Sudden Death in Addison’s Disease: Lead Poisoning-like Gum Appearance

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ABSTRACT

Fatal Addison’s disease is rarely found in forensic cases. We report the sudden and unexplained death of a 51-year-old woman on arrival at the emergency room. Previous clinical history revealed frequent hypotension, hyponatremia and persistent hyperpigmented skin consistent with Addison’s disease. However, the diagnosis could only be made during autopsy. The adrenal gland was completely absent. Postmortem blood cortisol was very low (0.86 µg/dL). The thyroid gland showed Hashimoto thyroiditis. The probable cause of Addison’s disease in this case was autoimmune adrenalitis.

INTRODUCTION

Chronic adrenocortical insufficiency is an uncommon disorder resulting from progressive destruction of the adrenal cortex [1]. Clinical features include progressive anaemia, bronze skin pigmentation, severe weakness, hypotension, nausea, vomiting, anorexia, weight loss and hypoglycemia [1-4]. Hyponatremia is observed in about 80% of acute cases whereas less than half present with hyperkalemia [5]. These clinical features were first reported as Addison’s disease in “On the Constitutional and Local Effects of Disease of the Supra-renal Capsules” in the London Medical Gazette, 1849 by Thomas Addison [2].

Nowadays autoimmune disease is the most common cause of Addison’s disease in western countries [2, 4, 6]. The remaining causes are tuberculosis, adrenomyeloneuropathy, systemic fungal infection, AIDS, metastatic carcinoma and isolated glucocorticoid deficiency [2, 4]. The prognosis is excellent [7], but mortality rate was more than 2-fold higher compared with the normal population in a 14 year period [8]. We report a fatal case of autoimmune Addison’s disease that was diagnosed only during autopsy.

CASE REPORT

A 51-year-old woman was found unconsciousness at home. Her relative called the Emergency Medical Service (EMS) for help. EMS personnel transported her to the emergency room of the provincial hospital. The patient was found apneic and pulseless.
The autopsy was performed on 3 November 2009 (21 hours after the pronounced death). The body was of a middle aged, poor nourished female, 153 cm in length with short black hair. Axillary and pubic hair was completely absent. Her skin looked generally dark especially on the palmar creases, all joint areas, and lips. The upper gum showed bluish-black patches, which looked like a lead line in lead poisoning (Figure 1).

Figure 1: Hyperpigmentation of the gum mimicking the lead line in lead poisoning

There was no wound or injection mark on the skin. The internal examination showed no evidence of vital organs injury. The brain had no pathological lesion. The pituitary fossa had no abnormal mass. The thyroid gland was normal shape and weighed 15 g. The airways showed no edema or foreign body obstruction. Both lungs showed mild edema with left upper lung consolidation. There was no pulmonary thromboembolism. The right and the left lung weighed 370 g and 470 g, respectively. There were some petechiae on the anterior external surface of the heart. The left anterior descending coronary artery showed 10% stenosis. The right main coronary artery showed no significant gross pathologic abnormality. Both kidneys showed a diffuse micronodular surface. No evidence of acute pyelonephritis was detected. The adrenal gland was normal shape and weighed 15 g. The airways showed no edema or foreign body obstruction. The myocardium showed hypertrophy with a small focal area of fibrosis. Some focal haemorrhages were present in the subepicardium and the myocardium. There were random contraction band necroses. The thyroid gland showed Hashimoto thyroiditis. There was pulmonary oedema with some focal hemorrhages in the lungs. The brain tissue was unremarkable.

DISCUSSION

Fatal Addison’s disease is rarely found in forensic practice, especially in Northern Thailand. Sudden death from Addison’s disease has been reported but mostly in Caucasians. Several studies showed that Addison’s disease could only be diagnosed during autopsy. The most specific sign of primary adrenal insufficiency is hyperpigmentation of the skin and mucosal surfaces which is due to the high plasma corticotrophin concentrations that occur as a result of a decrease of cortisol feedback. Malaise, hyperpigmented skin, hypotension and hyponatremia in our case were clues for diagnosis of chronic primary adrenal insufficiency. However, the dark line on the gum may be mistaken as the lead line in lead poisoning.

However, the detection of blood cortisol indicates some remaining cortisol secreting tissue. Autoimmune adrenalitis is the main cause of Addison’s disease and may occur alone or as a component of type I or II autoimmune polyglandular syndrome. The Hashimoto thyroiditis of this case indicated that the autoimmune disease was the probable cause of adrenocortical insufficiency.

Autoantibodies against 21-hydroxylase, one of the enzymes in steroid biosynthesis inside the adrenal glands, can be found in approximately 80% of the Addisonian persons. These autoantibodies thus clearly correlate with the disease and are useful for its diagnosis. Approximately 21% of persons positive for adrenal cortex autoantibodies (ACA) developed overt Addison’s disease within 5.2 years, while negative ACA persons maintained normal adrenal function during the observation period. ACA is also an additional marker to predict Addison’s disease. In conclusion, clinicians should not overlook hyperpigmentation of the skin combined with other significant clinical signs and basic laboratory tests for the correct diagnose of adrenal insufficiency which is potentially fatal if not recognised and promptly treated.
Figure 2: Photomicrograph of the thyroid gland tissue of the deceased (X 20 magnification): The thyroid parenchyma contains a dense lymphocytic infiltration with germinal centers (Hashimoto thyroiditis).

REFERENCES


