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A STUDY OF 89 CASES OF HYPOKALEMIC PERIODIC PARALYSIS IN EASTERN INDIA

General Medicine

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ABSTRACT

Hypokalemic periodic paralysis is a channelopathy involving calcium channel which is transmitted in an autosomal dominant pattern. A typical episode of hypokalemic periodic palsy is characterized by acute motor weakness during sleep or immediately after arousal after an unusually strenuous exercise or a meal rich in carbohydrate. It usually involves proximal limb muscles, predominantly legs and may be associated with myalgias. The patient usually recovers with no residual weakness after treatment with potassium supplementation.

KEYWORDS

INTRODUCTION:

Periodic paralyses (PP) is a heterogeneous group of muscle diseases characterized by episodes of flaccid muscle weakness occurring at irregular intervals. General characteristics of PP includes-[1]

a) They are hereditary
b) Most are associated with alteration in serum potassium levels
c) Myotonia sometimes may co-exist
d) Usually results from defective ion channels

Causes of periodic paralyses may be enumerated as-[2]

1. Hypokalemic periodic paralysis
2. Normokalemic periodic paralysis
3. Hyperkalemic periodic paralysis
4. Paramyotonia congenita
5. Andersen-Tawil Syndrome
6. Secondary kalemic periodic paralyses like- thyrotoxicosis, aldosteronism, 17 α-hydroxylase deficiency, barium poisoning, glycyrrhizic acid ingestion,abuse of thyroid hormone and renal tubular acidosis

Among the spectrum of diseases causing acute systemic weakness, hypokalemic periodic paralysis is very common. Inherited in an autosomal dominant pattern, it has a reduced penetrance, especially in women. The disease is characterized by acute episodes of weakness involving proximal muscles of limbs and trunk associated with low levels of serum potassium. A typical attack usually comes after an unusually strenuous exercise or a meal rich in carbohydrate. The attack may be preceded by excessive hunger, thirst, dry mouth, palpitation, sweating or diarrhea. After the attack abates, patient may sometimes complain of headache, exhaustion or diuresis. Starting in adolescence, the attacks often recur. The frequency of episodes decrease as the age advances. The defect in hypokalemic periodic paralysis may be within a dihydropteridine-binding, voltage sensitive, skeletal muscle calcium channel.[3] In Eastern India, periodic palsy is more common during summer season without a strong family history of any such episode.[4-5] Hyperkalemic periodic paralysis is an autosomal dominant chanellopathy characterized by episodic generalized weakness of rapid onset. It is invariably associated with a rise in serum potassium. Recent electrophysiologic studies have suggested that the fundamental defect may involve an increase in the muscle membrane sodium permeability.[6-7]

In paramyotonia congenita (Eulburg Disease), myotonia associated with weakness is seen. The episode is usually precipitated by cold exposure. It is also an autosomal dominant disorder.

Normokalemic periodic paralysis has all the features similar to hyperkalemic periodic palsy, except normal to sub-normal serum potassium.

Anderson-Tawil Syndrome is characterized by episodic weakness, cardiac arrhythmias and dysmorphic features like short stature, scoliosis, clinodactyly, hypertelorism etc.[8]It is due to KCNJ2 gene defect which subsequently affects the normal functioning of potassium channels.

AIMS AND OBJECTIVES:

To study the pattern of inheritance and clinical features of patients suffering from an acute attack of hypokalemic periodic paralysis in Eastern India.

MATERIALS AND METHODS:

A cross-sectional study of patients suffering from an acute attack of hypokalemic periodic paralysis in Eastern India. Serum potassium estimation, thyroid profile, ECG and EMG was done in all the cases under study. This study was done in the Department of General Medicine, Rajendra Institute of Medical Sciences, Ranchi in collaboration with Advanced Diagnostic Centre, Ranchi

INCLUSION CRITERIA:

Patients suffering from an acute attack of motor weakness with

1. low serum potassium
2. sub-normal serum potassium with early ECG changes suggestive of hypokalemia

EXCLUSION CRITERIA:

Patients with-

1. acute systemic weakness due to GBS, myasthenia gravis, porphyria etc
2. acute motor weakness with normal serum potassium and normal ECG (due to secondary causes like thyrotoxicosis etc)
3. weakness persisting for more than two weeks despite proper treatment
4. no clinical improvement despite appropriate administration of potassium

RESULTS:

Eighty nine patients of acute motor weakness with low to sub-normal serum potassium and who responded clinically to potassium supplementation were evaluated in this study. All but four patients were male (95.5%). Age of patients varied between 16 to 57 years with a mean of 32.75 years. On proper history taking, sixty five patients (73%) had one to five episodes, sixteen patients (18%) had more than ten episodes while three patients (3.4%) reported as high as more than fifty episodes of acute motor weakness in the past. The duration of each attack varied from anywhere between six hours to eight days (mean duration for which the weakness lasted was 28.4 hours). Time interval between two consecutive attacks varied from two days to thirty years.

Seventy six patients (85.4%) had attack during sleep or just after arousal. Forty one patients (46%) complained of myalgias, especially of lower limbs during the attack. Attacks could be related to heavy exercise in five patients (5.6%) and to fever, excessive sweating and to alcohol intake in three patients (3.4%) each. Similar episodes in the family could be established in four patients (4.5%) only.

On biochemical analysis, definite hypokalemia was present in seventy six patients (85.4%). Of the remaining thirteen patients with sub-normal serum potassium, ECG showed flattening/inversion of T-waves.

As per the history given by patients under study, attacks were more common during summer season. 55% of the attacks occurred between April and June. June to September accounted for about 25% cases, while only 20% of all cases of acute attack of hypokalemic periodic palsy were observed between October and March.

DISCUSSION:

Hypokalemic periodic palsy is a disease which usually affects young males. Most of the patients complained of no more than five episodes in their life. Though, a small number of patients reported more than fifty recurrent episodes. Most of the attacks subsided within one to two days with no or minimal residual weakness. A large number of patients typically developed the attack during sleep or immediately after waking up. Almost half of the patients complained of associated myalgias, especially of lower limbs, during the attack. Few patients could relate the attack to heavy exercise, alcohol consumption and fever.

Only four patients could give a positive family history of similar attacks in their family members.

The attacks were usually clustered during the period of April-June.

RESULTS:

Hypokalemic periodic paralysis is an autosomal dominant disorder. In our study, there is lack of positive family history in a majority of the patients. Also, most of the patients involved in this study were males. Hence, it would not be wrong to say that hypokalemic periodic palsy encountered in Eastern India behaves as a sporadic disease. This may be due to low penetrance of the disease, especially among females.

Most of the attacks are precipitated in summer season during sleep or immediately after arousal. Many patients of hypokalemic periodic palsy in Eastern India complain of associated myalgias during the attack. The attack subsides within one to two days with proper potassium supplementation.

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