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Acute Renal Failure with Severe Hyperkalaemia Revealing Schmidt Syndrome

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Abstract: **Case Report**

Background: Schmidt's syndrome, or type II autoimmune polyendocrinopathy, is characterized by the coexistence of autoimmune conditions, most commonly Addison's disease, autoimmune thyroiditis, and gonadal insufficiency. Addison's disease, though rare, may present with acute renal failure, complicating timely diagnosis and treatment. ***Case Presentation***: We report a case of a 21-year-old female presenting with nonspecific gastrointestinal symptoms, severe asthenia, hypotension, and secondary amenorrhoea. Laboratory investigations revealed acute renal failure, hyperkalaemia, hyponatraemia, hypothyroidism, and primary ovarian failure. Clinical re-evaluation noted hyperpigmentation suggestive of Addison's disease, confirmed by markedly reduced serum cortisol, elevated ACTH, and positive anti-21-hydroxylase antibodies. Imaging showed adrenal atrophy. Prompt initiation of intravenous hydration and hydrocortisone led to rapid clinical and biochemical improvement, including restoration of menstrual cycles. ***Discussion***: This case highlights the diagnostic challenge of Addison's disease, especially when masked by renal failure symptoms. The underlying pathophysiology involves combined glucocorticoid and mineralocorticoid deficiency, leading to volume depletion, electrolyte imbalance, and reduced renal perfusion. Early recognition and hormone replacement are crucial to prevent potentially life-threatening outcomes. ***Conclusion***: Addison's disease, particularly as part of Schmidt's syndrome, should be considered in patients presenting with unexplained renal failure, electrolyte disturbances, and endocrine abnormalities. Timely diagnosis and corticosteroid replacement can result in dramatic clinical recovery.

Keywords: Addison's disease, Schmidt's syndrome, Autoimmune polyendocrinopathy, Acute renal failure, Hyperkalaemia

INTRODUCTION

Schmidt's syndrome, or type II autoimmune polyendocrinopathy, is a relatively common syndrome that occurs mainly in adults. It combines several autoimmune diseases, including Addison's disease, thyroiditis and peripheral gonadal insufficiency [1].

Addison's disease is a rare disorder that often presents with non-specific clinical signs. It can present as acute renal failure associated with hyperkalaemia. Here, we discuss the possible mechanisms underlying acute renal failure in Addison's disease, which is part of the polyendocrinopathy, and the spectacular improvement observed after treatment with rehydration and intravenous hydrocortisone.

Case Presentation

A 21-year-old female presented 15 days prior to admission with a digestive complaint of nausea, vomiting, generalised asthenia and epigastric pain, all evolving in a context of weight loss. Clinical examination revealed an altered patient, hypotensive to 80/60 mmHg, normocardial to 70 bpm.

Paraclinical examinations revealed acute renal failure, hyperkalaemia (8 mmol/l) and hyponatraemia (124 mmol/l). Given the hyperkalaemia without electrical signs on ECG, the patient was treated with Kayxalate and calcium gluconate and hydration with saline and sodium; the blood ionogram improved. In the face of chronic constipation, a TSH was performed, returning to 11 uui/ml with anti- TPO antibodies at 1000. An ultrasound scan revealed thyroiditis, and the patient was put on levothyrox 50ug/d. The patient continued to suffer from profound asthenia and also reported secondary amenorrhoea for 4 months with negative BHCG and a hypophysiogram showing hypergonadotropic hypogonadism in favour of primary ovarian failure. On re-examining the patient, our attention was drawn to the presence of hyperpigmentation of the skin and slate-coloured patches, slate-coloured patches in the oral mucosa. The diagnosis of Addison's disease was suspected. A cortisol assay was ordered, which came back collapsed at 0.01µg/dl with ACTH elevated to 199pg/ml with anti-21-hydroxylase antibodies at 5.0U/ml (VN<0.4) confirming the diagnosis. The patient was started on intravenous hydration and hydrocortisone, with marked improvement in clinical and paraclinical parameters and restoration of menstrual cycles, followed by oral hydrocortisone and fludrocortisone. As part of the etiological diagnosis, an adrenal CT scan revealed adrenal atrophy (absence of visible adrenal parenchyma). Other autoimmune disorders in the context of polyendocrinopathy were ruled out in our patient.

