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Case Report

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Fibrocalculous pancreatic diabetes (FCPD) presenting as regression of puberty–a rare presentation of a rare disease

Abstract

Here we present a case of young female who presented to us with secondary amenorrhea causing regression of puberty which is quite uncommon as a presentation of FCPD which is occasionally encountered in our day to day clinical practice. The aim of this communication is to keep a high index of suspicion and to keep FCPD as a possible aetiology which can lead to regression of puberty.

Keywords: fibrocalculous, pancreatic, diabetes, amenorrhea, abdomen

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Introduction

FCPD is a rare form of diabetes mellitus which is secondary to chronic calcification of the pancreas in young non-alcoholic people and exclusively found in tropical countries.[1] The characteristics of the disease are the presence of abdominal pain in childhood and pancreatic calculi associated with dilation of the pancreatic duct and fibrosis of the gland in adolescence.[2] The patients generally have clinical trial of pain abdomen, steatorrhea, diabetes.

Case report

A 15 year old female presented to the outpatient clinic with history of amenorrhea for the past seven months and failure to thrive. Possibility of pregnancy was excluded by history and negative urine for pregnancy test. On physical examination, atrophy of breasts and gross emaciation was noted. Tanner stage 2 was assigned. However on further history taking it was discovered that patient suffered from recurrent bouts of pain abdomen in her childhood which used to get aggravated by meals and lying down position and relieved by stooping forward. On further examination, pallor was noted which could be attributed to her nutritional status. As a part of workup routine random blood glucose was done which came out to be unexpectedly high, which lead to suspicion of FCPD as a distinct possibility responsible for the current predicament of the patient? On further enquiry she confessed to having osmotic symptoms. Her urine analysis showed glycosuria and absence of ketone bodies. Her haemoglobin was 9 gm/dl while rest of the biochemical investigation were unremarkable.

Ultrasonography of whole abdomen was done which showed atrophic pancreatic parenchyma with heterogeneous and hypoechoic shadows suggestive of chronic calcific pancreatitis. Her follicle stimulating hormone level was 1mIU/ml and estradiol was 15pg/ml both were suppressed leading to functional hypothalamic amenorrhea. Her Vitamin D a level was also suppressed i.e. 10.77 ng/ml as a consequence of fat and vitamin malabsorption. Her fasting blood glucose level was 219 mg/dl, post prandial blood glucose was 347 mg/dl with an HbA1c of 13.1%. She was started on intravenous fluids to correct her dehydration with basal bolus regimen of insulin consisting of injection FIASP and injection Glargine. Injections FIASP is a rapidly acting Aspart and was chosen because of the flexibility of injection around meal times and she also received pancreatic enzyme supplementation of tablet Pancreatin 10000 units before breakfast, 25000 units before lunch and dinner. Her condition improved with treatment and patient was discharged in a haemodynamically stable condition.

Discussion

Fibrocalculous pancreatic diabetes as a form of secondary diabetes is a rarity. Extreme emaciation, a peculiar cyanotic hue of the lips , bilateral parotid gland enlargement and distension of the abdomen are some of the classic clinical features.[3–5] Genetic factors have been concluded as the most significant in causation with a link between the Serine Protease Inhibitor, Kazal type (SPINK 1 gene) and TCP (11,12). However in our case report except emaciation no other classical features were evident. In a series by Dr Mohan et al.,[6] malnutrition was observed in 25% cases although 70% were lean. Recently however there appears to be a shift in the paradigm in presentation of FCPD. Although despite uncontrolled hyperglycaemia, ketosis is absent because of residual beta cell function reflected by intermediate levels of C peptide.[6] Large number of patients require insulin for hyperglycaemia control.

Conclusion

FCPD is a rarity with existence of few cases in developing countries like India. It is a viral protease inhibitor that prevents unregulated or inappropriate activation of the pancreatic enzyme cascade by inhibiting trypsin activity.[7] Strangely macro vascular complications are less common.[8] Perhaps due to relatively young age of patients and leanness and low cholesterol level.[9,10] The aim of this communication is to aware physicians of the multitudinous presentation of FCPD and to keep it as a possibility while evaluating patients of secondary amenorrhea.[11,12]

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Conflicts of interest

The authors declare that they have no conflicts of interest.

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