

CASE REPORT

Transient Adenocorticotrophic Hormone deficiency in an infant

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ABSTRACT

Background: Isolated Adenocorticotropin Hormone (ACTH) deficiency is a rare disorder, characterized by secondary adrenal insufficiency with a low or absent cortisol production, normal secretion of other pituitary hormones and absence of structural pituitary defects.

Case summary: The patient was the product of a 28 weeks gestation, clomiphene induced to a 40-year-old, gestational diabetic mother. He was ventilated for 30 days, and was diagnosed to have grade IV Retina of Prematurity (ROP). He suffered from recurrent attacks of hypoglycemic, blood sugar of 1.6 mmol/L, low serum cortisol 65 nmol/L (normal; 150-630), and ACTH of 1.5 pmol/L (normal; 1.6-13.9), with suppressed serum insulin and normal thyroid, growth hormone and gonadal functions. Magnetic Resonance Imaging (MRI) was unremarkable. He was started on hydrocortisone 2.5 mg twice daily for two years, which was then slowly tapered and stopped. Later Serum cortisol was 175 nmol/L, and ACTH of 5.5 pmol/L with a normal shot ACTH stimulation test.

Conclusions: This case of a premature baby who presented with recurrent hypoglycemia had an isolated ACTH deficiency that proved to be transient. The pediatricians need to be aware of the existence of such condition.

Key Words: Adenocorticotropin Hormone, Isolated, Hypoglycemic, Transient

1. INTRODUCTION

Isolated Adenocorticotropin Hormone (ACTH) deficiency is a rare disorder that characterized by secondary adrenal insufficiency with low or absent cortisol production, normal secretion of other pituitary hormone and absence of structural pituitary defects.^[1] The defect is usually at the pituitary level but can be at the hypothalamus.^[2] Most cases are secondary to autoimmune process such lymphocytic hypophysitis.^[3,4] It may also occur after traumatic head injury^[5] and pituitary irradiation.^[6] Genetic cases are encountered in neonatal and childhood isolated ACTH deficiency.^[7] It might be associ-

ated with transient growth hormone (GH deficiency)^[8] or thyroid disorders.^[9]

In this report, we describe a premature baby who presented with recurrent hypoglycemia and proved to have transient isolated ACTH deficiency. The practicing pediatrician should be aware of the existence of such disorder.

2. CASE REPORT

The patient was the product of a 28 weeks gestation, clomiphene induced to a 40-year-old mother, who was suffering from gestational diabetes managed with diet. She

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was para 1+0. Spontaneous vaginal delivery with a birth weight of one kilogram. He was ventilated for 30 days and diagnosed with grade IV retina of Prematurity (ROP) which required laser therapy. There was no intraventricular hemorrhage. The patient suffered from recurrent attacks of hypoglycemia. At a blood sugar of 1.6 mmol/L, serum concentration of cortisol was low at 65 nmol/L (normal; 150-630), with also low ACTH level of 1.5 pmol/L (normal; 1.6-13.9). Serum concentration of growth hormone (GH) was 15, 12 and 8.5 ng/ml (normal; >10) serum concentration of insulin was 6.3 m μ /ml (normal; 5-21). Normal thyroid function tests; TSH of 4 mU/L (normal; 0.5-5) and FT4 of 15 pmol/L (normal; 10-30) Normal hypothalamic pituitary gonadal function; LH of 3.9 μ /L (normal; 0.5-3) and FSH of 1.3 U/L (normal; 1-5). Unremarkable MS/MS and VLCFA and urine organic acids. Magnetic Resonance Imaging (MRI) was unremarkable, see Figure 1.

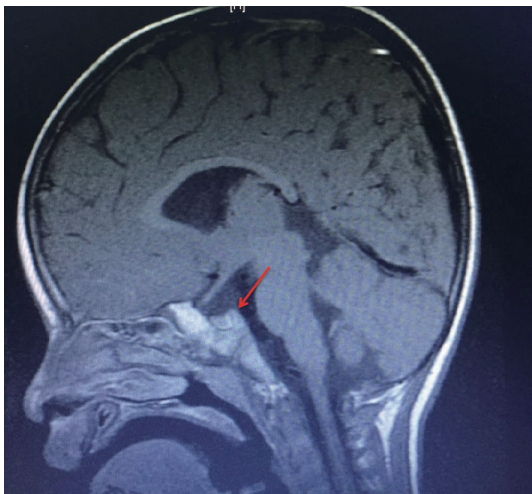


Figure 1. Sagittal view of pituitary Magnetic Resonance Imaging (MRI), showing normal pituitary gland (arrow)

He was started on hydrocortisone 2.5 mg twice daily for two years that was then slowly tapered and stopped. Currently at the age of seven years his adrenal function was normal, serum concentration level was 175 nmol/L and serum ACTH of 5.5 pmol/L, with a normal response to short ACTH stimulation test (see Table 1). He remains well and growing normally and asymptomatic.

3. DISCUSSION

Adrenal insufficiency is caused by either primary adrenal failure, mostly due to autoimmune adrenalism or hypothalamic pituitary adrenal impairment of the corticotrophic axis, predominantly due to pituitary or hypothalamic disorders. It is rare, but life threatening when overlooked.^[10] The diagnosis of isolated ACTH deficiency was based on clinical symptoms

of hypoglycemic and low serum concentration of cortisol and ACTH, while the remaining pituitary hormones were normal. The pituitary hypothalamic structure was normal on MRI.^[1,2] The patient improved with hydrocortisone therapy.

Table 1. ACTH stimulation test

Adrenal function (Base-line Results)	ACTH (1.6-13.9 pmol/L) 5.5 pmol/L	Cortisol (150-630 nmol/L) 175 nmol/L
Post Stimulation Cortisol	30 min 480	60 min 750

The present case report represent a patient with concurrent isolated ACTH deficiency in a premature infant who developed retina of prematurity (ROP) which required laser therapy. This may have been an acute stress related a disease of the elderly polymyalgia rheumatica (PMR), was associated with transient ACTH, as well as in the elderly, critically ill patient.^[11,12] Pathogenic significance of adrenal steroid status in relation to inflammatory cytokines (Interleukemia). Systemic inflammation in common critical illness.^[11,12] The concept of depressed adrenal function associated with the clinical picture of shock and systemic inflammation which can lead to suppression of ACTH synthesis or release and suppression of adrenal cortical synthesis is well known.^[12,13]

The possible pathophysiologic mechanism leading to this is poorly understood. A combination of factors are likely involved. Direct damage to the hypothalamic-pituitary-adrenal (HPA) axis components, ischemic injury to the metabolically active adenocortical cells. Also the patient's own immune and inflammatory response appears to play a role.

Prostaglandin (PGE) effects on the hypothalamic pituitary adrenal (HPA) axis activation induced by the inflammatory cytokines has long been suggested.^[14-16] However, the importance and identity of the PG mediated process have remained to be identified.

4. CONCLUSION

This case of premature baby who presented with recurrent attacks of hypoglycemia and isolated ACTH deficiency which proved to be transient should alert the practicing pediatrician to the co-existence of such disorders and try to find an explanation for the possible mechanism.

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CONFLICTS OF INTEREST DISCLOSURE

The authors declare no conflicts of interest.

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