Case Report

Recurrent rhabdomyolysis and myoglobinuric acute renal failure in a patient with polymyositis

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Introduction

We report on a 57-year-old woman with polymyositis who on two occasions presented with rhabdomyolysis and myoglobinuria, with resultant oliguric acute renal failures, the second episode of which required haemodialysis.

Polymyositis is a rare and gradually progressive autoimmune inflammatory disease of skeletal muscle that is characterized by muscle weakness, usually proximal and symmetric, elevated muscle enzymes, and distinctive findings on electromyography and muscle biopsy [1]. The disease may be associated with infections or with other collagen vascular diseases, such as Sjögren’s syndrome and sarcoidosis [1].

Rhabdomyolysis is a term that refers to disintegration of striated muscle, which results in the release of muscle cell constituents, in particular myoglobin, into extracellular fluid and the circulation. The released myoglobin is filtered by glomeruli and reaches the tubules, where it may cause obstruction and renal dysfunction [2]. The main causes of rhabdomyolysis include crush injuries, burns, infections, medications, as well as illicit drugs, and exercise [3]. Several patients who had recurrent rhabdomyolysis after intense exercise or generalized tonic-clonic convulsions were reported to have genetic errors of carbohydrate or lipid metabolism in the muscle [4,5]. Status epilepticus can cause recurrent rhabdomyolysis in subjects with normal glycolytic and lipolytic enzyme activity [6]. Younger et al. [7] also reported a case of recurrent myoglobinuria without acute renal failure in a patient with human immunodeficiency virus-associated polymyositis. The kidneys are generally spared in polymyositis [8] and renal failure is rare [8,9], although myoglobulinuria may occur in as many as 20% of patients with polymyositis.

Case

A 57-year-old woman was admitted to the emergency room at St Vincent’s Hospital, Suwon, Korea, with posterior neck pain and pain and weakness in her upper and lower extremities. The patient had felt well until 5 days before admission, when she began having difficulty walking and dressing and noticed swelling of her legs. The patient denied engaging in strenuous exercise or using alcohol or drugs. She had no family history of neuromuscular disorder and there were no similar cases in her family.

Six months before presentation, she had experienced recurrent episodes of pain in her shoulders and the upper and lower extremities, accompanied by gradually worsening proximal muscle weakness. She had developed oedema, decreased urine output, and dark-coloured urine, and for further examination was referred to our hospital with a diagnosis of rhabdomyolysis with acute renal failure (ARF). She had no history of recent trauma, administration of drugs, infections, physical exercise, or other factors that could cause rhabdomyolysis. At that time, her blood tests showed increased serum creatinine (maximum 4 mg/dl) and muscle enzymes, especially creatinine phosphokinase (CK; 5369 IU/l) and myoglobin (4346 ng/ml). Tc-99m-methylene diphosphonate (MDP) bone scan also demonstrated slightly increased radioisotope uptake in her calf muscles (Figure 1A). After appropriate hydration and bed rest, the patient’s renal impairment and muscle strength gradually improved without haemodialysis. She was discharged without any sequelae, and had been stable and without any pain and weakness in her upper and lower extremities until 5 days before her second admission.
On physical examination she appeared quite ill. Her temperature was 36.5°C, respiratory rate 24 breaths per minute, and blood pressure 160/100 mmHg. There was non-pitting oedema of the lower extremities bilaterally up to the upper thigh, and painful swelling of both of the upper arms. She had no skin rash and nail-fold capillary microscopy was normal. Her gait was limited by pain. Neurological examinations on admission revealed no abnormal findings. The admission laboratory tests revealed a normal blood cell count and an erythrocyte sedimentation rate of 50 mm/h. The urine test for myoglobin was positive, and urine dipstick test was positive for blood (3+); her urine was dark coloured, but without erythrocytes evident on microscopic examination. Her serum myoglobin was 63 559 ng/ml. The results of her other tests were as follows: serum creatinine 4.4 mg/dl, serum potassium 6.8 mEq/l, serum CK 50 470 U/l, the MM isoenzyme

