

Addison's disease masquerading hyperemesis gravidarum

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Abstract

Addison's disease is primary hypoadrenalism. It results from progressive destruction of the adrenals. We present a literature review and the case of a 24-year old lady presenting with repeated hospital admissions for 'hyperemesis gravidarum' since 10 weeks of POA. On clinical examination she was found to have a very low BMI with mucous membrane pigmentation and no other significant clinical findings. Biochemical tests confirmed the diagnosis of Addison's disease.

Key words: Addison's disease, hyperemesis gravidarum, vomiting, pregnancy.

Introduction

Addison's disease is primary hypoadrenalism¹. In this condition there is destruction of the adrenal cortex². Glucocorticoid, mineralocorticoid and sex steroid production from the adrenals are therefore reduced. Reduced cortisol levels lead, through feedback, to increased CRH and ACTH production³.

Case report

The patient was 24-year old lady gravida 3 para¹. Her 2 previous pregnancies were first trimester miscarriages. She had 9 hospital admissions from 10 weeks of POA onwards every 2-3 weeks apart until her delivery at 37 weeks of POA. She always presented with excessive vomiting and was initially managed as hyperemesis gravidarum at the local hospital. She was referred to Teaching Hospital, Peradeniya at POA of 32 weeks due to non response to treatment. She did not give a previous history of a similar illness at any stage of her life. Her medical history and family history did not reveal any significant illness. On examination

her BMI was 14.6 kg/m² with a body weight of 33 kg and a height of 1.5 m. She had slight pigmentation of her oral cavity with no cutaneous hyperpigmentation. Blood pressure was 110/70 mmHg. Serum sodium concentrations were between 125-135 mmol/l, serum potassium concentrations between 4.0-4.8 mmol/l, BU 3-3.5 mmol/l. Blood picture showed a normochromic anaemia with a haemoglobin of 10.2 g/dl. White blood cell and differential count was normal. A short ACTH stimulation test was carried out and Addison's disease was diagnosed and treatment initiated. Her basal cortisol level was 34.7µg/dl and following ACTH stimulation it was 40µg/dl. A normal individual should have a rise of more than 2.5 times (usually 3-5 times) the basal cortisol value. The foetus was also growth restricted and managed accordingly. She went into spontaneous labour at 37 weeks and delivered a baby weighing 1800 g. Postpartum period was uneventful.

Discussion

Addison's disease - Thomas Addison described this condition in his classic monograph published in 1855¹. The original description of Addison's disease - "general languor and debility, feebleness of the heart's action, irritability of the stomach and peculiar changes of the colour of the skin" - summarizes the dominant clinical features². This is a rare condition with an estimated incidence in the developed world of 0.8 cases per 100,000 and with an incidence of 3-4/million/year³. This disease may occur at any age and affect both sexes equally².

Addison's disease results from progressive destruction of the adrenals which may involve >90% of the glands before adrenal insufficiency appears². Worldwide, infectious diseases are the most common cause. Leading causes include tuberculosis, fungal infections (histoplasmosis, cryptococcosis) and cytomegalovirus. Adrenal failure may occur in AIDS¹. Autoimmune adrenalitis results from the destruction of the adrenal cortex by organspecific autoantibodies, with 21-hydroxylase as the common antigen³.

Adrenocortical insufficiency is characterized by onset of fatigability, weakness, anorexia, nausea and vomiting, weight loss, cutaneous and mucosal pigmentation, hypotension and occasionally hypoglycaemia. Depending on the duration and

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degree of adrenal hypofunction the manifestation vary from mild chronic fatigue to fulminating shock².

The diagnosis of Addison's disease in pregnancy is difficult, because so many of the features of Addison's disease may be associated with normal pregnancy (vomiting, syncope, weakness, hyperpigmentation). However, persistence of nausea and vomiting after 20 weeks' gestation and weight loss should be considered abnormal. The diagnosis will be made on the basis of high levels of endogenous ACTH and low plasma cortisol levels which do not rise 30 min after the patient is given intramuscular synacthen 0.25 mg (tetracosactide). Fortunately, most patients with undiagnosed Addison's disease usually tolerate pregnancy well. The diagnosis is often made when they suffer Addisonian collapse after delivery or following an obstetric catastrophe such as abortion⁴.

Routine biochemical profile will show hyponatraemia in 90% and hyperkalaemia in 65%, and some may show a high urea level. Mild to moderate hypercalcaemia occurs in 10-20% of patients. Blood glucose may be low, with symptomatic hypoglycaemia. Blood picture may show a normocytic anaemia, relative lymphocytosis, and moderate eosinophilia³.

Diagnosis is by short ACTH stimulation test. Absent or impaired cortisol response confirms the presence of hypoadrenalism but does not differentiate Addison's disease from ACTH deficiency or iatrogenic suppression by steroid medication³.

Measurement of serum ACTH may also help to distinguish primary adrenal insufficiency from hypopituitarism. Low baseline serum cortisol levels, coupled with ACTH levels greater than 250 pg/ml, confirm the diagnosis⁵.

Pregnancy usually proceeds normally in treated patients. Maintenance replacement of adrenocortical

hormones is provided by cortisone acetate 25 mg orally mane and 12.5 mg vesper. An alternative is prednisolone 5 mg mane, 2.5 mg vesper. Mineralocorticoid deficiency is treated with fludrocortisone acetate 0.05-0.1 mg daily. Stress doses of glucocorticoids should be given during labour and delivery. Women with undiagnosed Addison's disease may have a crisis during puerperium. Symptoms include nausea, vomiting, profound epigastric pain accompanied by hypothermia and hypotension. Treatment consists of glucocorticoids and fluid replacement. I.V hydrocortisone is given at doses of 100 mg repeated doses every 6 hours for up to several days. Mineralocorticoid replacement is indicated in cases of refractory hypotension or hyperkalaemia⁵.

Patients with adrenal insufficiency should wear an identifying bracelet.

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