

Case report

Respiratory muscle paralysis in a patient with hypokalemia associated with Southeast Asian ovalocytosis: A case report.Sameera Narasinghe^{1*}, Chamara Sarathchandra², Hemal Senanayaka², Prasanna Weerawansa²¹Postgraduate Institute of Medicine, University of Colombo, Sri Lanka.²Faculty of Medicine and Allied Sciences, Rajarata University of Sri Lanka, Saliyapura, Sri Lanka.**Abstract**

Hypokalemic periodic paralysis is a rare clinical syndrome characterized by low serum potassium and severe muscle weakness. The aetiology can be primary idiopathic or secondary to other disorders such as thyroid dysfunction and renal tubular acidosis. Hypokalemic periodic paralysis with distal renal tubular acidosis (RTAd) is seen in a cluster of the population in the North-central province of Sri Lanka particularly in association with Southeast Asian Ovalocytosis (SEAO). Here, we report the first case of severe respiratory muscle paralysis in a patient having RTAd with severe hypokalemia with associated SEAO from the North Central province, Sri Lanka.

Keywords: Hypokalemia, Renal tubular acidosis, Type 2 respiratory failure, Southeast Asian ovalocytosis**Copyright:** © 2022 Narasinghe S *et al.*  This is an open-access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.**Funding:** none**Competing interests:** None**Received:** 26.01.2021**Accepted revised version:** 22.05.2022**Published:** 15.07.2022*✉ **Correspondence:** nsajithsameera@yahoo.com. <https://orcid.org/0000-0002-6001-6472>

Cite this article as: Narasinghe S *et al.*, Respiratory muscle paralysis in a patient with hypokalemia associated with Southeast Asian ovalocytosis: A case report. *Anuradhapura Medical Journal* 2022; 16 (2): 41-43, DOI: <http://doi.org/10.4038/amj.v16i2.7681>

Introduction

Periodic paralysis is a neuromuscular disorder characterized by a defect in membrane ion channels presenting episodic muscle paralysis. Although hyperkalemia and thyrotoxicosis can cause periodic paralysis, hypokalemia is more common to cause periodic paralysis, with a prevalence of 1 in 100 000 population (1). Presentation with respiratory muscle paralysis requiring mechanical ventilation in hypokalemic periodic paralysis was not reported before. Here we report a case of hypokalemic periodic paralysis requiring ventilatory support secondary to renal tubular acidosis. The patient also had Southeast Asian ovalocytosis.

Case report

A 37-year-old lady from Anuradhapura, North Central Province of Sri Lanka presented to the emergency department with difficulty in walking upon waking up in the morning. She had experienced marked difficulty in

moving her legs with slight difficulty in moving her upper limbs, but there was no difficulty in breathing or dysphagia. There were no previous similar episodes, and there was no preceding diarrheal or respiratory illness. She had no hair loss, fever, arthritis, weight loss, palpitation, or heat intolerance. She was the 3rd child of a healthy family with no familial comorbidities.

On examination, she had a marked bilateral symmetrical proximal and distal grade two lower limb weakness and bilateral symmetrical grade three upper limb weakness. The limbs were hypotonic, and reflexes were diminished equally with an intact sensory system. Investigations revealed severe hypokalemia with a serum potassium of 1.8 mmol/l. Arterial blood gas showed compensated metabolic acidosis. (pH-7.3, HCO₃-13 mmol/l, PO₂-86 mmHg/PCO₂-21 mmHg, Urine potassium-30 mmol/L) Other metabolic panels and inflammatory markers were unremarkable, including normal calcium and magnesium levels. ECG revealed sinus rhythm with u waves.

Table 1: Laboratory investigation results of the patient

Parameter	Test result	Normal range
WBC	11 000 /mm ³	4000-11 000 /mm ³
Hemoglobin	11.3 g/dl	11-16 g/dl
Platelets	230 000/mm ³	150000-450000/mm ³
AST	32 u/L	5-40 u/L
ALT	28IU/l	7-55IU/l
CRP	5 mg/dl	<10mg/dl
pH	7.31	7.34-7.44
Bicarbonate	18mmol/l	18mmol/l
Chloride	107mmol/l	22-26 mmol/l
Sodium	137 mmol/l	135-145 mmol/l
TSH	2.2mIU/l	0.4-4 mIU/l
T4	8 micrograms/	5-12 micrograms/l
ESR	12mm/hour	24 mm/hour
Urine pH	7	4.6-8