Fig. 1. Whole body scan taken at the first admission of the patient (A) shows diffusely increased soft tissue Tc-99m-MDP uptake, with both kidneys visible, and faint soft tissue uptake in both kidneys. A bone scan at the time of the second episode (B) shows diffusely increased soft tissue Tc-99m-MDP uptake in both lower extremities and both upper arms with no activity over the kidneys, suggesting rhabdomyolysis with ARF. Mildly increased bone uptakes are also seen in the right shoulder, both elbows, both wrists and the lumbar spine, suggesting degenerative changes.
of CK (CK-MM level) 33 269 ng/ml, aspartate aminotransferase 1238 IU/l, alanine aminotransferase 647 IU/l, and lactate dehydrogenase 7070 IU/l. Rheumatoid factor was negative, and we detected no antibodies to antinuclear antigen, smooth muscle, ribonuclear protein, double-stranded DNA, or to Jo-1, Ro or La. The serological tests for various anti-viral antibodies and HIV antibody were also negative. Her thyroid function was normal. A Tc-99m-MDP bone scan demonstrated increased radioisotope uptake in various muscles, especially in the upper and lower extremities, with no urinary activity (Figure 1B). A diagnosis of ARF due to non-traumatic rhabdomyolysis was made. The oliguric ARF, hyperkalaemia, and metabolic acidosis did not respond to volume expansion and supportive treatment, and haemodialysis was initiated on the sixth inpatient day. Electromyography (EMG) performed of the deltoid, abductus pollucis brevis, quadriceps and temporalis anterior muscles revealed profuse fibrillations with sharp waves and low-voltage polyphasic units. Nerve conduction studies were normal. Biopsies of the gastrocnemius and quadriceps muscles demonstrated prominent necrotic and fatty degenerative changes in the muscle fibres and inflammatory cells clusters in endomyseal and perivascular sites (Figure 2). Histochemical stains showed normal myophosphorylase, phosphofructokinase and myoadenylate deaminase activity. She was started on prednisone, 1 mg/kg daily, resulting in her muscle pain, weakness and swelling being resolved. Her strength gradually improved, so that on the day of discharge she was able to walk with assistance. Twelve months after her admission, her condition is stable and her dosage of corticosteroids is being decreased further.

**Discussion**

Recurrent rhabdomyolysis can be caused by extreme physical exercise, centrally induced muscle cramps caused by toxic substances such as alcohol and cocaine, heatstroke, hyperthermia, infections (mycoplasma) and, in rare cases, hereditary diseases. Carnitine palmitoyl transferase II deficiency is probably the most frequent cause of hereditary and metabolic myopathies, but rhabdomyolysis can occur with any deficiency of glycolytic enzymes, with fatty acid oxidation disorders, and with many of the mitochondrial cytopathies. However, many cases of recurrent myoglobinuria are deemed idiopathic (Meyer–Betz disease) [10]. In this case, examination of muscle biopsies and the EMG confirmed the diagnosis of polymyositis. Therefore, we suggest that polymyositis should be added to the list of causes of recurrent rhabdomyolysis.

Our patient presented with recurrent rhabdomyolysis and myoglobinuria, and resultant ARF, in the context of an initially unrecognized polymyositis. A definitive initial diagnosis was delayed because the first episode of ARF resulting from rhabdomyolysis was alleviated with intravenous hydration and supportive treatment. During rhabdomyolysis, CK-MM concentration remains elevated in a more consistent manner than that of myoglobin, and it is more reliable for assessing the presence and intensity of damage to the muscles [3]. The rapid recovery of this patient from the first episode of renal dysfunction and the severe deterioration of renal function at the second episode were closely associated with elevated serum CK concentrations and Tc-99m-MDP bone scan uptake in various muscles.
scan findings, i.e., with the severity of damage to the muscles [3].

Both polymyositis and dermatomyositis can cause myoglobinuria [8,9]. Fewer than 10 cases of polymyositis-induced myoglobinemic ARF have been reported to date. However, polymyositis rarely leads to a myoglobinuric ARF that requires haemodialysis [2,7–9]. ARF might play a role in sustaining in circulation and urine massive amounts of myoglobin originating from extensive muscle fibre necrosis in patients with polymyositis. Usually, polymyositis has a gradual progression over a period of 3–6 months, and rarely presents as an acute episode, as it did in this patient. Early recognition and treatment is essential to minimize renal damage and optimally preserve renal function. Once ARF has been established, or severe hyperkalaemia and acidosis are present, the patient may require dialysis [3].

In summary, the diagnosis of polymyositis (or dermatomyositis) must be considered in any patient with recurrent and non-traumatic rhabdomyolysis-induced ARF. Scrupulous attention to renal function and aggressive use of immunotherapy are mandatory to prevent acute renal failure and morbidity in these patients.

Conflict of interest statement. None declared.

References


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