Acute Flaccid Quadruparesis In A Young Male: A Case Report
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Abstract
A 28 year old normotensive euthyroid male presented with recurrent lower motor neuron type of weakness without any sensory or autonomic involvement, with preserved reflexes. Systemic examination was significant for a mild hepatosplenomegaly. Investigations revealed persistent hypokalemic, hyperchloremic, normal anion gap metabolic acidosis with deranged liver functions. Urine pH was 6.0 even after an oral ammonium-chloride loading test. A case of Type I Renal Tubular Acidosis (RTA) was diagnosed. A search for the etiology revealed bilateral Kayser-Fleischer (KF) ring, with low serum ceruloplasmin levels and high urinary copper, confirming it to be Wilson's Disease (WD).

A 28 year-old male farmer was admitted with weakness of all the four limbs. To start with, the weakness involved the lower limbs and then progressed rapidly to involve the upper limbs making the patient bed-bound within the next day.

The weakness involved both proximal and distal group of muscles and was associated with significant flaccidity but without fasciculations, sensory abnormalities or diurnal variation. There was no history suggestive of alteration of higher functions, cranial nerve or sphincter involvement and of radicular pain or girdle sensation. Weakness was not preceded by vaccination, respiratory tract infection or diarrhoea and was not associated with food intake, exercise or change of temperature. He denied unexplained sweating, tremors, heat intolerance, prolonged vomiting, diabetes, hypertension or any drug intake over a prolonged period of time.

The patient had 2 such similar attacks in the past, but those were milder, subsided spontaneously and didn't require hospitalization. His personal and family history was non-contributory.

The patient had a normal higher function, and normal vitals with blood pressure of 110/80mm Hg. The neurological examination revealed marked hypotonia and weakness in all four limbs, the power being 1/5 in the lower and 2/5 in the upper limbs. There was no cranial nerve involvement, sensory impairment and no abnormality in any of the superficial or deep reflexes. Plantar reflexes were bilaterally flexor and coordination could not be tested. Both the lobes of liver were palpable 3c.m below the costal margin with firm consistency. He had a mild splenomegaly without any venous prominence and clinical evidence of free fluid in the abdomen.

The complete blood count, serum urea and creatinine were normal but the fasting plasma glucose was 182mg/dl and the post-challenge plasma glucose was 214mg/dl. The liver function test revealed the following: Bilirubin-0.9mg/dl, AST-168U/L, ALT-58U/L, Albumin-2.2gm/dl and Globulin-4.1gm/dl. The serum sodium, potassium, chloride and bicarbonate were 140mEq/L, 2.8mEq/L,108.1mEq/L and 20.4mEq/L respectively. The arterial blood gas analysis suggested mild metabolic acidosis with normal anion gap.

A provisional diagnosis of hypokalemic paralysis was considered and the electrolyte deficit was corrected with oral and intravenous potassium supplementation. The patient improved completely but a repeat estimation after 4 days showed a persistent hypokalemia (K=2.7meq/L) and metabolic acidosis. Measured calcium (9.1mg/dl) and magnesium (1.9mg/dl) were within normal limits.

He had a normal thyroid profile. Serum HIV serology, Serum electrophoresis, Creatine Kinase, Aldolase, Porphobilinogen, Cerebrospinal fluid study and Electrophysiological studies were all normal.

24 hours urinary potassium excretion was 242.76mEq.
Plasma and urinary osmolality were 296.76mosm/kg and 482.2mosm/kg respectively. Estimated transtubular potassium gradient (TTKG) came out to be 15.25.

To proceed with the diagnosis of a case of hypokalemia, we followed the algorithm given in Figure 1(1). A potassium excretion >15mmol/day and a TTKG of more than 4 narrowed our possibilities further. A diagnosis of renal tubular acidosis (RTA) was made and oral NH₄Cl test (0.1gm/kg) revealed worsening of acidosis(7.377 to 7.289) with a urinary pH of 6.

[Image:2]

Presence of distal RTA with deranged liver function prompted us to re-evaluate the patient. Careful systemic examination revealed a faint corneal ring (which was missed previously) (Figure 2) and slit lamp biomicroscopy confirmed KF ring. Ultrasonogram of abdomen documented coarse hepatic echotexture, mild splenomegaly, and minimal ascites. Liver biopsy revealed early cirrhotic changes. Diagnosis of Wilson's disease (WD) was confirmed with the serum ceruloplasmin level of 71.34 mg/dl and 24 hours urinary copper excretion of 115 microgram.

[Image:2]

**DISCUSSION**

Hypokalemia with paralysis is a potentially reversible medical emergency. It is primarily the result of an enhanced shift of potassium (K+) into cells, decreased intake or excessive loss. The urine K+ excretion and evaluation of blood acid – base status could be helpful in diagnosis and management (2). Episodic weakness with onset after age 25 years is almost never due to periodic paralysis with the exception of thyrotoxic periodic paralysis (3), which was ruled out in this case by a normal thyroid profile. The normal renal response to hypokalaemia is to retain potassium. High urinary potassium in the presence of hypokalaemia suggests the kidneys are the problem. If in doubt as to whether the cause is renal or extrarenal, measurement of the TTKG can help (4). The TTKG is a semi-quantitative index of the activity of potassium secretion from the distal convoluted tubule and the cortical collecting duct which is measured as

\[ \frac{\text{urinary K+}}{\text{plasma K+}} \div \frac{\text{urinary osmolality}}{\text{plasma osmolality}} \].

Hypokalaemia with a TTKG>4 suggests renal potassium loss due to increased distal potassium secretion (5). Our patient had a TTKG of 15.25 establishing a renal loss. Distal renal tubular acidosis results from ineffective addition of hydrogen ions to the lumen of the distal nephron. The syndrome is manifested by hyperchloremic metabolic acidosis often associated with hypokalemia with a normal serum anion gap (6). Absence of bicarbonaturia and worsening of systemic acidosis with urine pH above 5.5 following an oral ammonium chloride loading test confirm type 1 RTA. Renal affection is part of the clinical picture of developed Wilson's disease. The most frequent sign of affection is distal tubular acidosis (DRTA), more frequently in its latent form with a normal systemic pH, but inability to reduce the urinary pH below 5.5 after an acid load. DRTA is a frequent component of the clinical picture, in particular in Wilson's disease when diagnosed late (7,8). Our patient also had near normal blood pH with urine pH more than 5.5. Renal tubular acidosis can give rise to hypokalemic paralysis (9). Recurrent hypokalemic paralysis due to renal tubular acidosis as a rare initial presentation of WD has been reported (10). Interestingly enough, similar case of WD with diabetes mellitus presenting as RTA was reported from India (11). We present this case to highlight such rare neurological presentation of WD involving the peripheral neuromuscular system.

**CONCLUSION**

The idea of presenting this case is to highlight a rare presentation of Wilson’s disease and emphasize the meticulous workup of every case of hypokalemia to search for a cause.

**References**


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