A Patient with a Rare Condition Presenting as Ventricular Tachycardia: A Case Report

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ABSTRACT

Introduction: Hypokalemic periodic paralysis (HPP) is a rare neuromuscular disorder associated with episodes of hypokalemia induced muscle weakness and paralysis. There have been some studies that reported presence of cardiac arrhythmias, secondary to hypokalemia.

Case Report: A 49-year-old hypertensive, diabetic lady presented to a specialized cardiac hospital with sudden transient weakness of both lower limbs along with palpitations, for last 3 hours. She also had an episode of transient loss of consciousness for 2-3 minutes, 30 minutes back. On examination, her pulse & blood pressure were non-recordable & patient was drowsy. ECG was recorded & monitor was attached immediately which showed ventricular tachycardia (VT). She was immediately given synchronized DC cardioversion for 2 times & she was reverted to sinus rhythm with restoration of consciousness. Her vitals were stable after cardioversion. Her neurological examination revealed hypotonia of her both lower limbs with diminished jerks and bilateral plantar flexor reflex. Her detailed history & previous records showed similar history of transient lower limb weakness with hospitalizations twice with hypokalemia within last 2 years. All those episodes occurred following heavy meals. After cardioversion, the rhythm was sinus but features were suggestive of presence of hypokalemia including prolonged QT. All baseline investigations were done & she was found to be severely hypokalemic (2.2 mmol/L) along with anemia, hyponatremia & hypomagnesemia. IV potassium & IV magnesium were given along with oral potassium supplementation. All workups were done to find out the cause of her recurrent hypokalemia. Finally diagnosis was made as a case of Hypokalemic Periodic Paralysis with VT.

Conclusion: Although rare but hypokalemic periodic paralysis can present with malignant arrhythmias like VT. So ECG interpretation should be prompt along with prompt management.

Keywords: *Hypokalemic periodic paralysis, Hypokalemia, Ventricular tachycardia.*

INTRODUCTION

Hypokalemic periodic paralysis (HPP) is a rare genetic neuromuscular disorder characterized by episodes of focal or generalized skeletal muscle paralysis, associated with hypokalemia[1].These hypokalemic episodes are most commonly triggered by carbohydrate-rich meals, alcohol, or rest after strenuous exercise[2]. Several studies have reported the presence of cardiac arrhythmias, the majority being secondary to hypokalemia-induced changes. Several studies have described cardiac arrhythmias in context of HPP. Here, we report a case who initially presented with ventricular

tachycardia in the emergency. After proper history, examination & extensive workup, she was diagnosed as case of HPP.

CASE REPORT

A 49-year-old hypertensive, diabetic lady presented to a specialized cardiac hospital with sudden transient weakness of both lower limbs along with palpitations, for last 3 hours. She also had an episode of transient loss of consciousness for 2-3 minutes, 30 minutes back. On examination, her pulse & blood pressure were non-recordable & patient was drowsy. ECG was recorded & monitor was attached immediately which showed ventricular tachycardia (VT) (Figure 1).

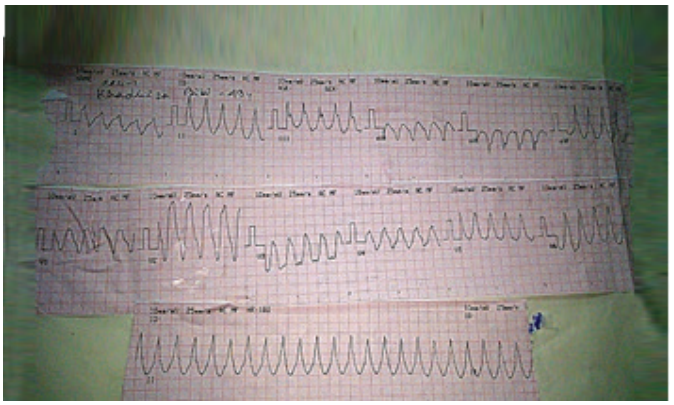

Figure 1: ECG: Ventricular Tachycardia

She was immediately given synchronized DC cardioversion for 2 times & she was reverted to sinus rhythm with restoration of consciousness (Figure 2).

