Persistent neonatal hypoglycaemia as a result of hypoplastic pituitary gland

Introduction

Hypoglycaemia is a common neonatal problem. In cases of refractory hypoglycaemia further investigations are required. The authors describe a neonate who presented with refractory hypoglycaemia as a result of prenatal hypopituitarism based on measurement of hormone levels, rapid response to replacement therapy and magnetic resonance imaging (MRI) of the brain.

Discussion

A Medline search between 1966–2004 reported a number of cases of congenital hypopituitarism in newborn infants (Kosaki et al, 1991; Van Hauthem et al, 1992; Kato et al, 1995; Geffner, 2002). However, this was the first case reported from Malaysia.

Although neonatal hypoglycaemia is a common problem, occasionally this may be a manifestation of congenital hypopituitarism (Geffner, 2002). If undiagnosed, infants with congenital hypopituitarism may develop life-threatening problems such as shock, hypoglycaemia and later growth failure. Therefore prompt investigations have to be instituted in infants with persistent hypoglycaemia.

In cases defined by a deficiency of anterior pituitary hormones the authors recommend an MRI scan of the brain to confirm the absence or hypoplasia of either an isolated anterior pituitary or the entire pituitary gland. BJHM

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Case Report

An Indonesian female infant developed hypoglycaemia shortly after delivery by lower segment caesarean section. Following admission to the neonatal intensive care unit, she continued to have episodes of hypoglycaemia despite adequate glucose intake of up to a maximum of 12 mg/kg/minute.

During the episodes of hypoglycaemia, repeated measurement of her blood samples revealed persistently low levels of serum insulin of <2 IU/ml, serum free thyroxine of 4.78 pmol/litre, thyroid stimulating hormone of 0.31 uIU/ml and growth hormone of 0.27 mIU/litre, all suggesting a diagnosis of hypopituitarism.

Empirical replacement therapy with hydrocortisone (10 mg four times daily) was started at 26 days. However, her hypoglycaemic attacks ceased only with commencement of additional hormone replacement therapy of growth hormone (0.1 mg once daily for 6 days per week) and thyroxine (50 μg once daily). The hydrocortisone dose was gradually tailed to a maintenance dose of 2.5 mg twice daily. Stimulatory tests such as adrenocorticotropic hormone and luteinizing hormone-releasing hormone tests and posterior pituitary function were not performed at that time as the infant developed *Klebsiella pneumoniae* and coagulase negative staphylococcus sepsis which may have affected the test results.

A magnetic resonance imaging (MRI) scan of her brain at 1 month of age showed a hypoplastic pituitary gland with a shallow pituitary fossa (*Figure 1*). The posterior lobe of the gland was undescended, lying ectopic at the base of the third ventricle with an absent pituitary stalk (*Figure 2*). The rest of the MRI was normal with no associated anomalies of the midline forebrain structures. No abnormalities were detected in detailed opthalmological examination of her eyes excluding the possibility of septo-optic dysplasia.

When followed up at 8 months of age, she remained well with improved hormonal profiles and normal levels of serum urea and electrolytes. Her weight, height and occipito-frontal circumference were all below the third percentile.

Figure 1. Magnetic resonance imaging of the brain in the sagittal view showing a hypoplastic pituitary gland with a shallow pituitary fossa.

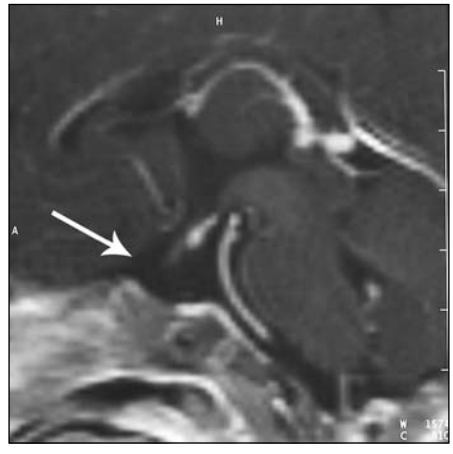
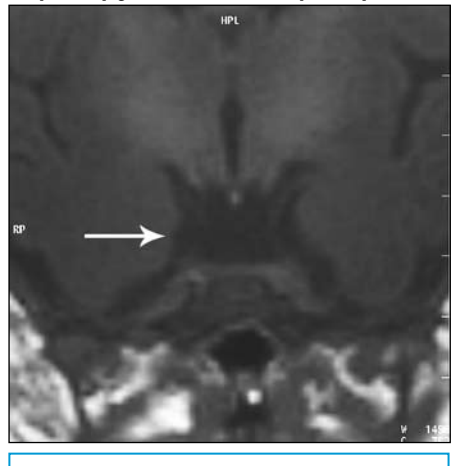


Figure 2. Magnetic resonance imaging of the brain in the coronal view showing the posterior lobe of the pituitary gland with an absent pituitary stalk.



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