DOI: 10.7860/JCDR/2014/8372.4483

Case Report

Paediatrics Section

A Case of Periodic Hypokalemic Paralysis in a Patient with Celiac Disease

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ABSTRACT

A 4-year-old male child presented with recurrent episodes of diarrhoea for 6-months, each episode associated with weakness of all four limbs and documented hypokalemia who on examination had some pallor, short stature, flaccid quadriparesis with absent DTR. The patient responded clinically and biochemically to potassium supplement. TTG and Intestinal biopsy confirmed celiac disease. Patient was put on gluten free diet and patient is doing well with no recurrence. We present a case of Recurrent hypokalemic paralysis with previously unsuspected celiac disease who was not in celiac crisis.

Keywords: HPP, Celiac disease, Flaccid quadriparesis

CASE REPORT

A 4-year-old male child presented with loose motion for 6 days,(4-5 times/day, semisolid, non foul smelling, no abnormal character of stools) and acute, non- progressive weakness of both upper and lower limb since last four days. Both the upper and lower limb were affected simultaneously, with more involvement of lower limbs and proximal portion. There was no associated difficulty in breathing, difficulty in swallowing, altered sensorium, seizures, visual problem or bulbar weakness.

He had history of similar illness in the past, two episodes of weakness, associated with floppiness of neck for past six-months. Each episode started with early morning weakness without any diurnal variation. It was preceded by loose stools for which he was hospitalised for 2-3 days. During every episode low serum potassium levels were documented, and treated with IV and oral potassium. The child was completely normal in between the episodes. There was no history of polyuria, recurrent UTI or similar complaints in the family. Birth history was normal and milestones were achieved normally. Feeding showed calorie deficit of 250 kcal/day and protein deficit of 5 g/day.

On examination, vitals were normal. Pallor was present and clubbing grade 2 was present. No features of vitamin deficiency were seen. Patient was conscious and oriented. Memory and speech were normal. Motor system showed normal bulk of the muscles, reduced tone in all four limbs, power 3/5 in all limbs, normal superficial reflexes, absent deep tendon reflexes and B/L flexor response in plantar. No involuntary movements and no sensory loss seen. Cerebellar and meningeal signs were absent. Fundus examination was normal. Chest, CVS, Abdomen examination were clinically normal.

Anthropometry showed weight 12 kg, height 96 cm (between 3rd and 50th percentile) and weight for height 11kg(below 3rd percentile).

Laboratory investigations showed hemoglobin 8.5g/dl, with TLC 13800c/mm, ESR 54mm/h, blood urea 12mg/dl, creatinine 0.3mg/dl, low potassium 2.8 meq/l, normal sodium (143 meq/l). ECG was showing hypokalemic changes. Serum CPK level was 55mg/dl, electromyography and nerve conduction studies were normal. Blood pH 7.346 with normal anion gap. Thyroid function test was normal. Normal calcium profile (Ca 9.3 meq/l Po4 5.4 meq/l, alkaline phosphatase 114 IU/L) normal Serum protein 6.9g/dL albumin 2.9g/dl were noted. Peripheral smear was showing microcytic hypochromic anaemia. Stool examination, USG abdomen, Kochs work up, Urine microscopy and C/S were found normal.

Hypokalemia persisted during the attack ranging from 1.6 to 3.2 meq/L. Hypokalemic paralysis was diagnosed based on clinical and biochemical parameters. His 24h urinary potassium excretion and calcium excretion were normal. Urine pH was within normal limit. The celiac serology was done in view of short stature, chronic diarrhea and anaemia. TTG value (50.9 U/ml) was found 5-fold increased.

Diagnosis of recurrent hypokalemic paralysis secondary to GI loss, most probably celiac disease was made. The child was treated with oral potassium and ORS. Serum electrolytes became normal after 24 hour (serum sodium 144 meq/l, serum potassium 4.1 meq/L) and quadriparesis improved after 3 days.

