Case report

Myopathy in Addison’s disease

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Summary Since the first description of primary adrenocortical insufficiency by Thomas Addison in 1855 several large series of patients with Addison’s disease have been published.1-3 The common signs and symptoms include: weakness, hyperpigmentation, weight loss, gastrointestinal complaints, and hypotension.3 It is rare for patients with Addison’s disease to present with musculoskeletal symptoms including flexion contractures,4 5 hyperkalaemic neuromyopathy,6 Guillain-Barré syndrome,7 migratory myalgia,8 sciatica-like pain,9 and low back pain.10 Myopathy has not been previously described in Addison’s disease. Herein we report a patient presenting with severe hyponatraemia and myopathy which resolved after steroid replacement therapy.

Key words: adrenocortical insufficiency, hyponatraemia, endocrine myopathy.

Case report

A 44 year old woman was admitted because of weakness, recurrent vomiting, and leg pain of two weeks’ duration. Past history included secondary amenorrhoea since age 24 due to premature ovarian failure after three normal pregnancies, and urinary tract infections for several years. The patient had increasing weakness and had lost 20 kg in weight over the previous two years. During the two weeks before admission the patient complained of severe pain in both thighs and knees aggravated by motion, generalised weakness, and recurrent vomiting after each meal.

On physical examination the patient was somnolent, with blood pressure 100/70 mmHg and pulse 88—regular; diffuse skin hyperpigmentation and scant axillary and pubic hair were noted. There were 60° flexion contractures of both knees with pronounced tenderness in the thigh muscles.

Laboratory studies showed white blood cell count 4.6×10⁹ cells/l; haemoglobin 135-2 g/l, serum sodium 103 mmol/l, potassium 5-0 mmol/l, chloride 75 mmol/l. Serum osmolality was 236 mosmol/kg, urine osmolality 268 mosmol/kg, urinary sodium 39 mmol/l, potassium 25 mmol/l. Blood urea nitrogen, creatinine, glucose, bilirubin, alkaline phospahatase, calcium, phosphorus, uric acid, total protein, and albumin were in the normal range. Serum aspartate transaminase was 183 IU/l (normal range 7-40), lactic dehydrogenase 260 IU/l (100-225), creatine phosphokinase 1670 IU/l (24-175), and aldolase 5.59 IU/l (7-6). Creatine phosphokinase isoenzymes showed an MM fraction only.

Endocrinological test results were: serum cortisol 8 am—0-02 µmol/l (0.22-0.55); 8 pm—0.02 µmol/l (0.11-0.22); adrenocorticotrophic hormone 28-50 pmol/l (0.95-3.8); synacthen test: basal cortisol 0-014 µmol/l, after stimulation 0-014 µmol/l; free thyroxine 19-5 pmol/l (12-36); tri-iodothyronine 1.35 nmol/l (1.35-3.15); follicle stimulating hormone 40 IU/l (1.3-4.3); luteinising hormone 8-95 IU/l (1.2-3.2); aldosterone 0.075 nmol/l (0.15-0.48).

Chest x ray and electrocardiogram were normal, intravenous pyelography—no calcification in the suprarenal area was seen. Computed tomography of brain and of abdomen were normal.

The patient received hypertonic saline infusions and from the second day corticosteroid therapy. Serum sodium rose gradually to normal levels within
Muscle group, 1 Fig. (EMG) electromyogram to (Fig. day was production potential duration, patient developed no and to two weeks later she oral was she normal while muscular relaxation, persistent motor neurone discharges at rest, and demyelinating neuropathy has also been described.

The syndrome of Guillain-Barré has been reported in three patients with Addison’s disease. Serum levels of muscle enzymes were normal and EMG was consistent with diffuse motor neuropathy.

Interstitial migratory myalgia was the presenting symptom of Addison’s disease in a 40 year old patient. Serum creatine phosphokinase, aldolase, and thyroxine were normal, and the symptoms responded to steroid replacement therapy. Two additional patients have been described in whom sciatica-like pain and chronic low back pain were the presenting complaint of Addison’s disease.

Our patient presented with muscle pain and tenderness, contractures of the knees, increased levels of muscle enzymes, and a myopathic pattern on EMG. Since both the myopathic features and the signs and symptoms of Addison’s disease responded concomitantly and dramatically to steroid replacement therapy we assume they are causally related. Theoretically, myopathy in Addison’s disease could result from mineralocorticoid or glucocorticoid deficiency, or both. On admission our patient had severe hyponatraemia and reduced aldosterone levels, reflecting significant impairment of mineralocorticoid function. In the literature we found two case reports of rhabdomyolysis in hypotnatraemia. In these patients the fall in serum sodium was rapid, in the clinical context of acute water intoxication. In view of the long history suggestive of Addison’s disease and the mild neurological symptoms relative to the degree of hyponatraemia we assume the metabolic derangement in our patient developed gradually, and hence we are inclined to relate the myopathy to the glucocorticoid deficiency. Since both the electrolyte imbalance and the steroid deficiency were corrected simultaneously, however, the relative role of these two factors in causing the muscle damage could not be ascertained.
Despite increasing knowledge of the metabolic effect of hormones, little is known about the pathogenesis of endocrine myopathies. Since the effects of physiological doses of steroids, or steroid deficiency on muscle, have not been examined, the possible relation of myopathy to Addison’s disease remains to be elucidated.

References