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General Medicine

CASE SERIES OF HYPOKALEMIC PERIODIC PARALYSIS

Dr. K. Padmanabhan MBBS, MD, PGCDM, HOD-Department Of General Medicine, Public Health Centre, West Mambalam, Chennai.

Dr. V. Deepika MBBS, DMO- Department Of General Medicine, Public Health Centre, West Mambalam, Chennai

ABSTRACT The Periodic paralysis syndrome are described as channelopathies that manifest as abnormal, often potassium-sensitive,muscle-membrane excitability and lead clinically to episodes of flaccid weakness or paralysis, sometimes in association with abnormalities of the plasma potassium level. Strength is initially normal between attacks, but progressive myopathic weakness may develop in up to one-third of patients. Hypokalemic periodic paralysis characterized by attacks that tend to occur on awakening,after exercise, or after a heavy meal and may last for several days. Patients should avoid excessive exertion. A low-carbohydrate and low-salt diet may help prevent attacks.An ongoing attack may be aborted by potassium chloride given orally or by intravenous drip, provided the ECG can be monitored and kidney function is satisfactory. We report a case series of 3 gentlemen with sudden onset paralysis of extremities and laboratory evaluation revealed a low potassium level. The patient paralysis resolved upon repletion of his low potassium and was discharged with no neurologic deficits.

KEYWORDS :

Case Report 1:

A 21 year old gentleman with no comorbidities presented to emergency room with weakness involving both proximal and distal muscles of all four limbs along with truncal weakness since last evening and progressive in nature. He had no respiratory symptoms or difficulty in swallowing and no history of trauma. Prior to this episode,patient had been healthy and denied any recent diarrhea,chest pain,shortness of breath.On examination, he was found to be comfortable and not in acute respiratory distress.His vital signs were as follows : blood pressure 130/80,pulse rate 90/min, Respiratory rate 21 breaths/ min Abdomino-thoracic breathing,sp02 99% in RA, temperature-98.8 F . His neurological examination showed weakness of both upper and lower limbs, equally low on both sides to power 2/5 with truncal weakness, flaccid tone of all 4 limbs and deep tendon showing hyporeflexia in all 4 limbs. Babinski showed bilateral plantar response. Higher motor functions were normal and Cranial nerve functions were intact. No apparent cerebellar signs and no signs of nystagmus.Other systemic examination happened to be normal.

On day of admission, laboratory investigations showed Hb 14.1,PCV 46.2,increased white blood cell count 18560 with neutrophils (88.7%),Lymphocytes(8.7%) .ESR and platelets were within normal limits.RFT was in normal limit. LFT was normal with mildy elevated Liver Enzymes,SGOT 57.0, SGPT 39.0 ; CRP 2.0 and CBG 108 mg/dl. Serum potassium was 2.5 mEq/l and chloride was 110 and Serum Sodium 144. Urine routine revealed 3 to 5 pus cells, 2 to 4 epithelial cells. 24 hour urine protein 91mg/dl.MRI brain showed Posterior annular bulge in C5-C6,L5-S1 levels indenting the anterior thecal sac , No significant neural foramen narrowing / nerve root compression ,Lumbarisation of S1 vertebra and Schmoral's node in D12 vertebra.Electrocardiogram showed prominent U waves. The patient was started on day 1 with intravenous KCL 40mEq over 4 hours,oral KCL TDS.On day 2,serum potassium 2.2 after correction, and treatment continued with Intravenous KCL 40mEq over 4 hours, oral KCL TDS. On day 3, serum potassium 3.9,Serum chloride 104,Serum sodium 143 and patient showed clinical improvement moving all 4 limbs, gained ability to walk with no proximal or distal muscle weakness.Same treatment was continued.On day 4, serum potassium was 4.2 mEq/l, patient improved and discharged with Syrup K MAC 5ml BD and advised to avoid carbohydrate loaded meal and with a follow up appointment with Serum electrolytes.