Urgent treatment with intravenous KCl was started, along with regular monitoring for respiratory rate and single breath count. Even after replacing 120 mmol of KCl, along with magnesium replacement, the weakness of the patient progressed, and she developed type two respiratory failure 6 hours after admission. Intravenous KCl was continued as an infusion, and her weakness started to improve along with serum potassium levels. She was extubated after 48 hours of mechanical ventilation and was having a steady increase in potassium. However, while the patient had hypokalemia with acidosis, the urine pH was >7, which led to a diagnosis of distal RTA (RTAd). The blood picture revealed a large population of oval macrocytosis compatible with southeast Asian ovalocytosis.

With continued potassium replacement, along with supportive care, she was able to walk by the fourth day with complete recovery of limb weakness. She was followed up at the medical clinic without any new complaints. Her family screening revealed two siblings with a similar condition.

Discussion

Hypokalemic periodic paralysis with distal renal tubular acidosis (RTAd) is seen in a cluster of the population in the north-central part of Sri Lanka particularly in association with South East Asian Ovalocytosis (SEAO). We are reporting the first occurrence of severe

respiratory muscle paralysis, which needed intubation in a patient having RTAd with severe hypokalemia with associated SEAO.

Hypokalemic periodic paralysis could be either primary (familial) or secondary. Secondary causes include renal tubular acidosis (RTA), primary hyperaldosteronism, Barter syndrome, Gitelman syndrome, liquorice ingestion, barium ingestion, thyrotoxicosis and GI loss (2). Sudden onset weakness ranging in severity from milder weakness to severe disability has been reported, but the clinical picture will depend on the severity of hypokalemia as well as the underlying cause. Muscle weakness is precipitated by heavy exercise, high carbohydrate meals or fasting (3). A thorough investigation is needed to identify the underlying cause of hypokalemic periodic paralysis. Most of cases, Familial hypokalemic periodic paralysis can occur sporadically either as autosomal dominant inheritance or spontaneously. This form of periodic paralysis is considered to be secondary to disordered potassium regulation (4)

However, our patient had normal anion gap metabolic acidosis with alkaline pH, which was compatible with renal tubular acidosis. Persistent high urine pH above 5.5 was suggestive of distal renal tubular acidosis, which can be primary or secondary to autoimmune conditions such as SLE, rheumatoid arthritis, and Sjogren's syndrome. An extensive evaluation didn't reveal any secondary cause for this RTA. Her blood picture revealed a large red cell population of ovalocytosis with transverse ridges without polychromasia compatible with Southeast Asian ovalocytosis.

Hypokalemic periodic paralysis is treated with potassium supplements. Non-pharmacological interventions such as avoiding high carbohydrate meals and refraining from heavy exertion can prevent further attacks. Hypokalemia with distal renal tubular acidosis is treated with alkali therapy either with sodium bicarbonate or potassium citrate after the initial correction of hypokalemia. However, potassium citrate is better tolerated than sodium bicarbonate as it has fewer gastric side effects. The drug of choice for distal renal tubular acidosis is potassium citrate, which provides potassium and correct acidosis simultaneously (5).

After initial resuscitation, she was started on oral KCl supplements as potassium citrate was not available. On review 2 weeks after discharge, her serum potassium was 4.5 mmol/L, and she was started on potassium citrate.

She remained well at 2 years follow-up. We arranged family screening, and the patient's father was found to have southeast Asian ovalocytosis.

In Sri Lanka, there is a cluster of the population with renal tubular acidosis associated with southeast Asian ovalocytosis especially concentrated in North Central province (6). Exact data is lacking about the incidence and prevalence of southeast Asian ovalocytosis among Sri Lankan population.

While the presence of both RTA and SEA is both prevalent in Anuradhapura, where CKDu is also

prevalent, further research is needed to the establishment of the association of these conditions.

This case reports the occurrence of respiratory muscle paralysis due to hypokalemic periodic paralysis secondary to distal renal tubular acidosis with Southeast Asian ovalocytosis. While further studies are required to evaluate the pathophysiological relationship of the co-occurring RTAd and ovalocytosis, this brings up the importance of anticipating life-threatening respiratory muscle paralysis in managing a patient with a similar condition.

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