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Creatine Phosphokinase-Linked Immunoglobulin in a Patient with Hypokalemic Myopathy

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A macromolecular complex between creatine phosphokinase (CPK) and an immunoglobulin has been identified in a variety of diseases including ischemic heart disease, neoplastic diseases and muscle disorders. However, the biological and pathological significance of the complex is not fully understood. We report here the first observation of the presence of the macro-CPK type 1 in a patient with hypokalemic myopathy.

A 49-year-old woman with no prior episode of muscle weakness was admitted because of increasing, symmetrical weakness in proximal upper and lower limbs over the previous month. She was unable to rise from a sitting position without using her arms. The affected muscle groups were hypotonic and tender without fasciculations. Laboratory data included: CPK 744 IU/l, aldolase 20.8 IU/l, lactate dehydrogenase 922 IU/l, serum Na 139 mEq/l, serum K 2.0 mEq/l, serum Cl 101 mEq/l. No autoantibodies were found. A CPK isozyme electrophoresis demonstrated an extraband which was found to be migrating in a position between the CPK-MM and CPK-MB bands (fig. 1). The extra-CPK band constituted approximately 9% of the total CPK activity. A combination of thin layer gel filtration and immunofixation electrophoresis revealed CPK-linked IgA (κ and λ). A morphological study of biopsied muscle demonstrated a selective atrophy of type 2B fibers in addition to a variation in size of muscle fibers and mild degenerative and regenerative changes without inflammatory cell infiltration.

Immediately after admission, intravenous supplementary therapy with potassium was started (fig. 2). A total of 420 mEq of potassium chloride was given during the first
7 days. The serum K concentration returned to normal within 4 days and the elevated muscle enzymes returned to normal in a week. Although oral potassium supplements were continued and the serum K level was kept in the normal range, the patient’s proximal muscles remained weak and tender despite a steady trend toward full recovery.

Although no underlying diseases or medicines to produce severe potassium depletion were identified, the neurological and laboratory findings were consistent with well-documented descriptions of hypokalemic myopathy [8,9]. This patient presented herein is the first to document a possible association between hypokalemic myopathy and macro-CPK type 1. The mechanism of the generation of a complex between CPK and immunoglobulin is at present unclear. Evidence is presented suggesting that CPK-linked immunoglobulin may be a circulating immune complex [4, 5]. An interesting finding in our patient is that the macro-CPK decreased and finally disappeared during the period of potassium therapy. This and other observations may indicate that CPK is transiently antigenic in certain pathological conditions [2, 6].

Although the immune complex theory is well accepted, whether the physicochemical form of CPK is altered in association with acute rhabdomyolysis remains to be addressed in future investigations of this phenomenon.

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References