Positron Emission Tomography in Congenital Hyperinsulinism

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We report a case of congenital hyperinsulinism with diffuse pancreatic abnormality diagnosed preoperatively by using [18-F]-L-DOPA positron emission tomography (PET). The infant was referred to us for recurrent hypoglycemia. Critical blood sample revealed increased insulin: glucose ratio. DOPA PET scan revealed diffuse involvement of pancreas. Subtotal (95%) pancreatectomy was done. Infant remained euglycemic on breast feeds at discharge.

Key words: *Congenital hyperinsulinism, [18-F]-L-DOPA positron emission tomography, Subtotal pancreatectomy.*

Congenital hyperinsulinism, previously called Primary islet cell hypertrophy (nesidioblastosis) is also known as Persistent hyperinsulinemic hypoglycemia of infancy (PHHI). It is characterized by nonketotic hypoglycemia in association with elevated insulin levels (>10 µU/mL). It is the most common cause of persistent and recurrent hypoglycemia in neonates [1,2]. Surgical treatment is indicated if medical therapy fails [2,3]. The distinction between focal and diffuse lesions is critical in planning surgical intervention but is not possible clinically. We are presenting a case of congenital hyperinsulinism with diffuse pancreatic abnormality diagnosed preoperatively by using [18-F]-L-DOPA Positron emission tomography (PET).

CASE REPORT

A female baby, product of a non-consanguineous marriage was born to 28 year old mother at term by vaginal route at a private hospital. Antenatal period was uneventful and mother's random blood sugar in third trimester was 86 mg/dL. At birth, baby cried after tactile stimulus and breastfeeding was started at 1 hour of life. On day 2, baby became lethargic, did not accept feeds and had multiple episodes of seizures. Blood sugar was 20mg/dL, sepsis screen were negative and lumbar puncture was normal. The infant was started on glucose infusion which was hiked to 12 mg/kg/minute on which she was maintaining euglycemia. Every attempt to decrease the glucose infusion rate resulted in hypoglycemia. Baby had multiple episodes of hypoglycemic seizures during first three weeks of life for which she was transferred to us. At admission, baby's random blood sugar was 18 mg/dL. Glucose infusion was maintained at 12 mg/kg/min to maintain euglycemia. It was not possible to decrease glucose infusion rate. Critical blood sample sent during an episode of hypoglycemia revealed blood sugar level 28 mg/dL, serum insulin 23µU/mL and serum insulin to blood glucose ratio 0.8. Serum pH and ammonia were normal and urinary ketones and reducing substance was negative. A diagnosis of congenital hyperinsulinism was confirmed. On day 30 of life, subcutaneous octreotide was started at 30 µg/kg/day in four divided doses. Dextrose infusion was subsequently tapered and baby was maintaining euglycemia on exclusive breast feeding. On day 36 of life, oral diazoxide was started and injection octreotide was stopped (so that baby could be discharged on oral drugs) but baby could not maintain euglycemia and injection octreotide was restarted. To ascertain the

type of pancreatic abnormality (focal or diffuse) [18-F]-L-DOPA PET scan was done on day 120 of life. It was suggestive of diffuse variety of congenital hyperinsulinism.

During the entire period, baby stayed in the hospital and remained euglycemic. Baby had developmental delay in all domains at 16 weeks of life. However, parents were not affording medical management and surgery was planned. Spleen preserving near total pancreatectomy was done at 5 month of life. Post operative period was uneventful. Histopathology revealed diffuse hyperplasia of islet cells. After two weeks of pancreactectomy, octreotide was gradually tapered and baby maintained euglycemia on exclusive breast feeding. Baby was discharged on post operative day 20 on exclusive breast feeding.

DISCUSSION

In 2003, researchers in Finland reported a new imaging technique using [18-F]-L-DOPA PET scanning to distinguish between focal and diffuse pancreatic lesion and to localize in the case of focal lesion [4]. The [18-F]-L-DOPA scan is currently available at only a few centers worldwide. This technique is 96% accurate for differentiating between focal or diffuse disease and 100% accurate in localizing the focal lesion [5]. However, there are no reports on the use of (18-F)-L-DOPA positron emission tomography in congenital hyperinsulinism till date from India.

