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Severe neonatal hyperkalemia presenting as life threatening arrhythmias in the pediatric emergency department: Case report

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Abstract
Context: Severe hyperkalemia is rarely seen in newborns, especially with critically high serum potassium. Hyperkalemia is a life-threatening condition; it requires early recognition and aggressive treatment. Severe hyperkalemia leads to a life-threatening arrhythmia and asystole.
Objective: To report a case of a newborn infant with vomiting, history of lethargy, decreased feeding and activity, and infected skin lesions in the face. The case was reported with severe life-threatening neonatal hyperkalemia (K 12.7 mmol/L) and associated with ventricular tachycardia converted by synchronized cardioversion. Ultimately diagnosed with pseudohypoaldosteronism.
Patient: This case was presented to the ED at the age of 14 days in septic shock. There was a family history of infant death. A two boys' siblings died during infancy after being diagnosed with autosomal recessive Pseudohypoaldosteronism type 1(PHA1). Our patient had ventricular tachycardia in ED due to severe hyperkalemia; while receiving fluid boluses, cardiopulmonary resuscitation, hyperkalemia treatment, and synchronized cardioversion were initiated. After stabilization, diagnostic workup demonstrated persistently low sodium, metabolic acidosis, and high potassium, which may require dialysis. Based on these results and family history, the patient was diagnosed with PHA1.
Conclusions and Outcome: This case highlights the significant hyperkalemia is a life-threatening electrolyte disorder and the vital cause of arrhythmias and a medical emergency that require urgent intervention in newborns and infants. Prompt treatment of hyperkalemia is imperative to prevent the further development of fatal cardiac arrhythmia.

Keywords: hyperkalemia, arrhythmias, synchronized cardioversion, pseudohypoaldosteronism

Introduction

Neonatal hyperkalemia is a medical condition that requires an emergent diagnosis and treatment to avoid morbidity and mortality. The causes of hyperkalemia in infancy include acute hemolysis, kidney disorders, and hormonal disorders. Common hormonal disorders leading to hyperkalemia in neonates and infants include congenital adrenal hyperplasia (CAH) and adrenal insufficiency, while pseudohypoaldosteronism (PHA) remains a rare cause. PHA can result in severe hyponatremia, hyperkalemia, and metabolic acidosis, which if undetected and untreated may cause neonatal mortality [1]. Hyperkalemia is a fundamental cause of arrhythmias and a medical emergency that need urgent treatment. The cause is usually multifactorial. It is most commonly caused by impaired potassium secretion, transcellular potassium shifts, and an increased potassium load [2]. The serum potassium concentration is tightly maintained within the range of 3.5–5.5 mmol/L. The threshold may be considerably higher in neonates (up to 6.5 mmol/L), especially if they are born prematurely [3]. Because hyperkalemia can cause lethal cardiac arrhythmia, it is one of the most severe electrolyte disturbances [2]. Hyperkalemia is usually caused by the concurrence of various factors: increased potassium intake, excretion disorders, and transcellular shifts [4]. In neonates, inborn errors of metabolism as congenital adrenal hyperplasia (CAH) and Pseudohypoaldosteronism (PHA) are a leading cause of severe hyperkalemia [4]. Hyperkalemia destabilizes myocardial conduction by decreasing the resting membrane potential, increasing cardiac depolarization and myocardial excitability [5]. It can cause a spectrum of adverse ECG abnormalities, from peaked T waves, beginning with levels above 6.5 mmol/L, to shortened QT intervals, prolonged PR intervals, heart conduction blocks, depressed P waves, and vast QRS complexes with levels 7.5-8 mmol/L, which tend to merge with the peaked T wave into 'sine-wave activity [6]. Severe hyperkalemia (above eight mmol/L) can precipitate fatal arrhythmias such as ventricular tachycardia and fibrillation, leading to asystole [6]. This study has reported a case of life-threatening severe neonatal hyperkalemia (with potassium level = 12.7 mmol/L with electrocardiogram changes (Ventricular Tachycardia) that converted with treatment and synchronized cardioversion in our pediatric emergency. We present the case of a 14-day-old boy who presented to the emergency department (ED) with life-threatening hyperkalemia and ventricular tachycardia that required hyperkalemia treatment, and synchronized cardioversion. He was treated for septic shock and CAH but did not respond to treatment, and it found that the hyperkalemia was due to PHA rather than CAH. The patient ultimately required aggressive hyperkalemia management to control his hyperkalemia.

