Case Report

Congenital hypopituitarism and renal failure

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ABSTRACT

Congenital hypopituitarism is potentially fatal in the newborn period but treatable if the diagnosis is made early. We report a neonate who presented with hypothermia and severe hypoglycemia. He also had undescended testis and micropenis. Initial screening revealed panhypopituitarism, which was corrected promptly. He developed renal failure due to initial cardiovascular compromise related to hypotension but recovered quickly with standard management. Magnetic resonance imaging revealed absent stalk of anterior pituitary.

Key words: Congenital hypopituitarism, hypoglycemia, hypothermia, renal failure

Introduction

While hypopituitarism is common, congenital hypopituitarism is rare. Even rarer is the successful diagnosis and management of this condition in the neonatal period, especially when accompanied by other comorbid conditions. This case highlights the clinical presentation and management of such a neonate.

Case Report

A male infant weighing 3.88 kg (50th–75th centile) was born at 42 + 1 weeks of gestation by emergency lower segment caesarean section performed for failed induction. His mother had active oral herpes at the time of delivery. The maternal medical history, family history, and antenatal history were unremarkable, including antenatal scans and maternal serology. He was born in good condition with a normal Apgar score and was transferred to the postnatal ward with his mother. His head circumference was 36.5 cm (75th centile) and length was 52.8 cm (25th centile). At 4 h of age, he was noted to be hypothermic (34.2°C) and hypoglycemic (blood glucose 1.0 mmol/l).

On examination, he was noted to have micropenis (stretched penile length 1.25 cm) and left testis was not palpable in the scrotum. He was hypotonic and lethargic. He was also noted to have a mild vesicular rash on his legs. No other congenital abnormalities were noted. A working diagnosis of congenital hypopituitarism (CH) with a possible herpes infection was made. He had a hypoglycemia screening and needed two dextrose boluses to normalize his blood glucose. He was placed in an incubator, to correct hypothermia, which slowly responded to high ambient temperature. He was noted to have hypotension and tachycardia for which he received two normal saline boluses, which improved his cardiovascular status. He was started on maintenance fluids and first-line antibiotics along with intravenous aciclovir. These antibiotics were later stopped as his bacterial and viral cultures from blood and vesicular fluid were negative.

By the second day of life, he had developed oliguria while being normotensive. He was unresponsive to fluid bolus and diuretics. His urea and creatinine increased rapidly, further suggesting renal failure. This was thought to be secondary to the initial cardiovascular compromise and hypotensive insult secondary to low cortisol levels. This was managed with standard renal failure guidelines including fluid restriction and careful monitoring of electrolyte

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status. By seventh day of life, his renal function started improving and he was gradually established on feeds and was normalized.

His initial hypoglycemia screening on admission revealed very low cortisol (<7 nmol/l), growth hormone (<0.05 μg/l), and low free T4 (11.4 pmol/l) concentrations. Thyroid function tests later revealed absent thyroid-stimulating hormone. A replacement dose of hydrocortisone and L-thyroxin was started. This improved his clinical status generally as he became more active with normal tone. The doses of hydrocortisone and L-thyroxin were optimized to 2 mg/kg four times a day and 50 μg once daily, respectively.

An ophthalmological review suggested a possibility of right hypoplastic optic disc, which was ruled out by magnetic resonance imaging (MRI). The MRI of brain revealed an anterior pituitary gland with absent pituitary stalk. The posterior pituitary gland was ectopic. There was a cavum septum pellucidum that was partially absent on the inferior aspect. No other structural abnormality of brain was revealed. Optic nerve and chiasm were normal. Parents were taught 'sick day rules'. They were also taught about the administration of intramuscular hydrocortisone and buccal administration of Hypostop. He was discharged under the care of a pediatric endocrinologist. Referrals were made for the surgical management of undescended testis. A follow-up at 5 weeks of age revealed normal growth and development with normal tone and activity.

Discussion

Pituitary deficiency is the second most frequent cause of persistent neonatal hypoglycemia, the first one being congenital hyperinsulinism. It is a potentially lethal but eminently treatable condition. The associated clinical features may include midline cleft lip or palate, single central incisor, or optic nerve hypoplasia. Male infants can have micropenis due to gonadotrophin deficiency.[1] Micropenis is an important sign in neonates since it may be the only clue to the diagnosis of panhypopituitarism.[2]

The diagnosis of hypopituitarism must be based on clinical grounds, especially when hypoglycemia, prolonged jaundice, micropenis, or midline alterations are found in the neonatal period.[3] Neonatal presentation may be mistaken for sepsis, which can present with similar clinical features of hypothermia, hypoglycemia, lethargy, and poor feeding. The initial investigation and management should be aimed at stabilizing the clinical status of the baby. But any suspicion in the presence of any or all of the above clinical features should be a trigger for the investigation of hypopituitarism.

By far anterior pituitary is the most common lobe to be involved, but both anterior and posterior pituitary may be affected. Adrenocorticotrophic hormone (ACTH), thyroid stimulating hormone (TSH), follicle stimulating hormone (FSH), luteinizing hormone (LH), growth hormone (GH), and prolactin may be deficient along with partial or complete absence of vasopressin. This may cause diabetes insipidus, although our patient did not have this condition. Hypothalamus may also be involved, although that is less common. Provocative testing may be required if the clinical features and biochemical abnormalities are subtle. Children with CH need to be followed up regularly for their ongoing management. It has been suggested that they can have an IQ that is below average when compared with the population norm and a reduced performance IQ when compared with sibling controls.[4]

In our patient, the presentation of hypoglycemia and hypothermia was very suggestive of CH in the presence of micropenis. Early diagnosis meant that treatment was instituted early. The onset of renal failure suggested that any further delay in the treatment could have been life-threatening. This case also highlighted that CH can be associated with a variety of metabolic disturbances, and renal failure is one of them.

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