Hypokalemic Periodic Paralysis: An Unusal Case

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ABSTRACT

Hypokalemic periodic paralysis (HPP) is a rare syndrome characterised by muscular weakness, arrhythmias, respiratory failure and death. The severity of attack varies from mild pain and weakness to total paralysis. The attack spontaneously abates and patient recovers within 36 hours. The Cognitive and sensory function remains intact and deep tendon reflex may be diminished or absent. Here we present an unusual case of young woman who presented to us with acute quadriplegia as a result of hypokalemia.

CASE REPORT

Thirty years old woman came to us in casualty with history of sudden onset weakness of both lower limb which ascended rapidly within 4 hours to involve both upper limbs and respiratory muscles. There were neither sensory symptoms nor any bladder or bowel involvement. She gave no history of fever or URTI preceding the illness. No history of diarrhoea, vomiting, vigorous exercise or excessive carbohydrate intake prior to onset of weakness. There was past history of similar episode one year back .

On examination, pulse was 100/min, BP was 130/70mm of Hg, RR of 22/min, and single breath count of 7. Neurologically patient was conscious and well oriented, no cranial nerves involvement, power grade 0 and hypotonia in all four limbs, deep tendon reflexes were absent, plantars were flexor bilaterally and sensory examination was normal. Routine haemogram, urine examination, renal function and liver function were normal. Random blood sugar was 110mg%. ECG and serum T3,T4 and TSH were within normal limit and serum potassium was 2.0 mEq/l.

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Muscle biopsy was done and showed no obvious pathology (normal morphology of skeletal muscles). In Nerve conduction velocity studies, right median and right ulnar nerve showed slightly decreased Continuous Muscle Action Potential amplitude (CMAP) as compared to left median and ulnar nerve.

She was treated with 90 mEq of potassium over 24 hrs after which she had complete neurological recovery and repeat serum potassium came out to be 5.0 mEq/l.

The interesting and unusual feature of this case were

1. Female patient
2. Age of the patient was 30 years
3. There was no precipitating cause.

DISCUSSION:

Acute systemic weakness is a common complain in the emergency department and has a wide differential diagnosis that includes neurological, metabolic, infectious and channelopathies causing HPP.

Channelopathies are due to disturbed function of ion channel subunits which regulate them.[1] These diseases may be either congenital (often resulting from a mutation or mutations in the encoding genes) or acquired (after resulting from an autoimmune attack

on ion channels).[2] Around 24 types of different Channelopathies of human skeletal muscles include hyper, hypo and normokalemic (high, low and normal potassium blood concentrations) periodic paralysis, myotonia congenita and paramyotonia congenita.[3]

All primary periodic paralysis have some features in common, they are all treatable and muscular weakness is reversible. They are characterised by episodes of flaccid muscle weakness occurring at irregular intervals.[4]

The tendency of hypokalemia can be due to number of causes. The commonest cause described in the Caucasian population is the familial Hypokalemic periodic paralysis.[5] This entity is due to a defect in the calcium channels of the muscles due to defect in Hypokalemic periodic paralysis type 1 gene. It occurs as an autosomal dominant condition in two third cases and is sporadic in remaining one third.

The exact incidence of HPP is not known, however Ariza-androea CR et al studied 18 cases of HPP in Spain, 16 males and 2 females out of which 5 had familial HPP, 4 had thyrotoxic and 9 were sporadic cases.[6] They concluded that age of onset in familial HPP was earlier and more common in males[6].Similarly A.N.Joshi et al studied 25 cases of HPP in India and showed more incidence of HPP in males as compared to females.[7]

However, our patient was female , with late age of onset and no precipitating factors[6, 7] being an unusual presentation.

The treatment of familial Hypokalemic periodic palsy is by acetazolamide and avoidance of high carbohydrate intake.

CONCLUSION

Hypokalemic periodic palsy is a rare but potentially treatable condition hence require high index of suspicion. Treatment with acetazolamide effectively stops further attacks of paralysis.

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