

Primary Hypokalemic Periodic Paralysis

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Primary hypokalemic periodic paralysis (PHPP) is a rare entity first described by Shakanowitch in 1882. Only a few cases of PHPP have been reported in Indian literature in adults(1). In children hypokalemic paralysis secondary to gastroenteritis and chronic renal disease is much more common than primary disease(2). We hereby report a case of PHPP in a child, successfully managed with acetazolamide and oral potassium supplementation.

Case Report

An 11-year-old boy weighing 15 kg presented with complaints of recurrent attacks of quadriparesis since 3 years of age. Each episode used to start with symmetrical lower limb weakness progressing to the upper limbs over a period of 3-4 hours. Spontaneous recovery occurred over

3-4 days every time. Most of the episodes started in the early morning hours without any particular precipitating factor. The patient used to have 3-4 such episodes every year. One of the attacks also involved respiratory muscles. The general examination was normal except for the presence of bilateral phlyctenular conjunctivitis. There was hypotonia and proximal muscle weakness in the limbs except right upper limb which was normal. Deep tendon jerks were depressed with bilateral flexor planters. No other neurological signs were present.

On investigations, the tuberculin test showed an induration of 20 mm. As shown in *Table I*, the patient had a consistent hypokalemia in the first week of hospitalization along with ECG showing prominent 'U' waves till the fifth day. Initial arterial blood pH was normal. Other related investigations were normal.

Considering the clinical pattern and biochemical abnormalities, a diagnosis of primary hypokalemic periodic paralysis was made and the patient was put on oral potassium supplements and acetazolamide (30 mg/kg/day) from the second day of hospitalization. Clinical recovery started from fourth day onwards gradually with return of normal power in all the limbs by the 7th day. Alternate day investigations till recovery revealed corresponding rise in serum potassium values to near normal levels by the end of the first week (*Table I*). On therapy the patient also developed metabolic acidosis (*Table I*) which resolved after omitting acetazolamide after two days of recovery. In addition, the patient was given antitubercular treatment for phlyctenular conjunctivitis and a strongly positive Mantoux test.

After recovery, electromyogram and nerve conduction studies were done which

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TABLE I—Correlation of Serum Potassium and Blood pH with Limb Weakness

| Parameter | Day of hospitalization | | | | | |
|--------------------------------|------------------------|------|------|------|------|------|
| | 1 | 3* | 5 | 7 | 11** | 15 |
| Serum K (mEq/L) | 1.7 | 2.2 | 1.8 | 3.1 | 3.8 | 4.5 |
| Blood pH | 7.34 | 7.29 | 7.21 | 7.19 | 7.28 | 7.35 |
| Blood HCO ₃ (mEq/L) | 14.6 | 20.4 | 13.4 | 7.9 | 15.6 | 20.8 |
| Muscle weakness | ++ | + | ± | - | - | - |

* Acetazolamide and K⁺ supplementation started on 2nd day.

** Acetazolamide omitted on 9th day.

were normal as also was the muscle biopsy. Both parents were asymptomatic with normal serum potassium values. During a one year long follow up on oral potassium supplementation, the patient had no relapse, in contrast to a frequency of 3-4 attacks/year before treatment. Periodic serum potassium estimations were also within normal limits.

Discussion

Familial or PHPP is an autosomal dominant disorder with incomplete penetrance in females or rarely sporadic in 20% of the cases. In the present case no family member was affected. The first attack occurs before 16 years of age in 60%(3). Involvement of respiratory muscles is usually uncommon during an attack(3). Our child however, did have a history suggestive of one such self limiting attack in the past. Though attacks can be induced by cold, emotional excitement, rest after exercise and a high carbohydrate meal—our case had no precipitating factor.

Diagnosis of PHPP rests upon excluding secondary causes of hypokalemia, *e.g.*, gastroenteritis, hyperaldosteronism, renal tubular acidosis, *etc.*(3). Intermittent attacks

and normal potassium values in between attacks points to a primary cause in the present case. Though the child also had tuberculosis, the association appears incidental. No case of HPP in association with tuberculosis has been reported in literature.

Though the exact etiology of hypokalemia in PHPP is not known, a possibility of abnormally reduced surface membrane permeability to potassium in muscles has been proposed(4). Thus, low serum potassium coupled with high muscle potassium levels produce hyperpolarisation of the muscle membrane, making it inexcitable(4). Accordingly, acetazolamide has been used in the treatment as it causes mild metabolic acidosis driving the potassium out of the cell(5,6). In one case clinical recovery and rise in serum potassium level was associated with progressively developing acidosis after starting acetazolamide therapy. Once the recovery is complete, acetazolamide can be omitted with continuation of potassium supplementation without a risk of recurrence, as the present case has been asymptomatic on follow up with only one potassium supplementation. Returning of blood pH to normal after stopping acetazolamide was not associated with clinical recurrence.

BRIEF REPORTS

Low carbohydrate low sodium diet, spironolactone and diclofenamid have also been tried in the management of HPP(7). None of these measures prevent progressive myopathic changes.

To conclude, in addition to rarity, the present case also emphasizes the usefulness of acetazolamide in the treatment and the case with which recurrent attacks can be prevented by potassium supplementation.

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Hyperimmunoglobulin E Syndrome

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Hyperimmunoglobulin E syndrome (HIE) is a rare immunodeficiency disorder which

presents with repeated pulmonary infections, pyoderma, otitis media and subcutaneous abscesses, in the child who has coarse features, short stature, eczema and ungal candidiasis. Serum IgE levels are usually greatly raised. Although 130 cases have been reported world-over, to the best of our

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