Polyglandular Autoimmune Syndrome Presenting as Acute Renal Failure with Severe Hyperkalemia

M.	Moses	Elisaf
H.	Haralampos	Pappas
K.C.	Kostas C.	Siamopoulos

Department of Internal Medicine, Medical School, University of Ioannina, Greece

K.C. Siamopoulos, Associate Professor of Medicine/Nephrology, University of Ioannina, GR-451 10 Ioannina (Greece)

Dear Sir,

Polyglandular autoimmune syndrome was first described by Schmidt in patients with both Addison's disease and lymphocytic thyroiditis. The disease has subsequently been expanded to include any combination of adrenal insufficiency, lymphocytic thyroiditis, gonadal dysfunction and diabetes mellitus [1]. Even though prerenal azotemia has been described in patients with Addison's disease [2, 3], to the best of our knowledge, acute renal failure (ARF) has not yet been reported in patients with Schmidt's syndrome.

A 38-year-old woman was admitted to the hospital because of ARF with severe hyperkalemia. She had developed a generalized vitiligo at the age of 9 years. The patient had been in a stable state of health until 3 months earlier, when anorexia, muscle weakness, somnolence and progressive weight loss developed. Laboratory investigation performed 2 months before the patient's admission revealed primary hypothyroidism, while the other parameters tested (including serum creatinine) were within normal limits. Thyroxine (75 µg/day) was prescribed. Despite treatment, the symptoms gradually worsened: muscle weakness became prominent, she was unable to move without any help and experienced nausea, vomiting and restlessness. On examination, low blood pressure was found (90/60 mm Hg), and she was admitted to the hospital. Laboratory parameters are shown in table 1. Microscopic examination of a urine sediment was negative and no proteinuria was found.

Because of severe hyperkalemia, one session of hemodialysis was performed. Correction of hypovolemia was followed by a progressive improvement in renal function. Glucocorticoid and mineralocorticoid replacement therapy was given and ameliora-

Table 1. Laboratory parameters of the patient

Urine values represent measurements in spot urine on admission. Figures in parentheses represent normal values. 1 Prolonged ACTH stimulation test was abnormal.

tion of the symptoms/signs, as well as restoration of electrolyte parameters were observed within a few days.

Our patient fulfilled the criteria for the diagnosis of polyglandular autoimmune syndrome [1]. Interestingly, even though cortisol levels were very low, skin hyperpigmentation was not evident. The absence of the expected skin hyperpigmentation was due to the generalized vitiligo. Hypovolemia was probably the cause of the patient's ARF, and could be ascribed to a decreased water intake, vomiting-induced fluid losses, as well

as salt and water losses due to both hypoaldosteronism and hypercalcemia. However, despite the hypovolemia, urine sodium was increased, while the ratio urea/creatinine was not raised. The inappropriate sodium loss could be ascribed to hypoaldosteronism, and the absence of an increased urea/creatinine ratio was possibly due to a decreased urea production. Both glucocorticoid deficiency, which decreases the protein catabolism, and hypothyroidism, which can reduce both the synthesis and degradation of proteins, as well as a decreased food intake lead

KA/GEI

E-Mail karger@karger.ch Fax + 41 61 306 12 34

© 1996 S. Karger AG, Basel 0028-2766/96/0723-0497\$10.00/0

to decreased hepatic urea production. In our patient, severe hyperkalemia was also prominent. However, due to coexistent hypercalcemia, no abnormalities in cardiac rhythm were present. Hyperkalemia was mainly the result of adrenal insufficiency. Nevertheless, effective circulating volume depletion could also have played a role in the significant increase in serum potassium concentration [4]. Even though serum chloride levels were within normal limits, a relative hyperchloremia (serum $\text{Na}^+/\text{Cl}^- = 1.33$) was apparent. The presence of hyperchloremic metabolic acidosis in our patient was probably the result of (a) a loss of aldosterone effect at the outer medullary collecting duct, (b) a diminished phosphate excretion from lack of glucocorticoid and (c) a depression of ammoniogenesis by hyperkalemia [5-7]. Finally, hypercalcemia was also present in our patient, possibly owing to adrenal insufficiency [8].

References

Leshin M: Southwestern internal medicine conference: Polyglandular autoimmune syndromes. *Am J Med Sci* 1985;290:77-88.

Loeb RF: Chemical changes in the blood in Addison's disease. *Science* 1932;76:421-422.

Burston RA, Garrod O: The variability of the lowered glomerular filtration rate in Addison's disease and panhypopituitarism, and the effect of cortisone therapy thereon. *Clin Sci* 1952; 11: 129-139.

Anderson HM, Laragh JH: Renal excretion of potassium in normal and sodium depleted dogs. *J Clin Invest* 1958;37:323-331.

Perez GO, Oster JR, Vaamonde CA: Renal acidification in patients with mineralocorticoid deficiency. *Nephron* 1976;17:461-473.

Szymlan P, Better OS, Chaimowitz C, Rosier A: Role of hyperkalemia in the metabolic acidosis of isolated hypoaldosteronism. *N Engl J Med* 1976;294:361-365.

Hulter HN, Slnicki LP, Harbottle JA, Sebastian A: Impaired renal H^+ secretion and NH_3 production in mineralocorticoid-deficient, glucocorticoid-replete dogs. *Am J Physiol* 1977; 232:F136-F146.

Walser M, Robinson BHB, Duckett JW: The hypercalcemia of adrenal insufficiency. *J Clin Invest* 1963;42:456-465.

498

Nephron 1996;72:497-498

Elisaf/Pappas/Siamopoulos