Idiopathic Addison's disease

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IN PAST YEARS, tuberculosis was the commonest cause of Addison's disease (Guttman, 1930). In Singapore, even though tuberculosis is common, Addison's disease, whether due to tuberculous destruction of the adrenals or idiopathic atrophy, is rare. Most cases of Addison's disease seen in Western countries nowadays is due to idiopathic atrophy of the adrenal glands (Friedman, 1948). In these cases, there is an increased prevalence of antibodies against adrenal gland tissue (Blizzard and Kyle, 1963). These antibodies are in many cases associated with antibodies against other organ-specific antigens, presumably due to the same underlying autoimmune disorder (Irvine et al., 1968). In this paper, we report a case of idiopathic Addison's disease associated with primary amenorrhoea and antibodies against the thyroid gland.

Case Report

The patient, a 45-year-old unmarried woman, was admitted on 9th March, 1971 with a 4-month history of progressive darkening of the skin. She also noticed that she was getting weaker, was mildly anorexic and had lost some weight. The symptoms were worse in the last few days before admission and she felt faint after exerting herself.

Further questioning revealed a history of primary amenorrhoea.

On examination, the patient had generalised hyperpigmentation, especially in the pressure areas. The blood pressure was 100/80, mm. Hg. and the pulse rate 72 beats a minute. Breast tissue was poorly developed; axillary and pubic hair was scanty. The uterus and cervix were normal and the adnexae were not palpable. A buccal smear and chromosomal analysis on a sample of peripheral blood revealed a normal karyotype.

A chest radiogram revealed right lower lobe pneumonia which required a course of tetracycline therapy. An abdominal radiogram revealed no evidence of adrenal calcification. Haemoglobin was 11.9 g% and the total leucocyte count 6,700/c.mm. with 63% polymorphs, 25% lymphocytes, 3% monocytes and 9% eosinophils. No ova or cysts were detected in three stool specimens. Fasting blood sugar was 85 mg%. Plasma cortisol levels at midnight and 8.00 a.m. were both 10 mcg%. A 24-hour urine collection (vol. 1550 ml.) for 17-ketosteroids and 17-OH corticosteroids gave values of 0.5 mg and 1.0 mg, respectively. A 1-hour A.C.T.H. stimulation test (Maynard et al., 1966) was performed. At 9.00 a.m., the plasma cortisol level was 9 mcg%. At 10.00 a.m., 1 hour after 25 units of A.C.T.H. were given intramuscularly, the cortisol level did not rise but remained at 9 mcg%. Soffer's test revealed poor excretion of a water load (170 ml. urine was collected over a 5-hour period after an initial loading dose of 1,500 ml. water). Antithyroid antibodies were positive to a dilution of 1/25.

During the first few days in hospital, the patient felt weak, anorexic and nauseated. She also felt faint on standing up for long periods due to orthostatic hypotension. However, with the clearing up of the pneumonia, together with cortisone replacement therapy, she became asymptomatic.

Discussion

The classical symptoms of Addison's disease are well known. However, in early cases, where symptoms are mild and non-specific, diagnosis is more difficult. Increased pigmentation, as in the present case, is an important sign of primary failure of the adrenal cortex. Other common symptoms and signs include weakness, weight loss, hypotension, anorexia, nausea and emesis. In early adrenal failure, the ability of the adrenal glands to respond to stress is impaired, and the signs and symptoms become more pronounced in periods of stress. The failure of the adrenal cortex to respond to stress forms the basis of the A.C.T.H. stimulation test, an important test in the diagnosis of Addison's disease.

While tuberculosis is fairly common in Singapore, Addison's disease is very rare in these patients, and is usually of the idiopathic variety. The reason for this is obscure.

It is postulated that the primary amenorrhoea in this patient arose on an autoimmune basis with damage and dysfunction of the ovarian tissues before menarche (Irvin et al., 1968).

Summary

A case of idiopathic Addison's disease associated with primary amenorrhoea and antibodies against the thyroid gland is reported. The rarity of Addison's disease inspite of the frequency of tuberculosis is stressed. The frequent association of idiopathic atrophy of the adrenals with other autoimmune disorders is noted. Early diagnosis is important as acute decompensation brought on by stress may be fatal.

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