Case Report

The patient came to the pediatric emergency department at the age of 14 days in septic shock. He was a child of

consanguineous parents and had a birth weight of 2.5 kg. There was a family history of infant death. A two boys' siblings died during infancy after being diagnosed with autosomal recessive Pseudohypoaldosteronism type 1(PHA1). Our patient presented to ED with a history of lethargy, decreased feeding and activity, and infected skin lesions in the face. While extracting the blood samples, the cardiac monitor showed a wide QRS complex, indicating ventricular tachycardia (figure 1). The immediate blood gas results showed pH = 6.9; serum bicarbonate = 6 mmol/L; hyperkalemia (14 mmol/L), hyponatremia (120mmol/L). The patient was promptly started on hyperkalemia treatment, including intravenous calcium gluconate, nebulized salbutamol, insulin, glucose infusion, and intravenous sodium bicarbonate, then converted to sinus tachycardia immediately after intravenous calcium gluconate. Initial laboratory findings showed hyperkalemia (12.7 mmol/L), hyponatremia (124 mmol/L), and acidosis. CBC showed WBC 35.6 Hb 13.7 g.dl PLT 665, serum lactate 13.6 mmol/L. We considered septic shock and adrenal insufficiency due to CAH because of the patient's hyperkalemia, hyponatremia, acidosis, and family history of infant death with consanguineous parents. The patient received three boluses of normal saline and started on vancomycin and cefotaxime and hydrocortisone IV. Before shifting the patient to the pediatric intensive care unit (PICU), he started again to have ventricular tachycardia on a cardiac monitor and 12 lead ECG (figure 2). The patient was immediately started on intravenous calcium gluconate, nebulized salbutamol, insulin, glucose infusion, intravenous sodium bicarbonate, and oral calcium resonium but still had persistent ventricular tachycardia. Hence, the amiodarone loading dose started but was not converted then the patient was sedated and received four shocks of synchronized cardioversion. The rhythm converted immediately to normal sinus rhythm after the 4th shock, then started on amiodarone infusion as advised by a pediatric cardiologist. Echocardiography done in ED showed normal cardiac function. After that, the intubation was performed, and the patient stabilized then shifted to PICU. After admission, the patient did not develop arrhythmia and was started on hydrocortisone and fludrocortisone by the endocrinologist as a suspected case of CAH. After 24 hrs, the patient responded to the treatment; his potassium fell to 7.6 mmol/L, his sodium improved to 130 mmol/L. After two days patient clinically improved and extubated, potassium fell to 5.1 mmol/L, sodium 138 mmol/L. He shifted from PICU to special care unit with a good clinical condition, on fludrocortisone, oral sodium bicarbonate, and oral hypertonic saline. Last laboratory results showed: Ph = 7.38, Hco3 = 18, K = 4.5 mmol/L and Na = 136 mmol/L. The patient was responding to treatment, but after two days, the serum potassium rising again to 7.29 mmol/L and becoming refractory and rising to 9.65 mmol/L and 11 mmol/L. The patient was again admitted to PICU due to this refractory hyperkalemia and ultimately may require dialysis to control his hyperkalemia but responded to hyperkalemia management, and potassium fell to 4.5 mmol/L. Again he shifted from PICU to special care unit with a good clinical condition, on oral sodium bicarbonate, potassium exchange resins, and oral hypertonic saline Laboratory studies obtained after admission subsequently revealed serum aldosterone markedly elevated at 312.40 ng/dl (range 1.72–23.2 ng/dl), serum cortisol markedly elevated at 1229 nmol/L (range 137.9–634.5), and normal ACTH 26.5 ng/L (range 5.0-60.0). These results ruled out CAH as the etiology for the infant's hyperkalemia and physiologic collapse. The patient's diagnosis of PHA type 1 was assigned depending on the family history, elevated cortisol, and aldosterone levels.

