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Laparoscopic Near Total Pancreatectomy for Infants with Congenital Hyperinsulinism

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1. Abstract

1.1. Aim

Pancreatic pathology is relatively rare in children. We report a child whounderwent near total laparoscopic pancreatectomy.

1.2. Results

An infant underwent near total pancreatectomy for congenitalhyperinsulinism and made an excellent postoperative recovery.

1.3. Conclusion

Laparoscopic near total pancreatectomy is feasible, safe andbeneficial for infants with congenital hyperinsulinism. Every attempt should bemade to preserve the spleen during surgery and these children should beimmunized preoperatively to avoid the risk of post-splenectomy sepsis.

2. Introduction

Persistent hyperinsulinemic hypoglycemia which is also called congenitalhyperinsulinism is the most common cause of recurrent and persistenthypoglycemia in infancy and childhood. The two main causes of recurrent and

persistent hypoglycemia are congenital hyperinsulinism and insulinoma. Inspite of improved radiological techniques, preoperative differentiation of congenital hyperinsulinism and insulinoma in infants and children is difficult [1]. Congenital hyperinsulinism is characterized by abnormal and dysregulatedsecretion of insulin from pancreatic beta-cells. Prompt diagnosis and earlytreatment of congenital hyperinsulinism is important to avoid recurrenthypoglycemia with its effect on the brain and the risk for subsequentneurodevelopmental disorders [2, 3]. The treatment of congenitalhyperinsulinism is medically to start with. Diazoxide and somatostatin (octreotide) are the mainstays of medical treatment for congenitalhyperinsulinism. Nifedipine was also used and reported to be successful [4, 5].Persistent hypoglycemia in spite of medical treatment is an indication forsurgical treatment. The standard treatment for congenital hyperinsulinism isnear total pancreatectomy for the diffuse type which is done openly. Recentlyand as result of advances in minimally invasive surgery especially for infants

and children, laparoscopic near total pancreatectomy was shown to be feasibleand safe in infants and children [6-10].In this report we describe a successful laparoscopic near total pancreatectomyfor an infant with congenital hyperinsulinism outlining the clinical features,diagnosis, treatment and outcome.

3. Case Report

A female newborn, a product of full term normal vaginal delivery to a G2P2L2 mother who had gestational diabetes which was controlled by diet. She washealthy and well but at the age of 3 days she started to have recurrent attacksof hypoglycemia. Her blood glucose level

dropped to 38 mg /dl (2.1 mmole / L).There were no convulsions and her physical examination was normal. She wasstarted on IV 10% dextrose and this was increased to 20% dextrose and enteralfeeding in an attempt to control her hypoglycemia. She required multipledextrose IV boluses and glucagon. On day 8, she was started on hydrocortisone10 mg / kg /dose every 8 hours, Diazoxide 20 mg / kg /day in three divided doses,Octreotide 10 mcg / kg/ dose every 5 hours and hydrochlorothiazide 1 mg / kg /dose every 12 hours. The hydrocortisone was discontinued after few days. Inspite of this she continued to have attacks of hypoglycemia. Her investigationsshowed hypoglycemia with a blood glucose level of 20 mg / dl (1.11 mmole / L),her serum insulin level was elevated (63.11 micro-IU / L), and C-peptide waselevated (7.35 ng / ml). Her growth hormone, liver function testes, renalfunction testes, abdominal and brain ultrasounds were normal. Her genetic testconfirmed an autosomal recessive homozygous form of congenitalhyperinsulinism (diffuse form) (ABCC8 mutation). Our patient was found tobe heterozygous for a pathogenic ABCC8 frameshift variant. Biallelic loss offunction pathogenic variant in ABCC8 cause Congenital hyperinsulinism(MIM256450).

