Neonatal Transient Idiopathic Hyperinsulinism: Case Report

Abstract

We report the case of a transient hypoglycemia secondary to idiopathic intermittent hyperinsulinism in a two-day-old newborn. The diagnosis was evoked on clinical data, intermittent hyperinsulinemia, endocrine screening and magnetic resonance imaging MRI brain and pancreatic normal. Neonatal hyperinsulinism intermittent transient idiopathic is a rare cause of neonatal hypoglycemia. Inappropriate secretion of insulin is responsible for unexplained severe hypoglycemia. An early treatment reduces the risk of neurological long-term sequelae.

Keywords: Hypoglycemia; Neonate; Hyperinsulinism; Transient

Case Report

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Radouani MA, Azzaoui S, Kabiri M and Amina Barkat*

**Department of Medicine and Neonatal Resuscitation, National Center for Neonatology and Nutrition, Morocco*

***Corresponding author:** Amina Barkat, Department of Medicine and Neonatal Resuscitation, National Center for Neonatology and Nutrition, Children's Hospital, Ibn Sina hospital, Ibn Rushd bd, 10100 Souissi, Rabat, Morocco, Tel: 0661385108; Email: barakatamina@yahoo.fr

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Introduction

The hyperinsulinism (HI) is a common cause of hypoglycemia in the newborn. Inappropriate insulin secretion by pancreatic beta cells is responsible for severe hypoglycaemia with a high risk of brain damage. It is usually found in hypotrophic newborns and in newborns of diabetic mothers by hyperinsulinemia or secondary to an increase in anti-stress hormones. Syndromic HIs are responsible for permanent hypoglycemia. The diagnosis of idiopathic HI can be done by high insulin and C-peptide in the blood with normal pancreatic and brain imaging and a negative balance metabolic and genetic levels. We report a case of a newborn who has severe idiopathic transient hypoglycemia.

Case Report

This newborn male was two days old admitted for dehydration with hypoglycemia. The mother is 32 years old, primiparous blood type A positive. The pregnancy was without difficulties. There is no parental consanguinity. The delivery was vaginal with immediate cry. Birth weight is 2700g.

Clinical examination showed, weight W = 2500g, size S = 47cm, head circumference HC 34 cm, temperature T = 37 °, heart rate HR = 160 b / m, Blood pressure BP = 55/30, blood glucose capillary GC: 0.13 g / l. The patient is polypneic, hypotonic with a fold dehydration and sunken eyes. The review noted no facial dysmorphia or micropenis and no hepatomegaly. The remainder of the physical examination was normal.

The newborn had received a saline remplissage and infusion of glucose solution 10% with antibiotics (3rd generation cephalosporins C3G parenterally). I1 is immediately found hypoglycemia requiring a significant increase in carbohydrate intake intravenous (12 g / kg / day at day 3, 16 g / kg / jh on day 4 and 18 g / kg / d after 5 days). The blood levels of insulin, C-peptide, 17OH progesterone, cortisol, ACTH adrénocortiotrophine and growth hormone (GH) were without abnormalities. Adrenal ultrasound came back normal. Brain and pancreatic MRI was without anomalies.

Chromatography of blood amino acids, chromatography urine organic acids, plasma lactate and plasma ammonia were normal.

Enteral nutrition was started at day 7 by continuous gavage (milk Pre) with good tolerance for stopping the perfusion was stopped at day 13. The daily carbohydrate intake is normal to 36 days of life. Subsequent developments are satisfactory.

Discussion

Congenital hyperinsulinism is a rare disease, its incidence is estimated at one in 50,000 in Western countries [1] and one in 3000 births in some populations of the Middle East [2]. HI newborn appears in the first 72 hours of life. The newborn tolerates episodes of hypoglycemia, may occur in the first hours of life through the production of ketone bodies in the brain used as an energy source. He can, unlike the premature and hypotrophic neonate, mobilize its reserves of hepatic glycogen and achieve gluconeogenesis from proteins and lipids [3]. The need for glucose in the premature newborn is 5 to 8 mg / kg / min and that of full term newborns 3 to5 mg / kg / min, but in case of hyperinsulinemia, glucose administered at these doses is insufficient to maintain normal blood sugar levels.

