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CASE STUDY

PRIMARY HYPOKALEMIC PERIODIC PARALYSIS: A CASE STUDY AND REVIEW OF LITERATURE

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ABSTRACT

Hypokalemic periodic paralysis is a condition in which affected individuals may experience paralytic episodes with concomitant hypokalemia (<2.5 mmol/L), and occasionally may develop late-onset proximal myopathy. The paralytic attacks are characterized by reversible flaccid paralysis usually leading to paraparesis or quadriparesis but typically sparing the respiratory muscles and heart [1]. The major triggering factors are carbohydrate-rich meal, vigorous exercise, viral illness, stress, emotional triggers. The age of onset of the first attack occurs before 16 years of age in 60% [1]; the frequency of attacks is highest between ages 15 and 35 and then decreases with age. We hereby report a case of primary (familial) hypokalemic periodic paralysis precipitated by carbohydrate rich food in an adolescent, successfully managed with potassium supplements.

INTRODUCTION

Periodic hypokalemic periodic paralysis (PHPP) is a channelopathy with episodic, short-lived, and hypo-reflexic skeletal muscle weakness, without sensory deficit and without loss of consciousness. Early in the course of disease, in primary or familial periodic paralysis, muscle strength is normal in-between the attacks. After many years of these attacks, inter-ictal weakness develops and may be progressive. This case study describes clinical features, pathophysiology, differential diagnosis, investigations and treatment of this relatively rare but treatable disorder.

Case Study

A 15 year old male patient presented with sudden onset bilateral lower limb and upper limb weakness. The patient reported to have consumed carbohydrate rich food (sweets) the previous night and woke up unable to move his upper and lower extremities. He had no respiratory or swallowing difficulty was able to move his facial and neck muscles. Patient denied any pain or parasthesia. There was no history suggestive of trauma/fever/diarrhea/convulsions/drug abuse. Patient reported to have similar episodes 6 times in the past (recovered with potassium supplements). There was a history of similar complaints in the elder brother.

On examination, the patient was afebrile, pulse was 84/min, regular; blood pressure 110/70mmHg; Examination of the heart, lungs and abdomen was unremarkable. Neurological examination revealed flaccid paralysis in bilateral upper and lower limbs which involved the proximal and distal muscles. Deep tendon reflexes were absent in both upper and lower limbs with bilateral flexor plantars but sensation was intact. Cranial nerve function was grossly intact. No bladder/bowel involvement.

Laboratory investigations revealed low serum potassium 1.3 mEq/L while other routine chemistry, blood gases, liver function tests, thyroid profile, complete blood picture and arterial blood gases were normal. CPK was 450 IU/L, 24 hour urine potassium was 19 mEq/L/day and CT Brain showed no abnormalities. ECG showed 'U' waves, ST depressions consistent with hypokalemia. Considering the history, clinical pattern and biochemical abnormalities, a diagnosis of primary hypokalemic periodic paralysis was made and patient put on oral potassium supplements. Clinical recovery started from next day onwards gradually with return of normal power by 3rd

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day. Serial investigations revealed rising potassium levels to near normal levels by the end of 3rd day.

DISCUSSION

Hypokalemic paralysis can be primary or secondary. Primary hypokalemic periodic paralysis is an autosomal dominant channelopathy [2], the mutations affecting CACN1S (Calcium channel) and SCN4A (Sodium channel). Secondary hypokalemic paralysis is non-genetic in origin, resulting from causes like thyrotoxicosis, barium poisoning, primary hyperaldosteronism, distal renal tubular acidosis, licorice ingestion, Sjogren's syndrome, Fanconi's syndrome, and gastrointestinal disorders like non-tropical sprue, laxative abuse, severe diarrhea and vomiting [3]. In patients with Primary HPP, there is generally a positive family history and first attack usually occurs before 16 years of age. Patients experiencing first attack in adulthood should be carefully screened for secondary causes. Men tend to be more affected by HPP as there is incomplete penetrance in women [4].

