

A possible association of hypokalaemic periodic paralysis, autoimmune thyroiditis and neuromyotonia

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Introduction

Acute hypokalemic periodic paralysis (HPP), a clinical syndrome characterised by acute systemic weakness and low serum potassium (K^+), is a rare but treatable cause of acute limb weakness. Hypokalemia can be caused by K^+ loss via the kidneys or extra renal routes mainly the gut, or due to transcellular potassium shifts where extracellular K^+ will move into the cell. In the latter situation, although there is hypokalaemia, there is no deficit of K^+ in the body. The main causes for intracellular shift of K^+ are familial hypokalemic periodic paralysis, thyrotoxic periodic paralysis, barium poisoning, insulin excess and alkalosis [1].

Although the association between thyrotoxicosis and HPP is known, HPP with hypothyroidism is extremely rare. We report a case of hypokalemic periodic paralysis associated with hypothyroidism and neuromyotonia.

Case report

A 30-year old male presented with acute onset muscle paralysis after awakening from his usual afternoon nap. He had not undergone strenuous activity or consumed a meal rich in carbohydrate prior to this episode. There had been a similar incident two months back which had resolved spontaneously. There were no features of thyrotoxicosis or hypothyroidism and no family history of familial hypokalemic periodic paralysis. Physical examination of limbs showed weakness of all four limbs,

the proximal muscles were weaker than the distal ones. Reflexes were intact and there was no sensory impairment. He had a positive Chvostek's sign but a negative Trousseau's sign and there was no goiter. The blood pressure was within normal limits.

Investigations showed low serum K^+ level of 2.2mmol/L. Urinary spot sample for K^+ was repeatedly less than 20mmol/L indicating no renal losses of K^+ . Arterial blood gas showed a pH of 7.45, PO_2 of 14kPa, PCO_2 of 4.9 kPa, HCO_3^- of 22.5 mmol/L and BE of 2. Serum ionized calcium 1.3 mmol/L (normal range 1.12 - 1.32 mmol/L) and magnesium 0/94 mmol/l (normal range 0.62-0.95 mmol/l) were within normal range. The ECG showed wide spread ST segment depression. Thyroid function tests showed a TSH of 16.5 mu/l (normal range 0.3 - 4.2) and normal FT4 levels. TPO antibodies were grossly elevated 324 IU/ml (>9 positive) and FNAC of the thyroid gland confirmed the diagnosis of autoimmune thyroiditis. The CPK values were marginally elevated 234 u/l (24 - 195) and the lipid profile showed high total cholesterol and triglyceride values.

Neurophysiological evaluation was done after recovery. Compound muscles action potentials were obtained by electrical stimulation of the nerves. However there was an unexpected finding of spontaneous myokymia with repetitive F waves seen in several nerves (Figure 1). These findings were suggestive of neuro-myotonia (Figure 2). A muscle biopsy performed subsequently was normal.

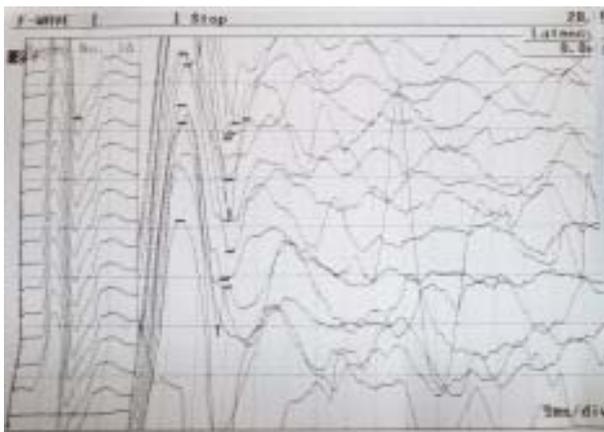


Figure 1. F wave latencies of median nerve.

