Survival After Cardiac Arrest With Instantaneous Rigorlike Stiffness: A Case Report

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Instantaneous rigor is the immediate appearance of rigor mortis after cardiac arrest. To our knowledge, no previous reports exist on resuscitation of such patients. A young athlete suddenly collapsed with cardiac arrest during a marathon; his legs stiffened with instantaneous rigorlike stiffness. This stiffening provoked hyperkalemia, rhabdomyolysis, and multiple organ failure. We decided to amputate both legs, with venoarterial extracorporeal membrane oxygenation support. The patient recovered and was discharged without neurologic impairment. This rare case highlights the potentially significant effect of instantaneous rigor. [Ann Emerg Med. 2019;73:393-396.]

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INTRODUCTION

Background

Instantaneous rigor, also known as accelerated rigor mortis or cadaveric spasm, is a rare condition in which extremely rapid muscle stiffening occurs after cardiac arrest, in contrast to normal rigor mortis that gradually occurs 3 to 6 hours after cardiac arrest.[1-3] Reports describe instantaneous-rigor cases in forensic medicine and a few reports exist in emergency medicine,[3,4] but all these reports discuss patients who died. To our knowledge, no reports describe therapeutic intervention.

CASE REPORT

A 14-year-old healthy boy suddenly collapsed after running 14 km of a marathon and was transferred to our emergency department, with basic life support. On arrival, he was in cardiac arrest (asystole). We immediately decided to apply extracorporeal cardiopulmonary resuscitation, commencing 12 minutes after arrival (22 minutes after collapse). Soon afterward, we found that both his legs were entirely stiffened, as in rigor mortis (Figure 1*A* and *B*). Muscle relaxants and sedative agents did not improve the stiffness. His first blood gas assessment revealed hyperkalemia (9.7 mmol/L), metabolic acidosis (pH 6.99, bicarbonate level 9.1 mmol/L, base deficit –22.3 mmol/L, and lactate level 18 mmol/L), and hypoglycemia (44 mg/dL). After confirming no obvious sign of hypoxic brain damage with computed tomography (CT), we initiated post–cardiac arrest care in the critical care unit. Despite correction of the acidosis (pH 7.27, base deficit –8.0, and lactate level 10.9 mmol/L) and hemodynamic management with venoarterial extracorporeal membrane oxygenation, the hyperkalemia worsened to 14.2 mmol/L within 2 hours

