

CONGENITAL PANHYPOPITUITARISM ASSOCIATED WITH IMPAIRED LIVER FUNCTION TESTS AND CONGENITAL HEART DISEASE

Z. Khalili-Matinzadeh*

Department of Pediatrics, School of Medicine, Tehran University of Medical Sciences, Tehran,
Iran

Abstract- The term congenital hypopituitarism defines deficiency of all of the pituitary hormones. Hypoglycemia and microphallus (in males) are common findings, and some infants have shown evidence of the neonatal hepatitis syndrome. We report a case of congenital panhypopituitarism with deficiency of six major hormones and association with severe hypoglycemia, impaired liver function tests and congenital heart disease.

Acta Medica Iranica, 44(2): 155-156; 2006

©2006 Tehran University of Medical Sciences. All rights reserved.

Key words: Congenital hypopituitarism, hormones, perinatal

INTRODUCTION

Congenital panhypopituitarism is a rare condition associated with deficiency of all six major hormones (LH, FSH, GH, thyrotropin, corticotrophin, and prolactin) (1). Hypopituitary states are associated with a deficiency of growth hormone with or without a deficiency of other pituitary hormones (2). Affected infants are being recognized by early hypoglycemia (1, 3-5) and microphallus (in males) (1, 2). Neonatal hepatitis syndrome (1, 6) and optic nerve hypoplasia (1, 2, 7) are common but definitive diagnosis rests on demonstration of absence of the major hormones. MRI is indicated in all patients with hypopituitarism (2, 8).

Here we report a case of congenital panhypopituitarism associated with impaired liver function tests and congenital heart disease.

Received: 7 Aug. 2004, Revised: 26 Jan. 2005, Accepted: 13 Apr. 2005

*** Corresponding Author:**

Z. Khalili-Matinzadeh, Department of Pediatrics, School of Medicine, Tehran University of Medical Sciences, Tehran, Iran
Tel: +98 21 88.64030,
Fax: +98 21 88037561
E-mail: Matinzadeh@bmsu.ac.ir

CASE REPORT

A ten day old female neonate was referred to our hospital due to generalized edema, respiratory distress and apneic spells.

She was born by cesarian section (due to fetal distress) with birth weight of 2550 gr. Her mother was 19 year old and her parents were first cousins. Hypotonia and hyporeflexia were apparent signs so sepsis workup was done and antibiotic therapy was started. Severe hypoglycemia (blood sugar of 12 mg/dl) was treated with 12.5% glucose, diazoxide and hydrocortisone. Echocardiography was done due to grade II/VI systolic murmur that confirmed left ventricular hypertrophy, mild pulmonary hypertension and small ventricular septal defect. Renal problems, hyponatremia (blood sodium, 124) and hypoglycemia resolved with convenient management but hypotonia continued. Brain magnetic resonance imaging (MRI) was normal. Optic nerve was normal in ophthalmologic exam. Liver transaminases were high. Thyroid function tests were performed. The results of laboratory tests are shown in table 1.

Table 1. Laboratory data

	Patient data	Normal range
T4 (µg/dl)	0.1	9-18
TSH (mIu/L)	0.1	1.7-9.1
Cortisol (8 am, nmol/l)	118	260-720
GH (mIu/L)	0.74	0-20
FSH (Iu/L)	0.05	1-4.2
LH (Iu/L)	0.08	-
Prolactin	0.1	3.7-40
AST(u/l)	176	5-40
ALT (u/l)	166	5-40
Bilirubin* (mg/dl)	9 (T), 1 (D)	

* On tenth day.

Abbreviations: GH, growth hormone; T, total; D, direct.

Low level of T4 with low TSH, conducted us to congenital hypopituitarism. Multiple anterior pituitary hormone deficiencies were found and finally confirmed by provocative tests.

Treatment with levothyroxine, growth hormone (after provocative test) and hydrocortisone was started. Now she is 5 month old with normal growth and development. T₄ = 13 µg/dl and BS = 90 mg/dl.

DISCUSSION

Hypopituitarism may occur after head injury and perinatal trauma (*e.g.* traction of delivery, anoxia and hemorrhagic infarction) (2, 9, 10). Although the increased incidence of breech presentation and birth trauma with neonatal asphyxia in congenital hypopituitarism has led some to suggest a causative role for these occurrences, but perinatal difficulties are to be the consequence rather than the cause of the abnormalities (1). The cause is unknown but young maternal age and nulliparity are strongly associated factors, which were present in our patient. Affected newborns are usually of normal size and weight at birth. They often present as neonatal emergencies such as apnea, cyanosis or severe hypoglycemia without hyperinsulinism, hypotonia and seizure. This neonate presented with these complications. Prolonged jaundice is common and some infants show evidence of the neonatal hepatitis syndrome but the relationship is obscure (2). Unilateral or

bilateral optic nerve hypoplasia is common. Definitive diagnosis rests on demonstration of absent or low levels of hormones. Brain MRI may show an empty sella turcica although it may be normal. Treatment should be started as soon as possible and replacement should be directed at hormonal deficiencies.

REFERENCES

1. Larsen PR, editor. Williams textbook of endocrinology. 9th edition. Philadelphia: W B Saunders; 1998. p. 1964-1965.
2. Behrman RE, Kliegman R, Jenson HB, editors. Nelson textbook of pediatrics. 16th edition. Philadelphia: W B Saunders; 2000. p. 1674-1680.
3. Fellner F, Triebe S, Hensen J, Baum U, Schmitt R, Flottemesch J, Bautz W. [Neuro-MR-findings in primary panhypopituitarism]. Rontgenpraxis. 1998; 51(7):243-247. German.
4. Kauschansky A, Genel M, Smith GJ. Congenital hypopituitarism in female infants. Its association with hypoglycemia and hypothyroidism. Am J Dis Child. 1979 Feb; 133(2):165-169.
5. Biggemann B. [Endocrine-induced forms of hypoglycemia]. Monatsschr Kinderheilkd. 1988 Sep; 136(9):612-615. German.
6. Spray CH, Mckiernan P, Waldron KE, Shaw N, Kirk J, Kelly DA. Investigation and outcome of neonatal hepatitis in infants with hypopituitarism. Acta Paediatr. 2000 Aug; 89(8):951-954.
7. Brodsky MC, Phillips PH. Optic nerve hypoplasia and congenital hypopituitarism. J Pediatr. 2000 Jun; 136(6):850.
8. Martinez M, Martinez A, Martinez Ramonde T, Jorge Mendez S, Vidal Vazquez O, Cordido F. [Utility of magnetic resonance in the etiologic diagnosis of hypopituitarism]. An Med Interna. 1996 Dec; 13(12):584-588. Spanish.
9. Gacs G. [Perinatal disorders in the etiology of hypopituitarism]. Orv Hetil. 1987 Feb 1; 128(5):243-246. Hungarian.
10. Craft WH, Underwood LE, Van Wyk JJ. High incidence of perinatal insult in children with idiopathic hypopituitarism. J Pediatr. 1980 Mar; 96(3 Pt 1):397-402.