Attacks are triggered by strenuous exercise, high carbohydrate meals, viral illness, or by drugs like insulin, beta-2 agonists, or steroids. HPP attacks can be paralytic or myopathic but paralytic form is more common and weakness may be mild and limited to certain muscle groups, or more severe and affect the arms and legs lasting several hours to days. Between 15-45 years of age paralytic attacks are common and affected individuals usually regain their muscle strength between attacks. As age progresses, there is decrease in frequency of paralytic attacks which may be replaced by abortive attacks. Abortive attacks are of long duration, fluctuating weakness which never progresses to paralysis and 25% of these attacks progress to myopathy or permanent muscle weakness. [5]

During episodes of hypokalemia, muscle cells get hyperpolarized which generates an abnormal gating pore current in patients having missense mutations in the L-type calcium channels or the skeletal sodium channel causing ineffective depolarization and hence ineffectual contraction (paralysis).[6]

Diagnosis of familial HPP rests on history, ruling out other causes of acute generalized weakness, excluding secondary causes of hypokalemia (reduced intake; trans cellular shift due to drugs like insulin, beta-2 agonists; renal losses; GI losses), routine chemistry, thyroid profile, EMG, muscle biopsy and genetic testing. [7]

Treatment includes oral potassium supplements, acetazolamide (which causes mild metabolic acidosis driving potassium out of the cells) [8, 9], intravenous potassium is reserved only for patients experiencing severe weakness and is unable to swallow.

Further attacks may be prevented by avoiding strenuous exercise, high carbohydrate diet and other identified triggers. HPP responds well to treatment, muscle strength is normal between attacks, although repeated attacks may eventually permanent weakness between attacks. Low carbohydrate low sodium diet, spironolactone and diclofenamid [10] have also been tried in the management of HPP. None of these measures prevent progressive myopathic changes.

CONCLUSION

Hypokalemia is an important differential in patients presenting with acute neuromuscular flaccid weakness. A 15 year old patient presented with paraparesis and after careful history taking and excluding secondary causes of hypokelmia a diagnosis of PHPP was made and the patient was treated successfully with oral potassium supplements and counseled on avoiding triggering factors.

References

1. Fenichel GM. Flaccid limb weakness in childhood. In: Clinical Pediatric Neurology, 2nd edn. Philadelphia, WB Saunders Co, 1993, pp 192-193.
2. Jurkat-Rott K, Lerche H, Lehmann-Horn F: Skeletal muscle channelopathies. J Neurol 2002; 249(11):1493-1502
3. Soule BR, Simone LN, "Hypokalemic Periodic Paralysis: a case report and review of the literature. Cases Journal 2008; 1:256
4. 21. Sternberg D, Tabti N, Hainque B, Fontaine B. Hypokalemic periodic paralysis. Gene reviews. http://www.ncbi.nlm.nih.gov/books/ 2006
5. Sansone V, Meola G, Links TP, Panzeri M, Rose MR. Treatment for periodic paralysis. Cochrane Database Syst Rev 2008; 23:CD005045.
6. Harrison's Principles of Internal Medicine. 19th edition, McGraw-Hill, 2015: Chapter 63
7. Lin SH, Lin YF, Chen DT, Chu P, Hsu CW, and Halperin ML: Laboratory tests to determine the cause of hypokalemia and paralysis. Arch Intern Med 2004; 164(14):1561-1566.
8. Griggs RC, Engel WK, Resnick JS. Acetazolamide treatment of hypokalemic period paralysis. Ann Intern Med 1970, 73: 39-40.
9. Vroom FFQ, Jarrell MA, Maren TH. Acetazolamide treatment of hypokalemic period paralysis: Probable mechanism of action. Arch Neurol 1975, 32: 385-386.
10. Sander C. Therapy of paroxysmal hypokalemic paralysis: Experience with diclofenamid. Monatsschr Kinderheilkd 1988, 136: 149-150

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