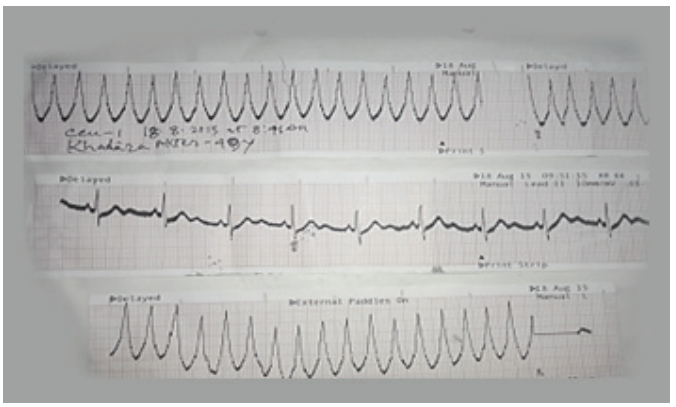

Figure 2: ECG: VT reverted to Sinus Rhythm following DC Cardioversion

Her vitals were stable after cardioversion. Her neurological examination revealed hypotonia of her both lower limbs with diminished jerks and bilateral plantar flexor reflex. Her detailed history & previous records showed similar history of transient lower limb weakness with hospitalizations twice with hypokalemia within last 2 years. Each episode was followed by intake of heavy meals. There was no similar family history. There was no history of stress, strenuous work, heavy exercise or thyroid illness. After cardioversion, the rhythm was sinus, but features were suggestive of presence of hypokalemia including prolonged QT with prominent U waves. T wave flattening was present & best noted in inferior leads (Figure 3). All baseline investigations were done & she was found to be severely hypokalemic (2.2 mmol/L) along with

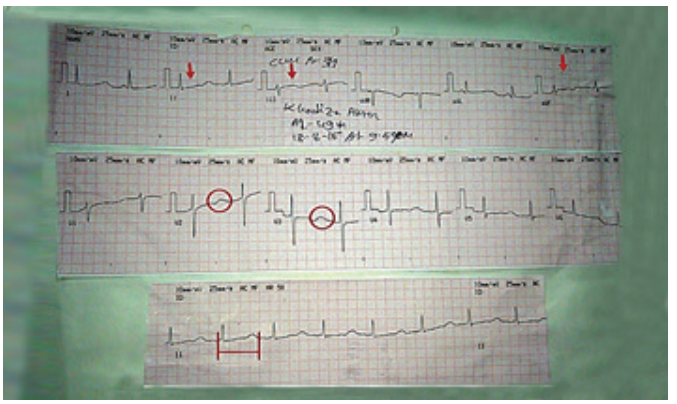

Figure 3: ECG in sinus rhythm showing flattening of T wave, prominent U wave & long QT interval

anemia, hyponatremia & hypomagnesemia (1.4 mg/dl). IV potassium & IV magnesium were given along with oral potassium and spironolactone, for potassium correction. All workups were done to find out the cause of her recurrent weakness & hypokalemia such as thyroid function test, serum calcium, Urinary 24-hour potassium, serum cortisol level, arterial blood gas analysis, chest X-ray & echocardiography. All reports were within the normal limit. Finally, diagnosis was made as a case of Hypokalemic Periodic Paralysis with ventricular tachycardia. Patient recovered well following potassium correction with final potassium 4.5 mmol/L on discharge.

DISCUSSION

HPP occurs in several settings and the diagnosis may require an extensive search for the underlying etiology since the treatment varies according to the cause. HPP may occur sporadically in the form of Familial Hypokalmic Paralysis (FHP), a poorly understood disorder which may occur spontaneously or as the result of autosomal dominant inheritance[3]. This form of Periodic Paralysis is due to the result of disordered cellular potassium regulation as a result of sodium or calcium channel abnormalities[4,5]. HPP is associated with heterozygous pathogenic mutations in the CACNA1S or SCN4A genes[2].

The first attack usually occurs between ages 5 and 35 years, but the frequency of attacks is highest between ages 15 and 35 years and subsequently decreases with age[6]. Our patient presented in her late 40s.