Case Report 2:

42 year old gentleman presented with complaints of fever for 3 days with weakness of all four limbs since 1 day.there was no history of seizure/bladder or bowel incontinence/respiratory difficulty/ trauma/ any medicine intake.he is a non smoker & non alcoholic. Patient was conscious,oriented, febrile (102.2 F), PR-58/min, BP-150/90mmhg,SpO2-97%@RA,CBG-243mg/dl.Neurological examination showed intact cranial nerves,normal higher motor function & normal sensory system,hypotonia present, reduced deep tendon reflexes seen, power-3/5 in all 4 limbs.blood investigations revealed hb-14.6mg/dl,TC-4,090, platelet-65,000,dengueNS1Ag-positive,urea-33,creatinine-1.2,serum potassium-2.5,normal liver function tests except for SGOT-108U/L INR-1.3.nerve conduction study-suggestive of consistent severe sensory motor axonal and demyelinating polyneuropathy (axonal loss) involving both lower limbs more than both upper limbs.urine potassium-22.3. ECG-sinus bradycardia.Echo- concentric LVH WITH NORMAL EF(65%). patient was treated with injection KCL infusion 40meq over four hours followed by oral KCL syrup.on day 2 of admission,weakness of limbs reverted and repeat potassium value was 3.8meq/l.oral KCL syrup was continued.on day 3 ,serum potassium was 4.1, patient had complete recovery of weakness ,able to walk by self and discharged.

Case Report 3:

A 24 year old male was admitted with a chief complaint of progressively increasing weakness of all 4 limbs for 2 days. He had history of high grade fever for 3 days. The patient stated that his weakness started first from lower limbs and within few hours it progressed to involve the upper limbs. Patient was not able to get up from the bed or not able to walk with or without support. There was no history of sensory disturbances, neck pain, respiratory or diarrheal illness. There was no past or family history of similar weakness.On general examination he was afebrile and his vitals were normal. On neurological examination, power was 3/5 in both upper and lower limbs with diminished deep tendon reflexes. There was no cranial nerve involvement, sensory deficit or any involvement of bladder or bowel. Blood investigations on admission were as follows Hb-13.3g/dl, Total leukocyte count-3100 cubic mm with polymorphs-62.4%, lymphocytes- 25.9% and monocytes- 10.8%. His platelet count was 28,000/cubic mm. His serum sodium was 147mEq/l and potassium 2.8mEq/l and his creatinine kinase was 2162 U/L. Arterial blood gas analysis showed a pH-7.44 mmHg, 20.8 mmol/l of Bicarbonate and an Anion gap 18.1 mmol/l. His renal functions were normal and liver function tests showed a normal serum bilirubin with SGOT and SGPT of 240 and 67 IU/L respectively. Urine analysis showed pus cells of around 8-12/hpf. ECG showed sinus bradycardia with LVH. His dengue NS1 antigen ELISA was positive while IgM and IgG was negative. A diagnosis of dengue fever with thrombocytopenia with hypokalemic paralysis was made and the patient was treated with intravenous potassium chloride infusion. His motor power improved rapidly and at 12 hours after starting treatment his power was completely normal. On the second day of treatment his repeat serum potassium was 3.6meq/l. On the 3rd day the patient's platelet count dropped to 0.05 Lac/cmm after which he was transfused with 4 units of platelet concentrate. On the 5th day of admission his platelet count was 0.43Lac/cmm. On the 6th day Nerve conduction study was done which was found to be a normal study. The platelet count progressively improved to 1.26Lac/cmm and he was discharged in a stable condition walking independently on the 8th day of admission.

Case Series Discussion:

We have described 3 cases studies. The mean age group was early 20's(2 cases, one was 42).all three were male. Presentation was acute episode of ascending type of quadriparesis, involving lower limbs more than upper limbs. 2 cases also had acute febrile illness (Dengue NS1Ag positive), whereas one had reduced urine output. No respiratory symptoms/dysphasia/ GTCS were noted. Neurological examination revealed flaccid paralysis, power 3/5 & 2/5 in 2 and 1 case respectively, higher motor functions were normal,cranial nerves were intact, normal sensory system, flexor in plantar reflex and hyporeflexia noted on deep tendon reflexes in all 3 cases. Mean potassium level on presentation was 2.5. Blood parameters showed normal hemoglobin, thrombocytopenia with leukopenia in 2 cases (dengue NS1Ag positive) and normal in 1 case. The renal functions were normal, liver function test were within limit except elevated SGOT levels in all 3 cases. Mean duration of recovery of weakness was within 12 hours. ECG showed sinus bradycardia in 2 cases and shallow T with U waves in 1 case. All 3 cases showed a complete recovery with iv potassium chloride followed by oral KCL.