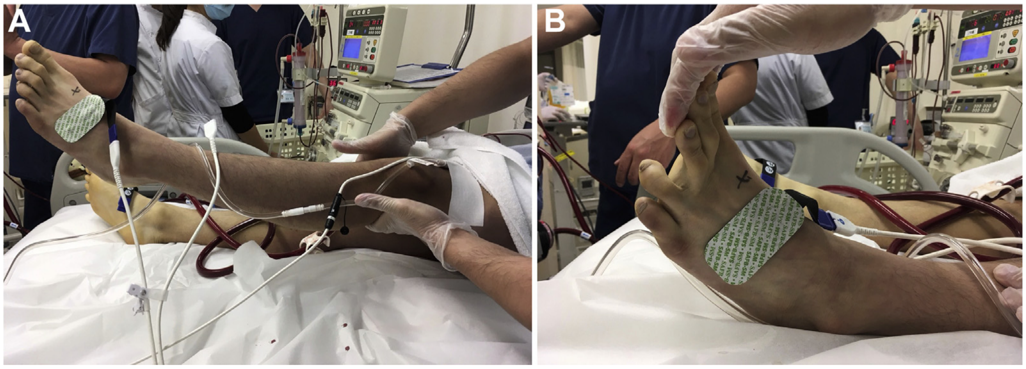
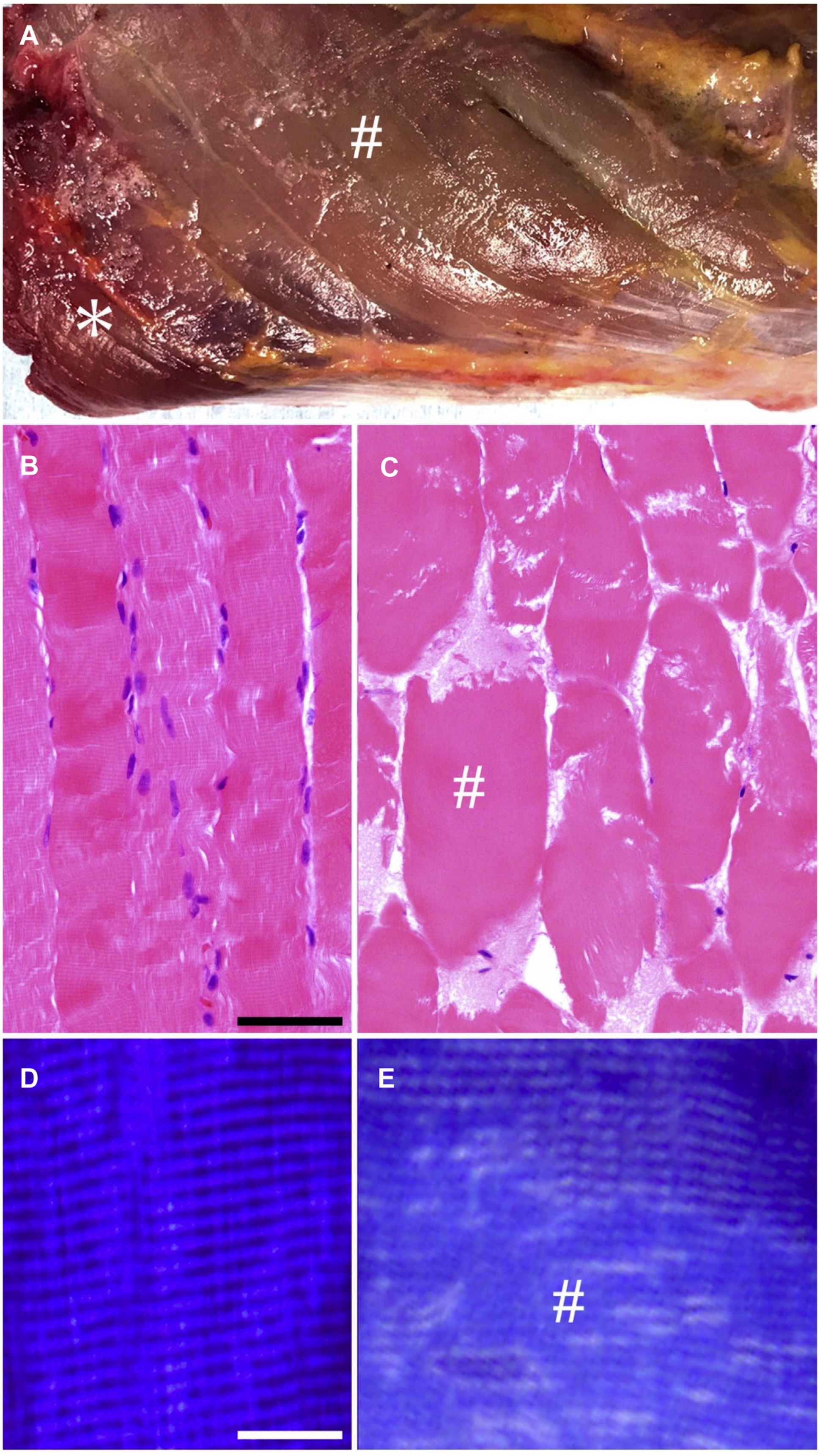


Figure 1. Physical findings on admission. *A*, The legs were entirely stiffened. *B*, The ankle and toes joints were not bendable.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of admission. Moreover, the serum creatine phosphokinase level increased to 50,305 U/L, and myoglobinuria was detected. The serum ionized calcium level gradually decreased from 1.17 mmol/L on admission to 0.73 mmol/L. The next day, both his legs from the hip to the toes were completely stiff, with creatine phosphokinase levels elevated to 500,000 U/L despite continuing hemodiafiltration.