Intestinal Biopsy was done which confirmed the diagnosis. Child was put on gluten free diet, After one year of regular follow up, patient is normal with no recurrence of paralysis and improvement in growth parameter.

DISCUSSION

Periodic paralysis may be primary or secondary type. The paralytic attack can last from an hour to several days and the weakness may be generalized or localized [1]. Disturbances of potassium equilibrium can produce a wide range of disorders including myopathy, marked muscle wasting, diminution of muscle tone, power, and reflexes [2]. The primary hypokalemic periodic paralysis is an autosomal dominant channelopathy and is exacerbated by strenuous exercise, high carbohydrate diet, cold, excitement and specific drugs, e.g., beta agonists, corticosteroids and insulin [1]. In the primary type, episodes of weakness recur frequently.

Hypokalemic periodic paralysis though common among Indian population varies greatly in disease spectrum and magnitude in our country due to the heterogeneous pattern of etiology behind it. In a study of 31 patients, there were 13 patients (42%) with renal tubular acidosis, 13 with primary hyperaldosteronism (42%), 2 each with thyrotoxic periodic paralysis and sporadic periodic paralysis, and I with Gitelman syndrome. Of the 13 patients with renal tubular acidosis, 10 had proximal and 3 distal renal tubular acidosis [3]. Secondary periodic hypokalemic paralysis has also been reported in association with gastroenteritis, diuretic abuse, Bartter syndrome, villous adenoma of colon, and hyperthyroidism.

Any disease entity that results in renal or GI potassium loss is capable of producing hypokalaemic paralysis. Clinicians should look for a secondary cause of hypokalaemia in patient who presents with paralysis, particularly when there are atypical metabolic features [3].

In this patient, findings such as normal urinary pH and lack of hyperchloremia during episode of paralysis also excluded the possibilities of renal tubular acidosis. The absence of hypochloremia, and hyponatremia ruled out Bartter syndrome. Normal serum magnesium and urinary calcium excretion ruled out the possibility of Gitelman's syndrome. Characteristic features of hyperaldosteronism like hypertension and polyuria were absent with normal adrenal gland in the USG of the abdomen. Normal thyroid function test ruled out thyrotoxicosis.

Malabsorption, weight loss and vitamin/mineral-deficiencies characterize classical celiac disease. Vitamin/mineral deficiencies are still common in newly "early diagnosed" celiac disease patients [4].

Presentation of celiac disease as autism [5], cerebellar ataxia in crisis [6], peripheral neuropathy [7], recurrent Guillain-Barré syndrome [8] and encephalopathy and seizure in celiac crisis [9], have been reported but our patient was not in state of celiac crisis. Immune-driven mechanism by an autoantibody, direct neurotoxic effect of gluten or dietary deficiencies were suggested as the pathogenic pathways of the neurologic injury [10].

No similar cases of recurrent hypokalemic paralysis in the presence of celiac disease without crisis have so far been reported. Few reported cases of HPP were in celiac crisis presenting as severe acute diarrhea with life-threatening metabolic derangement leading to paralysis, but our patient was not in celiac crisis. As this patient initially responded to potassium supplement with normalization of serum potassium level, we believe that HPP was due to potassium depletion and patient had secondary cause of periodic paralysis i.e. celiac disease.

CONCLUSION

We report a rare case of celiac disease presenting as hypokalemic periodic paralysis who was not in celiac crisis. Neurological manifestations, are increasingly known to be the presenting feature of celiac disease. Thus screening for celiac disease should be recommended in many neurological disorders especially when the aetiology is not obvious. Our case suggests that celiac disease should be considered in the differential of hypokalemic periodic paralysis in children.

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FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Jan 04, 2014**
Date of Peer Review: **Feb 19, 2014**
Date of Acceptance: **Apr 18, 2014**
Date of Publishing: **Jun 20, 2014**