Hypokalemic Periodic Paralysis Case

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Abstract

Objectives: In this case, a five year-old girl without a similar history in the family, having hypokalemic periodic paralysis attacks without an obvious trigger factor is presented.

Case: 5-year-old girl was brought to our clinic with repeated complaints of weakness. She had the history of polymyositis diagnosis and the use of deltacortril and received intensive care treatment in another clinic with a similar complaint about a year ago. There was no significant family medical history. Physical examination was normal. Pathology was not observed in the tests in previous clinics except hypokalemia at 1.6 meq/L (3.5-5 meq/L) level. Cranial and spinal MR in previous clinics was normal. Hypokalemic periodic paralysis (HypoPP) was considered. CACNA15 gene was sent for the diagnosis of the patient. Spironolactone and oral potassium therapy was started. The patient was discharged with recommending proposing outpatient follow-up. At outpatient follow-up, CACNA15 gene was found to be positive. Genetic counseling was offered to the family.

Results: It is important to keep in mind hypopotassemic periodic paralysis disease, although rare, in patients applying with weakness and malaise, and possible fatal cases can be prevented by giving genetic counseling to the family and early diagnosing the undiagnosed patients after the diagnosis is genetically verified. This study has been presented for the consideration of the rare HypoPP in patients presenting with sudden muscle weakness.

Keywords: Hypokalemia; Periodic paralysis; Weakness

Introduction

The primary periodic paralysis is a rare disease showing autosomal dominant inheritance characterized by recurrent muscle strength loss attacks, and that can occur due to mutations in the sodium (Na), potassium (K) or calcium (Ca) channels. It is classified as hypokalemic or hyperkalemic periodic paralysis according to serum potassium levels during attacks. The most common form of periodic paralysis in humans is hypokalemic periodic paralysis and is reported to have the prevalence of 1 in 100,000 births, but can be fatal if not treated [1].

Patients’ waking up in the morning with weakness is typical. Patients had numbness in four-extremities. Respiration and facial muscles are usually not affected, but deaths from respiratory failure have also been reported. Carbohydrate-rich diet, stress and extreme exercises are among the causes that trigger the attacks. Average 2-36 hours of attacks can be shortened with potassium replacement therapy [2-4]. In this case, a five year-old girl without a similar history in the family, having hypokalemic periodic paralysis attacks without an obvious trigger factor is presented.

Case

5-year-old girl was brought to our clinic with repeated complaints of weakness. She had the history of polymyositis diagnosis and the use of deltacortril and received intensive care treatment in another clinic with a similar complaint about a year ago. There was no significant family medical history. Physical examination was normal. Pathology was not observed in the tests in previous clinics except hypokalemia at 1.6 meq/L (3.5-5 meq/L) level. Cranial and spinal MR in previous clinics was normal. Hypokalemic periodic paralysis (HypoPP) was considered. CACNA15 gene was sent for the diagnosis of the patient. Spironolactone and oral potassium therapy was started. At outpatient follow-up, CACNA15 gene was found to be positive. Genetic counseling was offered to the family.

Discussion

Hypopotassemic periodic paralysis is a disease that that can be sporadic or familial and seen more frequent in men. Patients typically wake up in the mornings with weakness. The most severe patients cannot move his four extremities. Respiration and facial muscles are usually not affected, but deaths from respiratory failure have also been reported. There are no emotional symptoms during attacks [1-3]. Our patient also did not have the emotional symptoms. However, there was rarely observed respiratory distress. Our patient had an attack remitted about 16 hours 1 year ago and was treated in the intensive care unit of another center. Completely normal progress of the disease between attacks was current for our patient. The attacks can be triggered after heavy carbohydrate
foods. This is due to the increase of the potassium and glucose cell transition [3,4].

Our patient without the history of trauma, recent infections and diarrhea had normal brain scanning, but significant hypokalemia in routine blood tests. Complaints resolved in a short time with potassium replacement treatment. So, the patient was diagnosed with hypopotassemic periodic paralysis. Due to being able to be associated with thyrotoxicosis, viewed thyroid function tests were normal. In the disease with no relation between potassium level and the severity of muscle weakness, dramatic improvement can be observed as a result of potassium replacement treatment [3,5]. Then oral potassium application can be applied. However, treatment must be careful in terms of hyperpotasemia that can develop secondary. In addition, oral potassium therapy was applied. In parallel to the complete clinical progress of the patient at the first six hours, the patient’s potassium levels were also normalized simultaneously.

As a conclusion, it is important to keep in mind hypopotassemic periodic paralysis disease, although rare, in patients applying with weakness and malaise, and possible fatal cases can be prevented by giving genetic counseling to the family and early diagnosing the undiagnosed patients after the diagnosis is genetically verified. This study has been presented for the consideration of the rare HypoPP in patients presenting with sudden muscle weakness.

References