On the second day after admission, the leg stiffness reduced slightly and the legs could be flexed at the knee to 20 to 30 degrees passively. His creatine phosphokinase level increased to more than 800,000 U/L despite continuing hemodiafiltration, and the extracorporeal membrane oxygenation flow rate had to be continuously increased to sustain organ perfusion. We hypothesized that multiple organ failure was caused by reperfusion of necrotic tissue in both legs. We repeatedly discussed therapeutic strategies, as well as the ethical dilemma, with a multidisciplinary team and his family. His parents hoped to save his life by any means, so we finally decided to amputate both legs while providing ongoing venoarterial extracorporeal membrane oxygenation and hemodiafiltration. After amputation, the patient's creatine phosphokinase and myoglobin levels rapidly decreased and his general condition improved. On the fifth day after admission, the extracorporeal membrane oxygenation support was weaned off.

Macroscopic findings of the amputated legs showed that most of the upper and lower leg muscles had lost their elasticity and were pale, but with some parts remaining red (Figure 2*A*). Pathologic changes were prominent in the flexor muscles, especially in the lower leg muscles such as the soleus muscle. Microscopic examination revealed that the uninvolved muscle retained a normal appearance, including elongated nuclei at the periphery of the myofibers and cross striations, as shown on phosphotungstic acid–hematoxylin staining (Figure 2*B* and *D*). The involved area showed destruction of the normal muscle structures, resulting in a homogeneous and amorphous appearance on hematoxylin and eosin staining (Figure 2*C* and *E*).

The contrast-enhanced CT findings on the sixth day after admission revealed that his lower back muscles—the erector spinae, psoas, gluteus maximus, and the remnants of the amputated quadriceps muscles—showed a lower density than the upper body muscles. All the above-mentioned muscles atrophied and calcified 4 weeks later.

On day 22 after admission, he was discharged from the critical care unit to the general ward. His neurologic function recovered fully. He understood and accepted his amputations and tried rehabilitation. A few weeks later, he received a diagnosis of dilated cardiomyopathy in another hospital.

DISCUSSION

We report a case with cardiac arrest and sudden stiffness of both legs, mimicking instantaneous rigor. This is, to the best of our knowledge, the first reported case of resuscitation of a patient who developed instantaneous rigorlike stiffness. The stiffened legs provoked severe hyperkalemia, rhabdomyolysis, and multiple organ failure, resulting in a decision to amputate both legs.

Rapid muscle stiffening immediately after cardiac arrest is called *instantaneous rigor*. It is a very rare phenomenon, occasionally observed in cases of cardiac arrest that occur during intensive muscular activity such as running or fighting on a battlefield.[1-3] Some previous case reports[1,2] have prompted discussion of whether instantaneous rigor is a genuine pathologic phenomenon or whether it is a continuation of premortem muscular activity, such as when a gun or knife is gripped just before death. Other case reports do not follow such activities, suggesting that instantaneous rigor is a distinct phenomenon.[3-5]

We believe that the stiffness of both legs observed in our case was caused by a mechanism similar to that of instantaneous rigor. In healthy human beings, adenosine triphosphate hydrolysis is an essential mechanism for muscle relaxation. It destabilizes the myosin-actin bond and breaks the cross bridge to enable muscle relaxation.[1] In the postmortem period, the adenosine triphosphate concentration diminishes and is insufficient for relaxation. Therefore, existing muscle contractions last as rigor mortis until the decomposition process induces muscle softening. Although rigor mortis usually begins approximately 3 hours after death, when the adenosine triphosphate concentration has reduced sufficiently, it sometimes progresses rapidly in conditions wherein the premortem adenosine triphosphate concentration in the muscle is already low because of

Figure 2. Representative macroscopic and microscopic findings of the amputated muscles. *A*, Macroscopic findings of the biceps femoris muscle, with a majority of the involved muscles pale (pound sign) and a minority of uninvolved muscles reddish (asterisk). Histologic images of the uninvolved muscle (*B*) and involved muscle (*C*). A pound sign highlights the homogeneous and amorphous appearance without nuclei and cross striations in the involved area. Hematoxylin and eosin staining (bar=50 μm). High-magnification images of the uninvolved area (*D*) and involved area (*E*). A pound sign highlights the indefinite cross striations. Phosphotungstic acid–hematoxylin stain (bar=10 μm).

