Case Report

Diffuse nesidioblastosis diagnosed on a Ga-68 DOTATATE positron emission tomography/ computerized tomography

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ABSTRACT

The authors describe a 50 days old pre-term infant with persistent hyperinsulinemic hypoglycemia of infancy in whom Ga-68 DOTATATE positron emission tomography/computerized tomography scan showed diffusely increased tracer uptake in the entire pancreas with no abnormal tracer uptake anywhere else in the body, suggestive of a diffuse variant of nesidioblastosis.

Keywords: Ga-68 DOTATATE, hyperinsulinemic hypoglycaemia, infant, nesidioblastosis, positron emission tomography/computerized tomography

INTRODUCTION

Nesidioblastosis is a pathological condition caused by neo-differentiation of islets of Langerhans from pancreatic ductal epithelium. This condition later renamed as persistent hyperinsulinemic hypoglycaemia of infancy (PHHI) exists in two forms. One corresponds to a focal pancreatic adenomatous hyperplasia (focal PHHI) and the other is characterized by a diffuse cell abnormality (diffuse PHHI). The underlying pathology and treatment strategies are different for both the types however differentiation by clinical and biochemical parameters is not feasible. No definite role of imaging to differentiate diffuse versus focal condition has been reported in the literature.

CASE REPORT

A 50-day-old pre-term female infant presented with persistent hyperinsulinemic hypoglycemia. The child was born at 35 weeks of gestation, with breathing difficulty since birth. She developed cyanosis on day 2 of life with seizures thereafter. On investigation the child was found to be hypoglycemia (blood glucose level = 16.8 mg/dl) and hyperinsulinemia with serum insulin levels of 149.70 IU/ml (n = 2.0 IU/ml). Screening for sepsis was negative. The child required intravenous glucose infusion up to 10 mg/kg/min and oral feeds fortified with glucose to maintain euglycemic state. Ga-68 DOTATATE positron emission tomography/computerized tomography (PET/CT) scan showed diffusely increased tracer uptake in the entire pancreas with no abnormal tracer uptake anywhere else in the body, suggestive of a diffuse variant of nesidioblastosis [Figure 1]. The child is currently on injection octreotide but not fit for definitive surgical management (near total pancreatectomy).

DISCUSSION

Laidlaw in 1938 first identified the disease and coined the term nesidioblastosis to describe the neodifferentiation of islets of Langerhans from pancreatic ductal epithelium.[1] This condition later renamed as PHHI of infancy exists in two forms. One corresponds to a focal pancreatic adenomatous hyperplasia (focal PHHI) and the other is characterized by a diffuse cell abnormality (diffuse PHHI).[2-4] These two forms could not be differentiated by clinical or biochemical data, although their underlying pathological mechanisms and the treatment remains totally different.[5] F18-fluoro-dihydroxyphenylalanine (F-DOPA) PET scan has been used to detect the hyperfunctional pancreatic islet tissue and to differentiate between focal and diffuse PHHI with a reported accuracy of 96% in diagnosing focal or diffuse disease and 100% in localizing the focal lesion.[6,7] The principle behind the use of F-DOPA PET in PHHI is that neuroendocrine pancreatic cells have an affinity for taking up F-DOPA and

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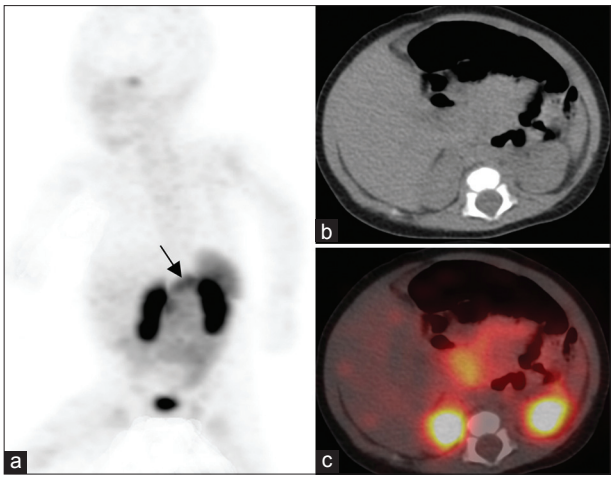

Figure 1: Ga-68 DOTATATE positron emission tomography/computed tomography (CT) (a) Maximum intensity projection (b) transaxial CT (c) transaxial fused images showing diffusely increased tracer uptake in the entire pancreas (arrow) with no abnormal tracer uptake elsewhere in the body, suggestive of a diffuse variant of nesidioblastosis

decarboxylate it into dopamine through aromatic amino acid decarboxylase.[8] Similarly neuroendocrine cells of pancreas also express high affinity somatostatin receptors (SSTR). 68-Ga DOTATATE specifically binds to the SSTR type II, which are highly concentrated in the pancreatic islet cells.[9] Previously only one case report was published where Ga-68 DOTATOC PET scan was used to differentiate focal versus diffuse nesidioblastosis with limited success.[10] This is the first case where Ga-68 DOTATATE PET scan had been used to successfully differentiate focal versus diffuse nesidioblastosis and thus help in tailoring the management in the infant.

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