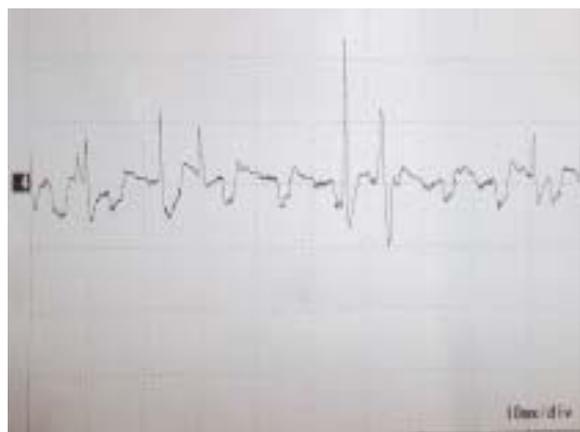


Figure 2. EMG of quadriceps showing myokimic discharges and doublets.

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The response to K⁺ replacement was dramatic and the patient was able to walk the following day. He was sent home on oral thyroxin 100 µg daily. K⁺ supplements were omitted subsequently as the serum K⁺ was normal. Thyroid functions improved and he did not develop any further paralytic attacks.

Discussion

This patient had features suggestive of HPP without evidence of K⁺ loss through the kidneys or the gut. The recurrent episodes of hypokalemia could most likely be due to intracellular shift of K⁺. Most forms of HPP are genetic, with autosomal dominant inheritance. These diseases are channelopathies, (caused by mutations in ion channel genes). The culprit genes encode muscle sodium (SCN4A), calcium (CACNA1S) and potassium (KCNE3) channels [2, 3]. Thyrotoxic hypokalemic periodic paralysis is a well-established phenomenon associated with HPP but our patient had subclinical hypothyroidism with autoimmune thyroiditis. A similar case was reported in Japan 1990 of recurrent hypokalemic paralytic attacks associated with hypothyroidism due to autoimmune thyroiditis. The patient had no further attacks following treatment with thyroxine [4].

Our patient had EMG evidence of neuromyotonia, which is a condition characterised by hyperexcitability of peripheral nerves manifesting as continuous muscle fibre activity. Neuromyotonia is well known to be associated with other auto immune diseases. Patients harbour

antibodies directed against voltage-gated K⁺ channels. These antibodies reduce the K⁺ current conducted by these channels and thus lead to a prolongation of the nerve action potential resulting in increased neurotransmitter release, manifesting clinically as muscle hyperexcitability [5].

This patient with subclinical hypothyroidism and autoimmune thyroiditis had HPP with EMG evidence of neuromyotonia which are diseases both related to K⁺ channels. This raises the possibility of a transient autoimmune process causing antibodies against K⁺ channels causing this patients disease manifestations. Detailed genetic and antibody studies for confirmation of channelopathy and neuromyotonia respectively were not performed due to non availability of these investigations in the country.

References

1. Ahlawat SK, Sachdev A. Hypokalaemic paralysis. *Postgraduate Medical Journal* 1999; **75**: 193-7.
2. Fontaine B. Periodic paralysis: new pathophysiological aspects. *Bulletin del' Academie Nationale de Med* 2008; **192**: 1543-8.
3. <http://neuromuscular.wustl.edu/mtime/mepisodic.html>
4. Kinoshita I, Usa T, Satoh A, Tsujihata M. A case of hypothyroidism associated with hypokalaemic periodic paralysis. *Rinsho Shinkeigaku* 1990; **30**: 100-2 (Japanese).
5. M. Benatar. Neurological potassium channelopathies. *Quarterly Journal of Medicine* 2000; **93**: 787-97.

A case of warm autoimmune haemolytic anaemia with intravascular haemolysis: a rare presentation

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Introduction

Autoimmune haemolytic anaemia (AIHA) is a rare (prevalence 0.2-1.0 per 100,000 population) but a potentially fatal condition [1]. Diagnosis of AIHA is based mainly on Direct Agglutination Test (DAT/Coomb's test) [2]. AIHA

is classified into warm (optimal autoantibody reactivity at 37°C) and cold (optimal autoantibody reactivity at 4°C) types. Based on the presence or absence of an underlying cause, it is divided into primary (idiopathic) and secondary (secondary to lympho proliferative disorders, autoimmune disease, drugs and non-haematological malignancies).

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