CASE REPORT

Hypokalemia-induced cardiac arrest in a Duchenne muscular dystrophy patient

Zikou X[1], Papathanakos G[1], Dounousi E[2], Nakos G[1], Koulouras V[1]

[1]Department of Intensive Care Unit
[2]Department of Nephrology
University Hospital of Ioannina, Ioannina, Greece

Abstract

Background: Duchenne muscular dystrophy (DMD) is a progressive myopathic disorder, inherited as X-linked recessive traits, in which muscle weakness is the primary symptom. Correlation between DMD and hypokalemia is reported in only three case reports. Recent investigations have focused on the nutritional management of patients with DMD. However, there are no data regarding recommendations on potassium intake.

Description of case: We report the case of a 15-year-old male patient with DMD, who developed sudden cardiac arrest following severe hypokalemia (K: 1.3 mEq/L) during a lower respiratory tract infection. Hypokalemia was gradually corrected with intravenous potassium chloride. The patient, after a prolonged hospitalization due to hypoxic encephalopathy, was discharged from the Intensive Care Unit (ICU) on mechanical ventilation.

Conclusion: Severe hypokalemia is a rare complication of DMD, with potentially lethal consequences. Therefore, in patients with DMD, potassium levels should be closely monitored and adjusted with appropriate diet or potassium supplements as needed. Hippokratia 2016, 20(2): 163-165

Keywords: Duchenne muscular dystrophy, hypokalemia, cardiac arrest

Corresponding Author: Dr. Vasilios Koulouras, Associate Professor, Department of Intensive Care Unit, University Hospital of Ioannina, Avenue Stavros Niarchos, 45500 Ioannina, Greece, tel: +302651099353, fax: +302651099343, e-mail: vpkoulouras@yahoo.gr

Introduction

Duchenne muscular dystrophy (DMD) is a progressive myopathic disorder that is inherited as X-linked recessive traits caused by a defective dystrophin gene[1]. The primary pathologic process is muscle fiber degeneration leading to muscular weakness, which becomes evident between the second and third year of age[2]. Moreover, although children frequently have varying degrees of mild cognitive impairment or global development delay, in most cases they have an average intelligence[3,4]. DMD also causes primary dilated cardiomyopathy (DCM) and a variety of arrhythmias, primarily supraventricular, especially during intercurrent infections or surgery[5]. The incidence of symptomatic cardiomyopathy and heart failure in those patients increases gradually in puberty, although the majority of children with DMD remain relatively asymptomatic until late in the disease course[6]. Patients with DMD are often confined to a wheelchair by approximately the age of 12 years and die in puberty from respiratory insufficiency or cardiomyopathy[7]. Nutrition is of critical importance for the long-term DMD management. Delayed growth, short stature, muscle wasting, swallowing difficulties and increased fat mass are characteristics of DMD and impact on the nutritional status. Also, steroid therapy can exacerbate obesity and cause bone demineralization, while malnutrition is a feature of end-stage disease[8]. Recent studies have focused on publishing recommendations related to nutrition that may attenuate disease severity and progression[9]. Vitamin D and calcium for steroid-induced osteoporosis, creatine monohydrate to improve muscle strength[8] and amino acids to maintain protein balance[10], have been proposed as nutritional supplements. According to the literature, there is little data from older studies about potassium homeostasis in DMD, while nonspecific recommendation exists about potassium intake in such patients. We report a case of a heart attack following severe hypokalemia in a patient with DMD.

Case Report

A 15-year-old boy was admitted to the Intensive Care Unit (ICU) of our Tertiary University Hospital after a successfully resuscitated cardiopulmonary arrest. He was diagnosed at the age of six years with DMD and was a wheelchair-dependent adolescent for the last two years. He had severe scoliosis with quite normal pulmonary function, and he was a high-performance student. Three days before the arrest, he had flu-like symptoms (low grade fever up to 38 ^{o}C and cough) and he was treated symptomatically. The day of his admission, he was found