The diagnosis of PP can be confirmed by genetic testing7. In the absence of an identified genetic mutation in approximately 30% of patients, periodic paralysis subtypes can be distinguished on the basis of clinical presentation, serum potassium levels during attacks, and pattern of abnormalities on long exercise testing[7]. Supportive diagnostic criteria for HPP include[8]: two or more attacks of muscle weakness with documented serum K <3.5 mEq/L, three of 6 clinical or laboratory features: onset in the first or second decade, attack duration (muscle weakness involving 1 or more limbs) > 2 hours, positive triggers (high carbohydrate rich meal, rest after exercise, stress), improvement with potassium intake, positive family history or genetically confirmed skeletal calcium or sodium channel mutation, positive McManis long exercise test; exclusion of other causes of hypokalemia (renal, adrenal, thyroid dysfunction; renal tubular acidosis; diuretic and laxative abuse); absence of myotonia (clinically or latent detected by needle EMG), except eye lids. As genetic testing was not available in our center, we followed the supportive diagnostic criteria which were consistent with our patient.

Hypokalemia represents a major arrhythmogenic factor, as it can lead to early and late after depolarizations, reentrant circuits, and altered activation-repolarization coupling[9]. Hypokalemia can be the cause of monomorphic as well as torsades de pointes or polymorphic VT, which are life-threatening situations[10]. Our patient presented with pulseless monophormic VT, which required urgent intervention by DC cardioversion.

The typical electrocardiographic pattern of hypokalemia consists of prominent U-waves >1 mm and U-waves greater than T-waves in the same lead with associated ST-segment depression (sometimes with inversion of T-waves)[11]. This finding is consistent with our patient.

Acute paralytic episodes are treated with potassium replacement and close monitoring of the cardiac rhythm and serum potassium levels. Spironolactone and acetazolamide have been used for prophylaxis with some success although long-term potassium supplementation may be necessary[4] although potassium is usually normal in between episodes. Intravenous potassium and magnesium were given to our patient, following which her hypokalemia was corrected along with resolution of lower limb weakness. Our patient was given oral potassium supplementation along with advice to avoid heavy meal, stress and heavy exercise on discharge.

CONCLUSIONS

We report a rare condition like HPP presenting with VT. Prompt diagnosis of VT & prompt management was essential to save the patient from such life-threatening condition. Finally physicians should be aware of different presentations of HPP.

CONFLICTS OF INTEREST

The authors declare that there are no conflicts of interest.

REFERENCES

1. Statland J.M., Fontaine B., Hanna M.G., et al. Review of the diagnosis and treatment of periodic paralysis. Muscle Nerve. 2018;57:522–530.
2. Fialho D., Griggs R.C., Matthews E. Periodic paralysis. Handb Clin Neurol. 2018;148:505–520.
3. Fontaine B, Vale-Santos J, Jurkat-Rott K, Reboul J, Plassart E, Rime CS, Elbaz A, Heine R, Guimaraes J, Weissenbach J, et al. Mapping of the hypokalaemic periodic paralysis (HypoPP) locus to chromosome 1q31-32 in three European families. Nat Genet. 1994, 6 (3): 267-272. 10.1038/ng0394-267.
4. Jurkat-Rott K, Lerche H, Lehmann-Horn F. Skeletal muscle channelopathies. J Neurol. 2002, 249 (11): 1493-1502. 10.1007/s00415-002-0871-5.
5. Wang W, Jiang L, Ye L, Zhu N, Su T, Guan L, Li X, Ning G. Mutation screening in Chinese hypokalemic periodic paralysis patients. Mol Genet Metab. 2006, 87 (4): 359-363. 10.1016/j.ymgme.2005.10.020.
6. Cannon SC. Channelopathies of skeletal muscle excitability. Compr Physiol 2015;5:761–790
7. Fournier E, Arzel M, Sternberg D, Vicart S, Laforet P, Eymard B, et al. Electromyography guides toward subgroups of mutations in muscle channelopathies. Ann Neurol 2004;56:650–661.
8. Venance SL, Cannon SC, Fialho D, Fontaine B, Hanna MG, Ptacek LJ, et al. The primary periodic paralyses: diagnosis, pathogenesis and treatment. Brain 2006;129:8–17
9. Tse G., Li K.H.C., Cheung C.K.Y., et al. Arrhythmogenic mechanisms in hypokalaemia: insights from pre-clinical models. Front Cardiovasc Med. 2021;8:1–11.
10. Lam L, Nair RJ, Tingle L. Thyrotoxic periodic paralysis Proc (Bayl Univ Med Cent). 2006;19:126–9
11. Helfant RH. Hypokalemia and arrhythmias Am J Med. 1986;80:13–22