Hypoglycemia is severe, revealed by a convulsion in half the cases, but it can also be revealing coincidentally. It immediately requires glucose intake very high (17 mg / kg per minute on average) who quickly put a central catheter. Such hypoglycemia should immediately evoke a HI and to administer glucagon by intravenous infusion or subcutaneous route to prevent neurological sequelae. A hepatomegaly is common [4].

Hypoglycaemia revealed later in the first months of life are more moderate, which explains the delay in diagnosis. Seizures reveal these hypoglycemia in 50% of cases [5]. The requirements are lower (12-13 mg / kg per minute) and rarely require a central catheter.

The first step to analyze this hypoglycemia is to distinguish hyperinsulinism of newborn of diabetic mother, Beckwith-Wiedeman syndrom, hypotrophy with poverty of subcutaneous adipose tissue (Donohue syndrom). A drug investigation in

the mother eliminates hypoglycemia due to hypoglycemiant drugs, beta-blockers or diuretics [6]. There is also uncontrolled hypoglycemia by medical treatment that may require surgery to search for nesidioblastosis. Other hormonal disorders such as growth hormone deficiency and / or cortisol are also mentioned. Another mechanism to evoke is that of insulin hyperactivity inhibition of gluconeogenesis induced by glucose infusion exclusive high flow in the absence of other substrates for gluconeogenesis [2].

Hypoglycemia in our patient was seen in the first 48 hours of life, revealed by neonatal seizures requiring high intakes of glucose. The etiologic appraisal was normal and the evolution was favorable at the first month of life, what makes us evoke an intermittent transient idiopathic hyperinsulinism. This entity is common in low birth weight, newborns of diabetic mothers but also in case perinatal asphyxia which was not the case in our patient. The mechanism of this type's HI is incompletely explained. Hypoxia, acidosis and alterations of blood flow and pressure fetal can stimulate the release of catecholamines that engage liver glycogen stores, in addition, hypoxia increases the rate of anaerobic glycolysis which accelerates glucose utilization. All this depleted glycogen fetal stores and places the child in a situation of risk of developing hypoglycemia.

Conclusion

The prognosis of congenital hyperinsulinism is reserved as a positive diagnosis is often delayed and it causes hypoglycemia resistant to medical treatment. Neurological and metabolic sequelae are important. The diagnosis of location is very important because it determines the surgical procedures and genetic counseling.

References

1. Bruining GJ (1990) Recent Advances in hyperinsulinism and the pathogenesis of diabete mellitus. Cur Opin Pediatr 2: 758-765.
2. Glaser B, Phillip M, Carmi R, Lieberman E, Landau H (1990) Persistent hyperinsulinemic hypoglycemia of infancy ('nesidioblastosis'). Autosomal recessive inheritance in seven pedigrees. Am J Med Genet 37(4): 511-515.
3. Kelly A, Ng D, Ferry RJ, Grimberg A, Koo-McCoy S, et al. (2001) Acute insulin responses to leucine in children with the hyperinsulinism/ hyperammonemia syndrome. J Clin Endocrinol Metab 86(8): 3724-3728.
4. De Leon DD, Stanley CA (2007) Mechanisms of disease: advances in diagnosis and treatment of hyperinsulinism in neonates. Nat Clin Pract Endocrinol Metab 3(1): 57-68.
5. Steinkrauss L, Lipman TH, Hendell CD, Gerdes M, Thornton PS, et al. (2005) Effects of hypoglycemia on developmental outcome in children with congenital hyperinsulinism. J Pediatr Nurs 20(2): 109-118.
6. Munns CF, Batch JA (2001) Hyperinsulinism And Beckwith-Wiedemann syndrome. Arch Dis Child Fetal Neonatal Ed 84(1): 67-69.

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