DISCUSSION:

There are largely two genes that are responsible for Hypokalemic Periodic Paralysis(HypoKPP).

HypoKPP type 1 is up to 70% of cases. The mutation localizes to a region containing the gene that encodes the alpha subunit of dihydropyridine sensitive L-type voltage gated calcium channel of skeletal muscle and missense mutations in the responsible gene (CACNA1S) on chromosome 1q32.1.

HypoKPP type2 is caused by a mutation in SCN4A gene on chromosome 17q23.3 encoding the pore-forming alpha subunit of the skeletal muscle voltage-gated sodium channel. Approximately 10%-20% of families with HypoKPP have this mutation. The usual pattern of inheritance is autosomal dominant with reduced penetrance in women (M:F ratio 3 or 4:1). Disease has become clinically apparent after adolescence.

The typical attack comes on during the second half of the night or the early morning hours, after a day of unusually strenuous exercise; a meal rich in carbohydrates favors its development. Excessive hunger or thirst, dry mouth, palpitation, sweating, diarrhea, nervousness, and a sense of weariness or fatigue are mentioned as prodromal symptoms but do not necessarily precede an attack. The attack may evolve over minutes to several hours. As in hyperkalemic paralysis, the muscular weakness in this disease is associated with a decrease in the amplitude, and eventual loss, of muscle action potentials and there is failure of excitation by supramaximal stimulation of peripheral nerve or by strong voluntary effort. A decline in strength precedes the loss of motor unit potentials and the failure of propagation of action potentials. Limbs are affected earlier and often more severely than trunk muscles, and proximal muscles are possibly more susceptible than distal ones. The legs are often weakened before the arms, but exceptionally the order is reversed. The muscles most likely to escape are those of the eyes, face, tongue, pharynx, larynx, diaphragm, and sphincters, but on occasion even these may be involved. When the attack is at its peak, tendon reflexes are reduced or abolished and cutaneous reflexes may also disappear. As the attack subsides, strength generally returns first to the muscles that were last to be affected.Myotonia is not seen; indeed, clinical or EMG evidence of myotonia essentially excludes the diagnosis of hypokalemic periodic paralysis. Electrocardiogram (ECG) changes such as increased PR and QT intervals, T-wave flattening, and prominent U waves also suggest an underlying hypokalemia. An accurate medical history is essential for the diagnosis because observation of attacks is unusual, and patients are often normal between attacks. Potassium concentrations are usually low during an attack, less than 3.0 mM, although concentrations less than 2 mM should suggest a secondary form of periodic paralysis.

Diagnosis at a time when the patient is normal may be facilitated by provocative tests. With the patient carefully monitored, including the use of ECG, the oral administration of 50 to 100 g of glucose or loading with 2 g of NaCl every hour for 7 doses, followed by vigorous exercise, brings on an attack, which then can be terminated by 2 to 4 g of oral KCl (the opposite of what pertains in hyperkalemic periodic paralysis). During an acute attack, the preferred method of treatment is oral potassium chloride given at 0.5-1.0mEq/kg, not exceeding 200mEq in a 24-hour period. If a patient is unable to take oral potassium (e.g. arrhythmia due to hypokalemia or airway compromise due to altered mental status), then intravenous potassium (KCl bolus 0.05-0.1 mEq/kg or 20-40 mEq/L of KCl) is indicated. Cardiac monitoring is important during the administration of potassium. Prophylactic administration of acetazolamide or dichlorphenamide can reduce attacks of periodic weakness.However in patients with HypoKPP type 2, attacks of weakness can be exacerbated with these medications. Regular exercise (not too strenuous) to keep the patient fit is desirable.

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