Every effort should be made, both before and during surgery, to identify or rule out a focal lesion. Finding a focal lesion can potentially prevent unnecessary pancreatic resection. Most focal lesions are too small to identify by CT scanning, MRI, or even intraoperative palpation. Invasive procedures like pancreatic venous sampling or intra-arterial calcium stimulation may help identify a focal lesion but they are technically difficult with low sensitivity [6].

Diagnosis of congenital hyperinsulinism is based on "critical" samples (samples drawn at a time of fasting hypoglycemia: plasma glucose <50 mg/dL). The finding of nonketotic hypoglycemia in association with elevated insulin levels (>10 μU/mL) and normal levels of free fatty acids (FFA) confirms the diagnosis of hyperinsulinism. The insulin-to-glucose ratio may range from 0.4-2.7 (normal <0.3) [7].

Surgical treatment is indicated if medical therapy fails, if a discrete lesion is identified, or if patient's family is unable or unwilling to comply with medical therapy. Subtotal (95%) pancreatectomy is the most widely accepted procedure for infants and children [8]. Hypoglycemia often persists even after a 95-98% pancreatectomy. Hypoglycemia may be easier to control after surgery and may resolve or persist throughout life. Patients who undergo pancreatectomy are at high risk for developing diabetes mellitus later in life. In some series, a high frequency of mental retardation, developmental delay, and non hypoglycemic seizures has been observed. These findings are generally attributed to minimal brain damage from early hypoglycemic events, although the existence of these disorders as inherent co-morbid conditions along with congenital hyperinsulinism has not been fully excluded [9]. Some data suggest that patients with early severe disease treated with early, aggressive surgery have a better neurodevelopment outcome [10].

To conclude, critical blood sample and [18-F]-L-DOPA PET scan are crucial in the diagnosis of congenital hyperinsulinism. In India,(18-F)-L-DOPA PET scanning is limited to some regional centers. Hence, it is all the more important to be aware of its utility for optimal utilization. Aggressive management is advocated to prevent neurological sequel.

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Ultrasonography for Masseter Muscle Cysticercosis

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Solitary cheek swellings can present a diagnostic dilemma. We managed two children 10 y and 8 y presenting with pain and swelling on one side of cheek for over 15 d and no constitutional symptoms. Sonography showed cysticercosis in both of them. We treated both with steroids and albendazole, with good response.

Key words: *Cysticercosis, Masseter muscle, Ultrasonography.*

Cysticercosis, the infestation with the encysted larval stage of the parasite *T. Solium* commonly infests the brain, but muscles are also often affected [1-6]. Intramuscular cysticercosis has non-specific manifestations and diagnosis can be difficult. High resolution sonography (USG) can demonstrate the classical cyst with scolex within, and is a convenient test for diagnosis [1-3]. We present two patients with solitary cheek swellings where USG helped diagnose masseter muscle cysticercosis.

CASE REPORTS

Case 1: A 10 year old girl, resident of Mumbai, was brought with a painful swelling over the right cheek for 2 months. There was no fever or other symptoms. The whole right cheek looked swollen and on palpation the swelling was tender, globular, 3 cm in diameter and felt firm in the center. We suspected a hematoma or soft tissue tumor. Blood counts were normal. High resolution ultrasonography of the swelling (***Fig.* 1**) revealed a well defined cystic mass with an eccentric echogenic nidus, the scolex, (arrow) within the masseter muscle fibers. There was surrounding edema fluid collection. An MRI of the swelling showed similar findings. We treated her with oral prednisolone 2mg/kg for 4 days and albendazole 15mg/kg for 28 days. The swelling disappeared after treatment.

Case 2: An 8 year old girl, resident of Nepal, presented with a similar, painless swelling over the left cheek for 15 days. There were no constitutional