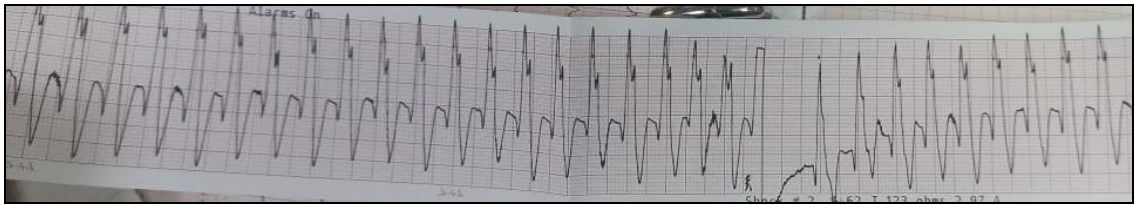

Fig 1

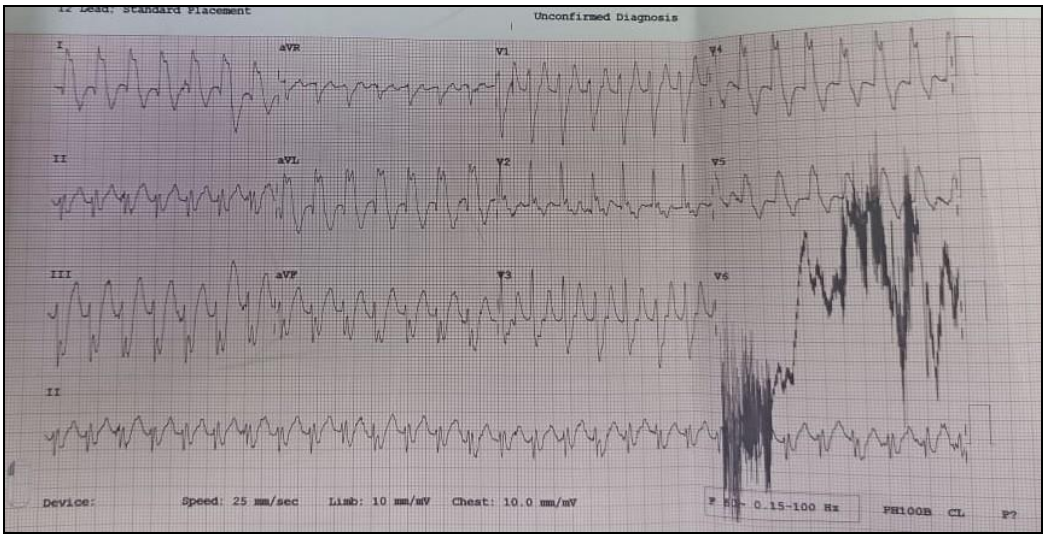

Fig 2

Discussion

We report a motivating clinical case of a newborn with severe hyperkalemia that led to a life-threatening arrhythmia. Such severe hyperkalemia is rarely seen in newborns, especially with critically high serum potassium [2]. Hyperkalemia is a life-threatening condition; it requires early recognition and aggressive treatment. Neonatal hyperkalemia is a medical condition that requires an emergent diagnosis and treatment to avoid morbidity and mortality [1]. Both CAH and Pseudohypoaldosteronism type 1(PHA1) are present in the neonatal stage with a similar clinical presentation (poor feeding, lethargy, and dehydration) and laboratory findings suggestive of adrenal crisis (hyperkalemia, hyponatremia, and acidosis). Management with hydrocortisone should be started in all patients with no index case of PHA because hydrocortisone results in a good response in the case of CAH [7]. Our patient is male, serum aldosterone markedly elevated at 312.40 ng/dl (range 1.72–23.2 ng/dl), serum cortisol 1229 nmol/L (range 137.9–634.5) and normal ACTH 26.5 ng/L (range 5.0-60.0). These findings assigned the diagnosis of PHA type as the cause for the infant's physiologic collapse and hyperkalemia. Aldosterone is a steroid hormone that regulates potassium and sodium homeostasis; it acts primarily on epithelial cells in the renal collecting ducts [8]. The electrolyte abnormalities of hyponatremia, hyperkalemia and metabolic acidosis are similar in CAH and PHA. However, in contrast to CAH, serum cortisol and aldosterone levels are elevated in PHA, indicating normal glucocorticoid production and resistance to aldosterone activity. The patient presented here clinically fits the picture of systemic PHA1 with severe refractory hyperkalemia that may require dialysis at diagnosis and continued requirement for salt supplementation. Management of PHA1 should be considered as an emergency and life-saving measure. Treatment of PHA1 comprises adequate rehydration, salt loss replacement, and correction of hyperkalemia and acidosis in the acute phase. After initial stabilization, potassium exchange resins and salt supplementation are the mainstays of treatment. To our information, our case is one of the first cases reported with severe life-threatening neonatal hyperkalemia (K 12.7 mmol/L) and associated with ventricular tachycardia converted by synchronized cardioversion.

The patient stabilized and had no cardiac arrest. The case reported by Kavčič *et al*. exhibited salt-wasting like our patient, but unlike ours, the serum potassium level of their case was nine mmol/L [1]. One of the cases reported by Attia & Marzouk, 2016 [7] exhibited hyperkalemia like our patient. Still, unlike ours, the serum potassium level of their case was 8.6 mmol/L, and the child developed cardiac arrest [7]. The case reported by Sakar *et al*. exhibited like our patient, but unlike ours, the serum potassium level of their case was 8.8 mmol/L [9]. Also, one of the cases reported by Schweiger *et al*. exhibited hyperkalemia like our patient, but unlike ours, the serum potassium level of their case was nine mmol/L [10]. So, our case is the first case reported with severe life-threatening neonatal hyperkalemia (K 12.7 mmol/L) associated with ventricular tachycardia converted by synchronized cardioversion and patient stabilized. The patient had no cardiac arrest. The patient's diagnosis of PHA type 1 was assigned depending on the family history, elevated cortisol, and aldosterone levels.

In conclusion, hyperkalemia is a life-threatening electrolyte disorder and the vital cause of arrhythmias and a medical emergency that require urgent intervention in newborns and infants. Prompt treatment of hyperkalemia was imperative to prevent the further development of fatal cardiac arrhythmia. First, intravenous calcium gluconate should be infused in patients with electrocardiographic changes to stabilize membrane potential. The insulin with dextrose infusion, intravenous or nebulized beta-adrenergic agonists (salbutamol), intravenous sodium bicarbonate, or oral calcium resonium can increase potassium shift from extra- to intracellular space [11].

Abbreviations

PHA1: Pseudohypoaldosteronism type 1
PHA: Pseudohypoaldosteronism
CAH: Congenital adrenal hyperplasia
ED: Emergency department
PICU: Pediatric Intensive Care Unit
ECG: Electrocardiogram

Funding

None

Conflict of interest

None declared

Disclosure

Abdullah M. Kamel is a Pediatric Emergency Consultant. King Saud Medical City, Riyadh; he received and managed the patient in ED, and reviewing patients' files.

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