HGVS DESCRIPTION: NM_ 001287174.1.c.4479_4480dupp. (Arg1494Profs*5)Location: GRCh37 (hg19)

Chr11: g.17394335_17394336dupHer father and mother were also found to be heterozygous for a pathogenic

ABCC8 frameshift variant. The risk that their next pregnancy will be affectedby congenital hyperinsulinism is 1 in 4 (25%). She was planned to undergo near total pancreatectomy. Preoperatively she wasgiven PCV 13, HiB and MCV4 vaccines. She underwent near total laparoscopicpancreatectomy with splenic preservation. Postoperatively, she did well andwas started on oral feeds on the second postoperative day. Her postoperative

blood glucose increased to 6.9 then 7.8 and then 10 mmol / L and she wasdischarged home on the 7th postoperative day. Histopathology of the resected specimen showed diffuse islet cell enlargementconsistent with diffuse hyperinsulinism. Microscopic examination revealed apreserved pancreatic lobular architecture. There were nodules of variably sizedand irregularly shaped islets. The percentage it occupied in each lobule varied

from 10-60%. Within these enlarged irregular islets cell nodules, largehyperchromatic endocrine cell nuclei were noted (up to 3x the size of adjacentcell nuclei). Few small ductuloinsular complexes were noted. The findings wereconsistent with diffuse hyperinsulinism (Figures 1a, 1b and 1c). On follow up inthe clinic she was gaining weight, on no medications and maintaining normalblood glucose levels.

