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Nephrology

AUTOIMMUNE ADRENALITIS REVEALED BY ACUTE KIDNEY INJURY.

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ABSTRACT A 44-years old female with no medical or drug history was presented with weakness, nausea and vomiting. On exam she present hypotension, tachycardia and signs of dehydration. Laboratory tests showed renal failure, metabolic acidosis and severe electrolytic disorders. Urine culture revealed a bacterial urinary tract infection. Kidney function, hyponatremia and hyperkalemia did not fully improve despite intravenous hydration and appropriate antimicrobial therapy. Repeated episodes of hypoglycemia in a context of weakness and lethargy. The diagnosis of autoimmune adrenalitis was made in the presence of low serum cortisol levels and positive anti-21 hydroxylase antibodies.

KEYWORDS : primary adrenal insufficiency, autoimmune adrenalitis; anti-21 hydroxylase antibodies; acute kidney injury.

Introduction

Autoimmune adrenalitis or autoimmune adrenal insufficiency is a rare endocrine disorder related to the destruction of the adrenal glands by anti-adrenal autoantibodies. It is currently the leading cause of primary adrenal insufficiency (PAI) in about 80% of cases [3]. The diagnosis is often elusive, requires some awareness of the disease, knowledge of its signs and symptoms (nonspecific) in order to be clearly established; in other words, it will only be worn during acute decompensation. Exceptionally, acute kidney injury (AKI) may dominate the clinical picture, only a few cases have been described in the literature [6]. We report a new observation that illustrates this atypical and extremely rare presentation.

Observation:

A 44-year-old female with no medical or drug history, was addressed to our department suffering from weakness, nausea and vomiting for about ten days. The clinical examination was normal except for arterial hypotension at 90/50 mmHg, tachycardia at 116 beats / min and signs of dehydration.

Laboratory tests showed renal failure: high blood urea nitrogen at 1.03 g/l (0.21-0.43 g/l), serum creatinine at 30 mg/l (5-12 mg/l) with severe electrolytic disorders: hyponatremia at 119 mmol/l (135-145 mmol/l), hypochloremia at 85 mmol/l (95-105 mmol/l) and hyperkalemia at 6.92 mmol/l (3.5-5.1 mmol/l), and metabolic acidosis: serum bicarbonates at 15 mmol/l (22-29 mmol/l); hypoglycemia at 0.70 g/l (0.72-1.07 g/l) and CRP at 143 mg/l (0-4.5 mg/l). The blood count showed a leukocytosis at 13000 cells/mm^3 (4000-10000 cells/mm^3) predominantly neutrophilic (neutrophils at 11000 cells/mm^3).

Urine culture isolated a *Klebsiella Oxytoca* which was susceptible to ceftriaxone. On renal ultrasonography, both kidneys were of normal size, with regular contours and normal cortical thickness (Figure 1).

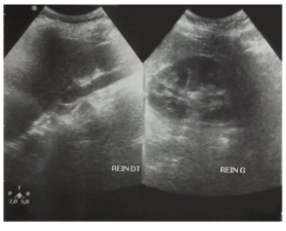

Fig.1: Ultrasonography showing normal sized kidneys with good corticomedullary differentiation.

After intravenous hydration and appropriate antimicrobial therapy, CRP and leukocyte count decreased, while renal function, hyponatremia and hyperkalemia did not fully improve. The patient was still experiencing nausea, vomiting, hypotension and suffering from repeated episodes of hypoglycemia in a context of weakness and lethargy.

The diagnosis of adrenal insufficiency was then evoked, a serum cortisol concentration returned at 37 nmol/l (171-536 nmol/l). The patient was then treated with intravenous hydrocortisone resulting in marked clinical improvement with correction of renal parameters and electrolyte disorders. As part of the etiological diagnosis, anti-21 hydroxylase antibodies were positive at 13.3 U/ml (≤ 1 U/ml). Abdominal CT showed atrophic adrenals (Figure 2). The etiological investigation for an associated autoimmune disease remained negative. The patient was later put on oral hydrocortisone replacement therapy at a dose of 30 mg / day and an Addisonian card was delivered after therapeutic education.

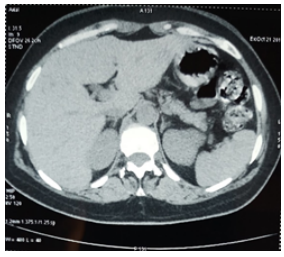

Fig.2: CT abdominal axial view, showing atrophy of the adrenal glands.

