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Periodic paralysis: what clinician needs to know?

Abstract

Acute flaccid paralysis is a diagnostic challenge in the emergency department. Periodic paralysis syndromes are characterized by recurrent episodes of flaccid hyporeflexic paralysis in association with potassium abnormalities. Periodic paralysis with hypokalaemia may be genetic, secondary to systemic hypokalaemia or associated with thyrotoxicosis. Genetic syndrome result from mutations in sodium (SCN4A) or calcium (CACNA1S) channels, inherited in autosomal dominant pattern. Diagnosis is established by demonstrating recurrent nature, family history and abnormal serum potassium during an episode. Thyrotoxic periodic paralysis is often sporadic but possibly has a genetic predisposition. Presence of thyrotoxicosis and hypokalaemia during an episode confirms the diagnosis. Management of acute episode is by cautiously correction of potassium abnormality. Long term therapy depends on the cause. Pathogenic mechanisms, differential diagnosis and treatment principles are discussed.

Keywords: periodic paralysis, hypokalaemia, hyperkalaemia, channelopathies, acute flaccid paralysis

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Abbreviations: ABG, arterial blood gas analysis; ACTH, adrenocortical stimulating hormone; AST, aspartate transaminases; CACNA1S, calcium voltage-gated channel subunit alpha1 S; CPK, creatine phosphokinase; ECG, electrocardiogram; EMG, electromyogram; hyperKPP, hyperkalaemic periodic paralysis; hypoKPP, hypokalaemic periodic paralysis; IV, intravenous; KCl, potassium chloride; LDH, lactate dehydrogenase; NormoKPP, Normokalaemic periodic paralysis; PP, periodic paralysis; REM, rapid eye movement sleep; RTA, renal tubular acidosis; SCN4A, sodium voltage-gated channel alpha subunit 4; TFT, thyroid function tests; TPP, thyrotoxic periodic paralysis

Introduction

Periodic paralysis (PP) is a rare group of clinical syndromes characterized by episodes of paralysis lasting minutes to days as a result of abnormal ion channel fluxes in skeletal muscles. Mutations of ion channels inherited in autosomal dominant pattern are the aetiology for primary PP syndromes.[1,2] They are associated with changes in serum potassium and manifest in childhood to young adult age. Hypokalaemia of other aetiologies may also lead to limb weakness, and are referred to secondary periodic paralysis. Estimated global prevalence of heritable hypokalaemic and hyperkalaemic are 1:100 000 and 1:200 000 respectively.[3] Men are affected more commonly than women.

Knowledge on genetic basis and therapeutic options of this group of diseases has rapidly expanded in the recent past. This review summarizes current knowledge on periodic paralysis syndromes with particular emphasis on thyrotoxic periodic paralysis.

Clinicopathological syndromes

Skeletal muscle channelopathies account for a spectrum of diseases ranging from myotonic syndromes to PP syndromes while some have overlapping features of both (Figure 1). PP syndromes are associated with mutations in sodium or calcium channels of skeletal muscles.

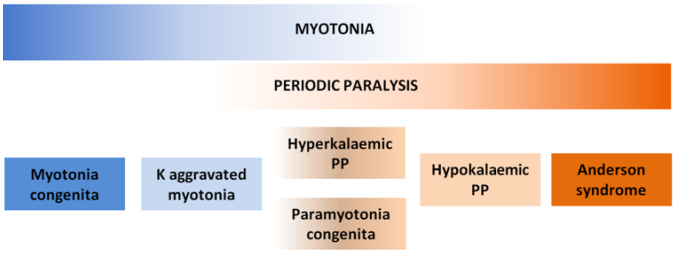

Figure 1 Spectrum of skeletal muscle channelopathies.

Myotonia and periodic paralysis are the cardinal manifestations of skeletal muscle channelopathies. Different mutations cause different syndromes with varying degrees of myotonia and periodic paralysis. PP, periodic paralysis; K, potassium.

Hypokalaemic periodic paralysis (hypokpp)

Periodic paralysis due to hypokalaemia may occur in three settings. 1. Inherited channelopathy causing hypokalaemia and paralysis, 2. thyrotoxicosis precipitating hypokalaemia and paralysis, probably in genetically predisposed, and 3. hypokalaemia by other mechanisms causing paralysis (secondary hypoKPP). Two inherited channelopathies have been identified to cause hypoKPP, sodium channel mutation (*SCN4A*) and calcium channel mutation (*CACNA1S*). Those two in general, have similar pathogenic mechanisms, clinical profiles and treatment options. However subtle differences in the phenotypes of the two types have also been recognized (see *differential diagnosis*). A variety of mutations in *SCN4A* or *CACNA1S* responsible for disease has been identified and are inherited in autosomal dominant pattern.[4]

In normal health, low cytoplasmic causes hyperpolarization of skeletal muscle membrane, but when severe, leads to paradoxical depolarization, which leaves the muscle fibres insensitive to signals from neurons, thereby causing paralysis. In patients with mutated *SCN4A* and *CACNA1S* channels, minor reduction in serum potassium is adequate to produce the paradoxical depolarization.[4] Thus, the hypokalaemic paralysis develops with a minor degree of hypokalaemia

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