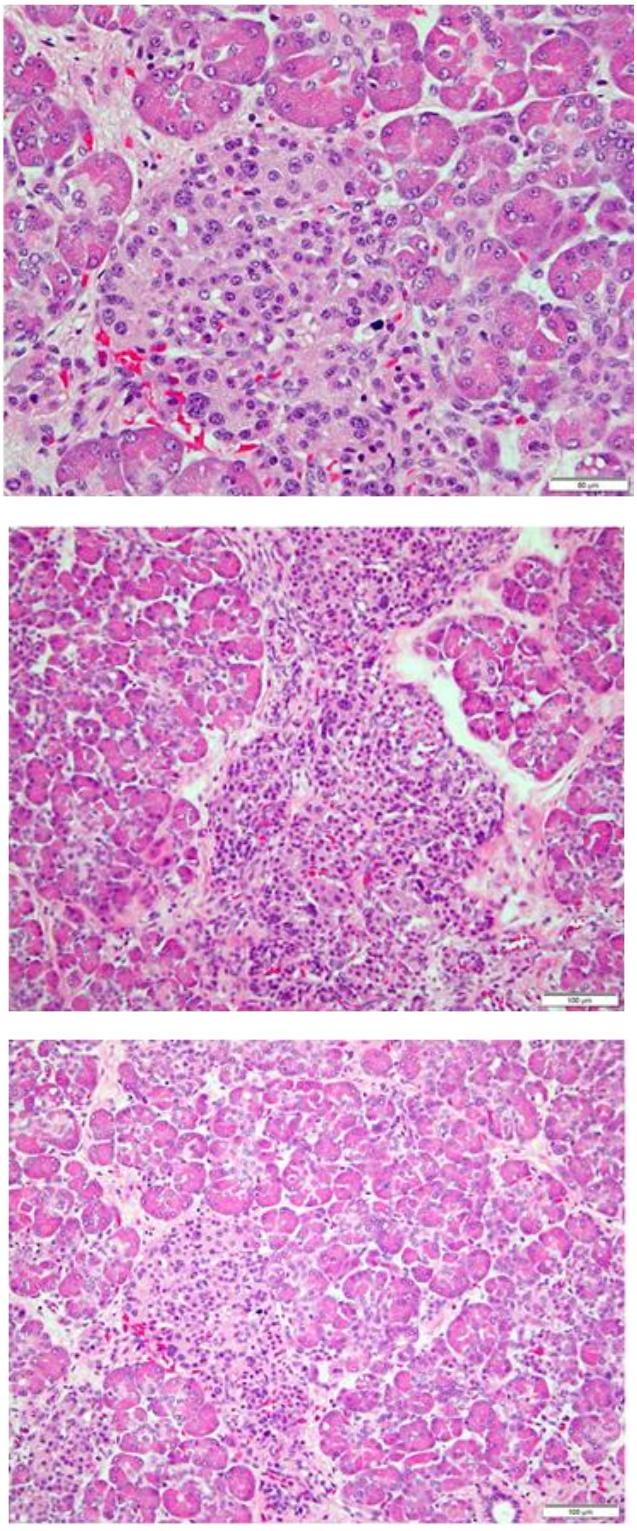

Figure 1a, 1b and 1c: Microscopic examination of the resected pancreas showing a preserved pancreatic lobular architecture. There were nodules of variably sized and irregularly shaped islets. The percentage it occupied in each lobule varied from 10-60%. Within these enlarged irregular islets cell nodules, large hyperchromatic endocrine cell nuclei were noted (up to 3x the size of adjacent cell nuclei). Few small ductuloinsular complexes were noted. The findings were consistent with diffuse hyperinsulinism.

4. Discussion

Congenital hyperinsulinism is a major cause of persistent hyperinsulinemichypoglycemia in newborns and infants. Histologically, congenitalhyperinsulinism is divided into three major subtypes: diffuse, focal, andatypical forms. The diffuse form which is the commonest is inherited as anautosomal recessive (or autosomal dominant). The focal form is inheritedsporadically. The focal form was reported to account for 40–70% of allcongenital hyperinsulinism cases [11,12]. The surgical management of thediffuse form is a near-total pancreatectomy, whereas the focal form can bemanaged with a limited pancreatectomy [13-18). It is importantto differentiate between the two types as near-total pancreatectomy carries a high risk of iatrogenic diabetes, whereas patients with the focal type can becompletely cured by a limited pancreatic resection [19,20]. The distinctionbetween the two types preoperatively is however difficult. There are invasiveand technically difficult techniques to achieve that such as pancreatic-venoussampling or pancreatic arterial calcium stimulation tests but these have notbeen used widely [18,19]. Timo Oton Koski and his colleagues found that it ispossible to identify the focal forms of congenital hyperinsulinism usingnon-invasive fluorine-18-L-dihydroxyphenylalanine ([18F]-DOPA) PET. Thiswill lead to focal accumulation of [18F]-DOPA in the pancreas in those withfocal congenital hyperinsulinism [20].The treatment of congenital hyperinsulinism is medical to start with. Diazoxideand somatostatin (octreotide) are the mainstays of medical treatment ofcongenital hyperinsulinism [4,21,22). Nifedipine is another drug that wasdocumented to be beneficial in cases of congenital hyperinsulinism [4,5].Failure of medical treatment is an indication for surgical resection [3]. Neartotal and partial pancreatectomy are the procedure of choice for diffuse andfocal congenital hyperinsulinism respectively. Patients treated with near totalpancreatectomy are however at risk of developing diabetes during childhood orat puberty [2]. Lovvorn and his colleagues retrospectively reviewed 101children treated for congenital hyperinsulinism who required pancreatectomy(subtotal (< 95%) or near-total (95–98%). They recommended that 95%pancreatectomy be the initial procedure of choice for newborns and infantswith congenital hyperinsulinism [23]. For many years, subtotal or near-totalpancreatectomy was done via the open technique but as result of advancementin minimal invasive surgery, laparoscopic near total pancreatectomy is nowfeasible and safe both in infants and children. Our patient underwent near totallaparoscopic pancreatectomy and made a good recovery and postoperativelyand on follow up she remained euglycemic and on no medications. Thistechnique however, requires experience in laparoscopic surgery.In conclusion, laparoscopic near total pancreatectomy is feasible and safe ininfants and children. It is associated with less morbidity, fast recovery andshorter hospital stay when compared with the open technique. Our patientunderwent laparoscopic near total pancreatectomy for congenitalhyperinsulinism and made an excellent postoperative recovery. Every attemptshould be made to preserve the spleen during laparoscopic pancreatectomy toavoid the risk of post-splenectomy sepsis. These children however, should be immunized prior to surgery.

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