Discussion

Autoimmune adrenalitis, also called autoimmune adrenal insufficiency or cortical retraction of the adrenals, results from the destruction of the adrenal cortex by an autoimmune mechanism. It is currently the leading cause of PAI, with an incidence of 4.4 to 6 / million inhabitants / year [9]. It is a rare disease that may appear isolated or associated with other autoimmune endocrine diseases [1]. Pathophysiological mechanisms imply cellular and humoral immunity. The target antigens are the steroidogenesis enzymes (CYP21A2, CYP17 and CYP11A1), associations with the HLA B8, DR3 and DR4 phenotypes have been found [4].

The clinical presentation is nonspecific and differs according to whether the deficit is manifested as an acute onset (acute adrenal insufficiency) or as the form of chronic insufficiency. Typical signs of cortisol deficiency are weakness, depressed mood, weight loss, anorexia, abdominal disorders (vomiting, nausea or diarrhea), arthralgia or myalgia [8]. Melanoderma is a pathognomonic sign of the primitive origin of adrenal insufficiency, resulting from increased stimulation of melanocortin type 1 receptors in the skin due to the high concentration of circulating ACTH and other cleavage of pituitary pro-opiomelanocortin (POMC) [7], this cutaneous hyperpigmentation was not detected in our patient because of her dark skin phototype.

During PAI as in the reported observation, patients present with severe hypotension, gastrointestinal symptoms and hypovolemic shock secondary to acute dehydration. Laboratory tests may reveal

hyponatremia, hyperkalemia, metabolic acidosis, hypercalcemia and sometimes hypoglycemia. The determination of the morning serum cortisol concentration allows the diagnosis of adrenal insufficiency, a high rate of ACTH confirms its primary origin. In our case, clinical symptomatology and renal failure were initially related to the urinary tract infection. It was the lack of improvement despite adequate hydration and appropriate antimicrobial therapy and the observation of repeated low blood glucose levels that made the diagnosis of adrenal insufficiency suspect. Serum cortisol level is then required and has returned low. The autoimmune origin is based on the detection of anti-21-hydroxylase antibodies, which are found in the majority of cases [5,2]. The adrenal CT shows, as in our case, bilateral adrenal atrophy. Treatment is based on lifetime replacement with glucocorticoids sometimes in combination with mineralocorticoids.

Although rarely reported, adrenal insufficiency is a well-described cause of acute renal failure [6]. Indeed, glucocorticoids promote the retention of water and sodium and potassium leakage. They also increase renal glomerular filtration and sodium excretion in the urine and potentiate the vasoconstrictor effect of catecholamines on the cardiovascular level. Thus, the glucocorticoid deficiency leads to resistance to catecholamines and therefore hypotension. The latter is reinforced by the sodium depletion associated with vomiting, diarrhea and mineralocorticoid deficiency. The combination of glucocorticoid deficiency and mineralocorticoids induces a reduction in extracellular volume and a decrease in cardiac output resulting in a reduction in renal perfusion rate and glomerular filtration rate, which result in the occurrence of acute renal failure [3].

Thus, for the diagnosis of adrenal insufficiency to be evoked in the presence of acute renal failure, a certain awareness of the disease must be necessary for nephrologist, in order to avoid any diagnostic delay and the realization of unnecessary additional exams. In the reported case, AKI was initially related to urinary tract infection, but it's persistence despite rehydration and antibacterial therapy led to a reconsideration of the diagnosis. The association of clinical signs of hypovolemia with hyponatremia, hyperkalemia and hypoglycemia in a patient receiving no otherwise treatment was suggestive of the diagnosis of adrenal insufficiency.

The usual treatments of hyperkalemia by glucose infusion and intravenous insulin should be avoided in patients with adrenal insufficiency; not only is it ineffective, it may also provoke or potentiate dangerous hypoglycemia, to which these patients are already predisposed due to their low levels of glucocorticoid [3]. Hyponatremia with inappropriately raised urine osmolality and sodium concentrations most commonly results from SIADH, for which the first-line treatment is fluid restriction. However, these findings may also arise in adrenal insufficiency, the treatment of which is fluid resuscitation and steroid replacement [3]. These therapies may be life-threatening, whereas simple saline rehydration and hydrocortisone replacement may correct clinical and paraclinical symptoms.

In conclusion, this case shows that adrenal insufficiency may cause AKI, and it can present as hypotension associated with hyponatremia, hyperkalemia and hypoglycemia. After excluding more common diseases, and before making unnecessary further investigations or examinations, the possibility of adrenal insufficiency should be considered as the cause of AKI, even in patients with no prior history of glucocorticoid therapy.

Disclosure

The authors report no conflicts of interest in this work.

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