambaran klasik dari DMTM adalah pasien DM muda dengan gejala nyeri epigastrium berulang, steatorroea, tanda-tanda malnutrisi dan defisiensi mikronutrien. Walaupun kasus ini disebabkan oleh resistensi terhadap ketoasidosis, namun pada kasus tertentu dengan penyulit seperti infeksi, trauma, dan sebab lainnya, ketosis dapat terjadi.

Kata kunci*: ketoasidosis, komplikasi, Diabetes terkait Malnutrisi*

Background

In a tropical country ketosis resistant, insulin dependent diabetes mellitus occurred in young lean (BMI < 19) patient who also shows evidence of malnutrition and pancreatic calculi with or without fibrosis with intermittent abdominal pain maybe regarded as FCPD provided that alcoholism and biliary disease have already been excluded.[1] Diabetes onset in early adulthood, was severe and usually insulin dependent although ketosis is rare.[2] Though, by definition these cases are ketosis resistant, but under special circumstances particularly after stress e.g. infection, trauma, etc, ketosis could be occurred.[1]

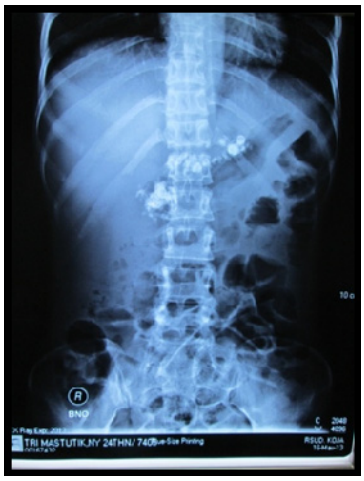
Case Presentation

A 24 years old female patient was admitted to internal medicine unit with shortness of breath (dyspnea) since one day before hospital admission. Dyspnea occurred when patient working and resting, and becames better when patient lying down. There was no trauma, allergic, or asthma history. She also had a fever with some sore throat since one day before. Additionally, she complained abdominal pain located at the epigastrium and it radiates to the back on either sides and was better when lying down. Patient said thatshe had felt abdominal pain since a long time ago, but she forgot when exactly it's occurred. Since patient had history abdominal pain, she also had white stools. She had a two years history of generalized body weakness associated with progressive weight lost. She also had polydipsia and polyuria.

Physical examination revealed a young lady with a low BMI of 13.88 kg/m^2. The tonsil was at stage T3-T3, and the pharynx hyperemia. On the neurological examination, she was fully conscious. Deep tendon reflex, joint position and vibration are normal. Musculoskeletal examination revealed generalized muscular hypotrophy.

The haematological investigations done included a raised random blood sugar level of 404 mg/dl. Keton 3.6 the complete blood count showed a leukocytosisof 20.000 u/mm^3. The blood gas interpretation showed compensated metabolic acidosis with pH 7.275, pCO_2 11.0, pO_2 176.2, HCO_3 5.1, BE -21.9.The abdominal x ray showed pancreatic calcification. The thorax x-ray was normal. An abdominal ultrasound showed diffuse calcification of the pancreas.

A diagnosis of FCPD with malnutrition and complicated by ketoacidosis with tonsilopharyngitis was established. The patient has been given NaCl 0,9% infusion and insulin injection for her ketoacidosis and and antibiotics for her pharyngitis. After ketoacidosis disappeared, insulin injection was still administered continuosly. She also received dietary counseling for her malnutrition. She was discharged from the hospital when she had much improvement.



Picture 1. Pancreas Calcification

Discussion

In this case report, we report a case of a young patient presenting with clinical features of FCPD with ketoacidosis complication. It was a classical form as described by Geevarghese,. FCPD has several distinct characterictics.[2] The classical recognized clinical picture of FCPD was primarily of a young diabetic patient (between the ages of 10 years and 40 years) presenting with recurrent epigastric pain, steatorrhoea, signs of malnutrition and micronutrient deficiencies.[3]Mohan et al have proposed the following criteria for the diagnosis of FCPD, based on their own studies and extensive review of the literatures (see table 1).[4]

Table 1. Diagnostic Criteria for Fibrocalculous Pancreatic Diabetes (FCPD)[4]

1. Occurrence in a "tropical" country. 2. Diabetes by WHO Study Group criteria. 3. Evidence of chronic pancreatitis : pancreatic calculi on X-ray or at least three of the following : a) Abnormal pancreatic morphology by sonography b) Chronic abdominal pain since childhood c) Steatorrhoea d) Abnormal pancreatic function test 4. Absence of other causes of chronic pancreatitis, i.e. alcoholism, hepatobiliary, disease or primary hyperparathyroidism etc.

Presentation of prolonged abdominal pain, steatorrhoea, hyperglycemia, and pancreatic calculi on plain abdominal x-ray and ultrasound scan confirmed the diagnosis as shown by the patient.

Features like clinical malnutrition, young age at onset and absence of ketosis are useful clues but were not diagnostic criteria by themselves.[2] Etiopathogenesis of FCPD was relatively hardly explained, but several possible mechanisms of causation have been suggested to explain development of FCPD, they include malnutrition, which was particularly protein energy malnutrition, Casava hypothesis, food toxin, xenobiotics, and genetic factors.[1]

FCPD is should be ketosis resistant, many studies have offered several explanations. Earlier studies has suggested a number of mechanisms such as low adipose tissue mass and delayed mobilization of free fatty acids from adipose tissue.[5] Recent studies shown patient with malnutrition diabetes has low glucagon levels after administration of oral glucose. The lack of glucagon response may be one of the factors responsible for protection against ketoacidosis.[6]In one study on pancreatic B-cell function using C-peptide as a marker shown that FCPD patients have some residual B-cell function.[7]

Although this case could be regarded as ketoacidosis resistant, under special circumstances particularly after stress e.g. infection, trauma, etc ketosis can be developed, as demonstrated in this case.[1] Beside that, it has been found that when FCPD were well feeding and when they gain weight they may develop ketosis.[1]

Management of FCPD cases was similar to the principle of management of all diabetes.[1]Since ketoacidosis was occurred in FCPD we give therapy similar with ordinary ketoacidosis occurred in diabetes mellitus. However, there were some points worth mentioning in FCPD like this case should be no calorie restriction and fatty food restriction if steatorrhoea was present.[1]

Management of FCPD cases ~~is~~was similar to the principle of management of all diabetes.[1]However, due to insufficient of its endocrine function, FCPD need insulin treatment for longterm (continuously). The patient will not be able to use any oral hypoglycemic agent. Diabetes was usually quite severe and insulin dependent type.[8]

Conclusion

Fibrocalculous Pancreatic Diabetes still exist in Indonesia. Although based on the pathogenesis this case caused by ketoacidosis resistant, but it could also caused by stress or weight gain. However, the management of FCPD caseswas basically~~is~~ similar to the principle of all diabetes management.

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