Figure 1: Specular disappearance of melanoderma under hydrocortisone

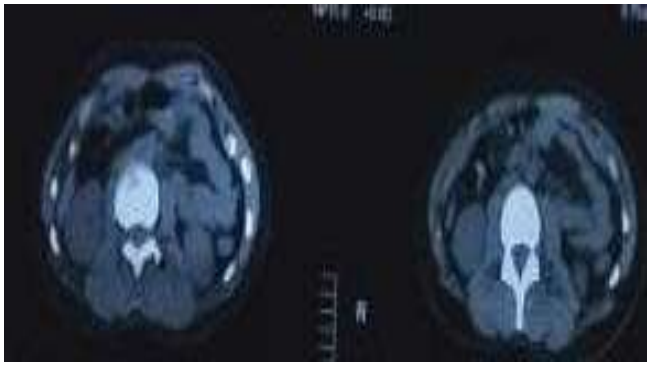

Figure 2: Adrenal Atrophy

DISCUSSION

Primary adrenal insufficiency or ‘Addison's disease’, first described by Thomas Addison [2], is defined by a deficit in the secretion of adrenocortical hormones (glucocorticoids, mineralocorticoids and androgens). Adrenal insufficiency is most often caused by an autoimmune disease. Autoantibodies to steroidogenic enzymes in the adrenal cortex are present in 86% of patients with primary autoimmune adrenal insufficiency, the most common antibody being anti-21-hydroxylase. Addison's disease in ISAP is associated with a polymorphism of genes in the HLA system [3]. Addison's disease is a rare cause of acute renal failure [4]. Mineralocorticoids promote sodium reabsorption in exchange for potassium and hydrogen secretion in the kidney, which explains the occurrence of hyponatremia and hyperkalaemia in mineralocorticoid deficiency. Mineralocorticoids also increase vascular tone and cardiac inotropism. On the other hand, glucocorticoids increase renal glomerular filtration and urinary sodium excretion, and also potentiate the vasoconstrictive effect of catecholamines on the cardiovascular system [5]. The combination of glucocorticoid and mineralocorticoid deficiency leads to a reduction in extracellular volume and a fall in cardiac output, resulting in a reduction in renal perfusion rate and glomerular filtration rate. This was the most likely mechanism explaining the onset of acute renal failure in this patient. They may be mistakenly considered as a manifestation of renal failure, which increases the likelihood of delayed diagnosis in the management of the disease. Based on this clinical case, a number of points should be discussed: the non-specificity of the clinical signs of Addison's disease may be

responsible for a delay in diagnosis and therapeutic management; the association of an increase in creatinine, hyperkalaemia and hyponatraemia in Addison's disease may be considered as a manifestation of acute renal failure. Hyperkalaemia in Addison's disease can be potentially dangerous. The usual treatment of hyperkalaemia with intravenous insulin and glucose should be avoided in this case, as it is unnecessary and dangerous in patients predisposed to hypoglycaemia due to hypocorticism. The hyperkalaemia subsided as soon as she was put on saline hydration and intravenous hydrocortisone. Fortunately, in our case, the patient benefited from Kayxalate and calcium gluconate to treat the severe hyperkalaemia. 90% of patients with primary ovarian insufficiency and Addison's disease have at least one positive antibody, including StCA, anti- 17-hydroxyprogesterone and P450scc antibodies. Italian studies have shown that primary ovarian insufficiency occurs on average two years before adrenal insufficiency [6]. When amenorrhoea occurs secondary to primary ovarian insufficiency, oestroprogestogenic treatment is introduced, either contraceptive or solely substitutive. This combines oral or transcutaneous 17-estradiol with a progestin administered continuously or sequentially (10 to 14 days per month). It is continued until at least the physiological age of menopause. In our patient, we did not use oestroprogestogenic treatment because once hormonal balance was achieved, the menstrual cycle was re-established with normal gonadotropic axis tests.

CONCLUSION

Although rarely reported, Addison's disease is a cause of acute renal failure, the course of which can be fatal in the absence of early diagnosis and correct initial management, based on rehydration and intravenous hydrocortisone, which leads to a spectacular improvement in symptoms. It may also be part of a polyendocrinopathy.

Compliance with ethical standards

Acknowledgments

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Disclosure of conflict of interest: The authors declare no conflict of interests.

Statement of ethical approval: The present research work does not contain any studies performed on animals/humans subject by any of the authors.

Statement of informed consent: Informed consent was obtained from all individual participants included in the study.

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