previous consumption; for example, in intensive muscular activity such as long-distance running before cardiac arrest.[1-3,6,7] Additionally, deletion of glycogen or ionized calcium concentration, acidemia with an increased lactate level, and the soleus muscle of the lower extremity are also associated with rapid progression of rigor mortis.[1,7,8] Our patient had almost all the above-mentioned conditions. Moreover, the stiffness began to reduce on day 2, which is when rigor mortis begins to decrease in cadavers.[1] In line with these findings, although we are inevitably unable to provide the definitive data such as adenosine triphosphate measurement, we believe it is reasonable to hypothesize that the stiffness of the legs in this case occurred because of mechanisms similar to those in instantaneous rigor.

This case also highlights that stiffened legs can be a cause of multiple organ failure. We suggest that instantaneous rigorlike stiffness provoked rapid muscle breakdown and that the cellular components released into the extracellular fluid acted as damage-associated molecular patterns, resulting in multiple organ failure.[8-13] Thus, although evidence for this procedure is inevitably very limited, amputation of stiffened parts of the body was considered as a therapeutic option. The decision to amputate the legs of an unconscious patient is a serious ethical dilemma because of the psychological trauma associated with an unanticipated amputation. Engaging in discussions with the family to ensure that such treatment is likely to be in accordance with the patient's wishes is essential, as is psychological support.

Conclusion

We describe a case wherein the patient was resuscitated after cardiac arrest, with both legs stiffened with instantaneous rigor and with rhabdomyolysis, leading to refractory multiple organ failure. This rare case highlights the potentially significant effect of instantaneous rigor.

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REFERENCES

1. Saukko P, Knight B. *Knight's Forensic Pathology.* 4th ed. Boca Raton, FL: CRC Press; 2015.
2. Bedford PJ, Tsokos M. The occurrence of cadaveric spasm is a myth. *Forensic Sci Med Pathol*. 2013;9:244-248.
3. Mesri M, Behzadnia M, Dorooshi G. Accelerated rigor mortis: a case letter. *J Res Med Sci*. 2017;22:126.
4. Lee JH, Jung KY. Emergency cricothyrotomy for trismus caused by instantaneous rigor in cardiac arrest patients. *Am J Emerg Med*. 2012;30:1014.e1011-2.
5. Fierro MF. Cadaveric spasm. *Forensic Sci Med Pathol*. 2013;9:251-252.
6. Kobayashi M, Takatori T, Nakajima M, et al. Does the sequence of onset of rigor mortis depend on the proportion of muscle fibre types and on intra-muscular glycogen content? *Int J Legal Med*. 1999;112:167-171.
7. Kobayashi M, Takatori T, Nakajima M, et al. Onset of rigor mortis is earlier in red muscle than in white muscle. *Int J Legal Med*. 2000;113:240-243.
8. Chavez LO, Leon M, Einav S, et al. Beyond muscle destruction: a systematic review of rhabdomyolysis for clinical practice. *Crit Care*. 2016;20:135-146.
9. Gonzalez D. Crush syndrome. *Crit Care Med*. 2005;33:S34-S41.
10. Wei Q, Baihai S, Ping F, et al. Successful treatment of crush syndrome complicated with multiple organ dysfunction syndrome using hybrid continuous renal replacement therapy. *Blood Purif*. 2009;28:175-180.
11. Sever MS, Vanholder R. Crush syndrome: a case report and review of the literature. *J Emerg Med*. 2015;48:730-731.
12. Kawai C, Kotani H, Miyao M, et al. Circulating extracellular histones are clinically relevant mediators of multiple organ injury. *Am J Pathol*. 2016;186:829-843.
13. Kono H, Rock KL. How dying cells alert the immune system to danger. *Nat Rev Immunol*. 2008;8:279-289.