Acute myopathy leading to severe rhabdomyolysis: an unusual presentation

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Sri Lanka Journal of Child Health, 2021; **50**(1): 165-167
DOI: http://dx.doi.org/10.4038/sljch.v50i1.9422
(Keywords: Acute rhabdomyolysis, acute myopathy)

Introduction

Rhabdomyolysis means "dissolution of skeletal muscle" and is the end result of several processes which cause skeletal muscle injury and subsequent release of creatine kinase (CK), myoglobin, potassium and phosphorus into the plasma[1]. We present a 14 month old girl with acute myopathy and rhabdomyolysis, who, in spite of very high levels of CK, made a full recovery and is developing normally.

Case report

A one year and two month old girl was brought to the accident and emergency department with a history of acute generalised weakness of around 12 hours duration. She had been completely well before this episode. The preceding day she had breakfast, refused lunch and had drunk a bottle of milk before going to bed. She woke up the following morning and her mum noticed her to be floppy and lethargic. There was no impairment of consciousness. The child has no known medical problems, was vaccinated up to date and was not on any medications. The family members are all fit and well and do not take any prescribed or over the counter medications. The family have not travelled abroad to any exotic locations. The child had a very short history of being generally unwell with food refusal. She was brought to the hospital because of her generalized weakness which was slowly getting worse and the family were very anxious about the child's condition. The child was seen by the paediatric team and was admitted for observation as the child's weakness was progressively worsening.

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(Received on 20 April 2020: Accepted after revision on 22 May 2020)
The authors declare that there are no conflicts of interest
Personal funding was used for the project.

On admission, the child was miserable and listless with no focal neurological deficit. Since the child did not have any choking or drooling and was maintaining the airway, it was decided to observe the child in the high dependency unit and investigate as appropriate. At presentation, the child had a power of 5/5, deep tendon reflexes (DTR) were present and there was gradual deterioration, the lowest level of power being 1/5. However, DTR were present during the entire period of hospital stay right up to hospital discharge. She was apyrexial and demonstrated severe hypotonia together with truncal and limb muscle weakness. No muscle tenderness was noted. The eye movements were normal and no ptosis was present. There were no clinical signs of seizures.

Urine dipstick demonstrated the presence of albumin, ketone and myoglobin. The urine only had 1+ albumin and this too was transient. Her serum CK was 2993 units/L (normal range 22 to 198 U/L). Her plasma potassium was 4.7mEq/L. Serum creatinine was 37mg/dL. Over the next 24 hours she became more and more lethargic and her CK increased to 16,720 U/L and later to 430,000U/L. The deteriorating respiratory function and weakness of respiratory muscles necessitated supportive airway support and invasive ventilation. She was intubated and ventilated for a week and then extubated. While being ventilated she was noted to have a persistent tachycardia (150-160/min) and a systolic blood pressure ranging from 110-130mmHg (95th centile 110 mmHg). The hypertension was attributed to idiopathic causes since the child was extensively investigated and the blood pressure returned to normal range post intensive care unit (ICU) stay. Her CK gradually decreased to 368,714 U/L (day 3), 2047 U/L (day7) and 663 U/L (day 9). During this phase her renal functions were within normal limits. In her outpatient follow up she had regained her motor functions completely and she continues to achieve normal developmental milestones.

The following investigations were done:

- Complete blood count
- Renal function (daily screening for albumin)
- Liver function tests. The serum aspartate transaminase (AST) was normal.

- Muscle enzyme tests: serum CK as shown above was very high. Isotypes of CK like CK-MB were not done. AST, done as part of the liver function tests, was normal but serum aldolase was not done.
- Cerebrospinal fluid (CSF) - Culture negative, microscopy showed no cells, normal opening pressure
  Herpes and enterovirus polymerase chain reaction (PCR) in CSF- Negative,
- Immunoglobulins- IgA 0.26 (Normal range 0.33-1.85)
- Anti-neutrophil cytoplasmic antibodies (ANCA), anti-deoxyribonuclease B (Anti-DNA se B) -Negative
- Echocardiography
- Computed tomography (CT) scan and magnetic resonance imaging (MRI) of brain
- Electromyography (EMG) - evidence of myopathy
- Electroencephalography (EEG)
- Renal ultrasound and dimercapto succinic acid (DMSA) scan
- Serum organic acids and amino acids
- Plasma ammonia, renin, aldosterone, angiotensin and cortisol
- Deoxyribonucleic acid (DNA) for malignant hyperthermia screens
- Red blood cell (RBC) acetyl cholinesterases
- Serum carbamate, plasma metanephrines
- Urine amino acids and organic acids
- Urine toxicology and drugs screen
- Urine porphyria screen
- Urine 4-hydroxy-3-methoxyamphetamine (HMA)/creatinine
- Homovanillic acid (HVA)/creatinine ratio,
- 4-hydroxy-3-methoxy methamphetamine (HMMA)/creatinine ratio
- Screening for fatty acid oxidation defect was negative (normal free carnitine and acyl carnitine profile).

The child showed progressive deterioration neurologically, primarily her airway and breathing being involved and hence needing acute respiratory support, but with complete recovery without any neurological deficit.

Discussion

Data on rhabdomyolysis in children are scanty. The characteristic clinical features are myalgia, muscle weakness, and muscle swelling which develop over hours to days[2,3]. Two small case series, each with less than 20 patients, reported acute renal failure (ARF) rates of 42%[4] and 50%[5]. However, a much larger series comprising 191 children reported ARF in only 4.7% of children with rhabdomyolysis[1]. In our patient except for transient albuminuria there was no evidence of renal involvement.

Rhabdomyolysis is diagnosed primarily on the basis of myoglobinuria or grossly elevated serum CK levels[6]. Following muscle injury, myoglobinuria generally disappears within 24 hours whereas serum CK levels peak at 3–5 days after injury, and decrease over the next 6–10 days[6]. CK elevation 5 times the upper limit of normal is the defining biochemical abnormality[3]. This was present in our child.

In the study by Mannix R, *et al*[1] the common causes of paediatric rhabdomyolysis were viral myositis (38%), trauma (26%), and connective tissue disease (5%). Hypokalaemia-associated rhabdomyolysis has been reported in a 10 year old boy on amphotericin B[7]. Acute rhabdomyolysis can also occur as a complication of acute respiratory failure secondary to status asthmaticus[8]. In our patient, though investigated extensively, a cause was not identified and parents declined muscle biopsy. However, viral myositis should be considered as a possible aetiology in our patient.

Acknowledgements

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