cyanotic and pulseless by his mother. He recovered after 20 minutes of advanced cardiopulmonary resuscitation and was admitted, intubated, to our ICU. On admission, he was hypotensive, with adequate ventilation and oxygenation (pO_2: 149 mmHg, receiving a 0.40 fraction of inspired oxygen), under mechanical ventilation. His vital signs were as follows: temperature 36.6 ^oC, pulse rate 135/min, blood pressure 75/50 mmHg and pupils of equal size with normal light reflex. Arterial blood gas analysis revealed metabolic acidosis (pH: 7.26, pCO_2: 38 mmHg, HCO_3^-: 15 mmol/L, Lactate: 2.1 mmol/L, anion gap: 25 mEq/L), and severe hypokalemia (K: 1.3 mEq/L), which was confirmed by biochemical analysis by our laboratory (K: 1.4 mEq/L). All other biochemical values, including urea, creatinine, and magnesium, were normal. Potassium deficit was repleted at an initial rate of 40 mEq/h infusion. The electrocardiogram revealed sinus tachycardia with right bundle branch block and depression of ST-segment in leads V_4-V_6. The echocardiographic examination revealed impaired contractility of the left ventricle with diffuse hypokinesia and normal diastolic function. Computed tomography scan of the brain and thorax revealed mild brain edema and lung infiltration on the right pulmonary lobe, compatible with the presence of lobar pneumonia. The patient received empiric treatment for community-acquired pneumonia with azithromycin and ceftriaxone and antiviral agent oseltamivir phosphate. After the initial six hours of aggressive potassium repletion, the rate of correction was gradually decreased. Hypokalemia was resistant to treatment and was eventually corrected on the 2nd day. The patient had a prolonged hospitalization due to hypoxic encephalopathy and weaning failure from mechanical ventilation and was discharged to home from the ICU two months later on mechanical ventilation and gastrostomy feeding. Three months later, the patient was in a minimally conscious state and he was weaned from mechanical ventilation, being able to sustain spontaneous breathing on T- piece.

Discussion

In the reported case, a young DMD patient suffered a cardiac arrest due to severe hypokalemia following a lower respiratory tract infection. The patient had no previous history of hypokalemia, no episodes of diarrhea or vomiting, and did not receive medications causing hypokalemia i.e. diuretics, laxatives, or antipsychotic drugs. To the best of our knowledge, only three previous reports describe DMD patients with clinically significant hypokalemia; two of them were associated with respiratory tract infection[11,12].

According to the formulated hypothesis, DMD patients have diminished total body potassium, which appears to correlate with the severity of muscular involvement[13] and not with renal wastage of potassium[14]. In fact, there are some studies, although old, documenting diminished total body potassium[13,15,16] in DMD patients and their non-dystrophic parents and siblings. Total body potassium may be decreased[13] or within the normal range[16] initially, and gradually decreasing as the disease progresses. It's decreased levels have been considered to be associated with the loss of functional muscle mass. A study reported that the total body potassium reflects lean body mass which is mainly composed of rich in potassium muscle and therefore potassium deficiency in muscular dystrophy is only the consequence of the wasting of dystrophic muscle[15]. Other investigators demonstrated that total body potassium depends not only on muscle bulk but also on intracellular potassium concentration in affected muscle cells[17]. On the contrary, Blahd et al demonstrated that intracellular potassium concentration in dystrophic patients is normal and supported that the low levels of total body potassium are the results of the replacement of muscle cells by collagenous tissue[13]. The finding that nondystrophic relatives had also reduced body potassium levels suggests that hypokalemia might be genetically determined[18]. Regardless of the diminished total body or intracellular potassium concentration in DMD patients, the level of serum potassium is generally normal[12]. It has been suggested that patients with advanced DMD develop hypokalemia from minor insults, such as vomiting, diarrhea, fever, that have little effect on healthy subjects[11].

Although there is not an obvious cause for severe hypokalemia in the reported patient, we postulated it was developed due to the combined effects of the respiratory infection, dehydration, and the already existent abnormal potassium reservoir and that it provoked severe arrhythmia leading to the cardiac arrest. However, although dilated cardiomyopathy is a frequent manifestation in DMD patients, we did not find any ultrasound evidence of overt cardiomyopathy in this patient. If we take into consideration the fact that hypokalemia causes weakness of the respiratory muscles[12], and the coexistence of marginal pulmonary function[19] due to severe scoliosis, we could hypothesize that the severe hypokalemia might have additionally contributed to the cardiorespiratory failure.

With this case report, we emphasize a rare but potentially lethal complication which can occur in DMD patients during pathological conditions, such as infections. Available literature for hypokalemia complicating DMD patients is sparse and limited to older studies. Recognizing that severe hypokalemia, although uncommon, might occur in DMD patients with lethal consequences, it may be beneficial to exhibit more attention in maintaining serum potassium levels in the normal range. Therefore, patients with DMD should be monitored for potassium levels, and potassium intake should be adjusted accordingly. They should be encouraged to follow a high-potassium diet or to take oral supplementation if necessary. The risk of a fatal arrhythmia and sudden cardiac death is rare but real; therefore, more attention should probably be given to the potassium homeostasis in patients with DMD.

Conflict of interest

Authors declare no conflict of interest.

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