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Trifascicular heart block, genetically inherited or hyperkalemia-induced; a rare case report

Abstract

We report a case of trifascicular block with hyperkalemia, due to a familial heart block and/or hyperkalemia. The patient's symptoms and electrocardiogram (ECG) evidence of trifascicular block resolved with correcting serum potassium level. This patient was also diagnosed with a progressive familial heart block (PFHB) type I. This would be the first report of a PFHB type I documented in Iran; this paper highlights an infrequently reported dysrhythmia associated with hyperkalemia which emergency physicians should be familiar with.

Keywords: hyperkalemia, trifascicular block, hereditary bundle branch system defect

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Introduction

Progressive familial heart block (PFHB) type I am an autosomal dominant congenital cardiac conduction disorder which might develop into complete atrioventricular (AV) heart block. In 1986[1] it was clarified the clinical and electrocardiography (ECG) aspect of the condition. Type I PFHB has beendesignated by right bundle branch block (RBBB), prolonged PR interval, left anterior fascicular block (LAFB), and/ or complete AV block with wide QRS complexes.[1,2] PFHB-I has widely been widespread among patients in Africa, and there have been very few reports of this condition among Asians population; it has especially never been introduced among Iranian society. Hyperkalemia is a life-threatening electrolyte imbalance necessitatingimmediate.[3] Serum potassium of above 5.5mmol/l is considered hyperkalemia which is frequently asymptomatic till plasma potassium runs above 6.5-7mmol/L; leading to lethal arrhythmias, therefore it's named as silent killer. Hyperkalemia is linked with vital instabilities in heart conduction system, ranging from shortening of QT interval, to lengthening of PR interval as well as QRS widening. Bundle branch blocks or intraventricular conduction delay and Reversible fascicular blocks as well may be seen.[4] Furthermore; hyperkalemia is recognized to induce potential deadly dysrhythmias comprising ventricular fibrillation (VF), ventricular tachycardia (VT), idio ventricular rhythms, beside a systole.

Case report

A 59year-old Fars male presented to our Emergency Department of Sayyad Shirazi Hospital, Golestan University of Medical Sciences, Northern Iran, with complaints of retrosternal chest pain and diaphoresis. He was a known case of systemic essential hypertension diabetes mellitus. Patient had not been received any treatment while presentation. His blood pressure detected as 163/91mmHg with pulse rate of 42/minute. Atropine was injected then he was immediately transferred to Coronary Care Unit (CCU). With more examination in CCU, his blood pressure was detected as 160/90mmHg, with pulse rate of 73/minute and cardiac auscultation revealed no significant finding; all other examinations were unremarkable. Patient's first electrocardiogram showed heart rate of 78/min, PR interval of 253milli-sec, and QRS duration of 144milli-sec with mean QRS axis which was 264°. ECG showed as trifascicular block i.e. first degree atrioventricular (AV) block, right bundle branch block (RBBB), left anterior fascicular block (LAFB). His random blood sugar was 351mg/dl. He had experienced two episodes of Stokes Adams attack following which isoprenaline drip was initiated. Management of acute coronary syndrome begined for him as well. ABG reported metabolic acidosis with compensatory respiratory alkalosis and hyperkalemia (pH 7.35, pCO2 19.2 mmHg, bicarbonate 10.7mmol/L, S. Na+ 132mmol/L and S. K+ 7.3mmol/L). Laboratory results showed leukocyte count of 15300/mm3, hemoglobin of 6.1g/dl, with hematocrit of 21.4%. His serum creatinine was 4mg/ml with blood urea of 95.8mg/dl. Anti-hyperkalemic measures (including calcium gluconate, kayexalate, and insulin and albuterol nebulization) were immediately launched and our patient underwent urgent hemodialysis; packed cell was transfused carefully. On the second day, hyperkalemia and acidosis were improved (S. K+ 4.1mmol/L, pH: 7.52, pCO2: 37.4mm of Hg, HCO_3: 28.8mmol/L). His ECG on the second day indicated the heart rate of 85/min, PR interval of 136 millisec, and QRS duration of 103 millisec with mean QRS axis -52.5°. His ECG (Figure 1A) was diagnosed as LAFB; so AV block- first degree- and RBBB were improved after correction of hyperkalemia. CPK-MB and Troponin-I were measured to rule out coronary artery disease (CAD); CPK-MB was 1.3IU/L and Troponin-I was 0.1ng/ml. His 24hour Holter-monitoring showed ventricular pause up to 17seconds during syncopeepisodes (Figure 1B). At the time, a temporary pacemaker was implanted. Radiofrequency catheter ablation (RFCA) with bidirectional cavo-tricuspid isthmus block of atrial flutter was done. After RFCA, the ECG showed marked first-degree AV block and bifascicular block (RBBB and LAFB) and (Figure 1C); the 24hour Holter specified an alternating second-degree AV heart block. Owing to frequent episodes of symptomatic high-degree AV block, permanent DDD type of pacemakerwas implanted for him (Figure 1D). His siblings' ECGs presented trifascicular block and their pedigree indicated an inheritance of autosomal dominance as well. Two-Dimentional echocardiography was unremarkable with LVEF 62%. He was discharged after 5days of admission.

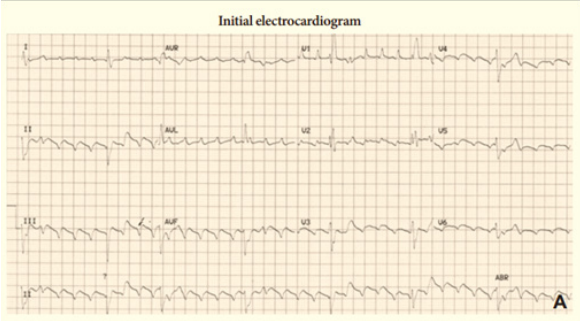

Figure 1A Initial ECG shows atrial flutter with bifascicular block; RBBB and LAFB (A). Twenty four-hour Holter-monitoring illustrates asystole.

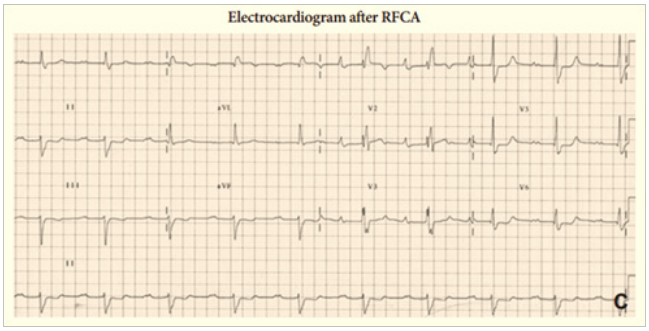

Figure 1B ECG after RFCA shows trifascicular block; first-degree atrioventricular block, LAFB and RBBB

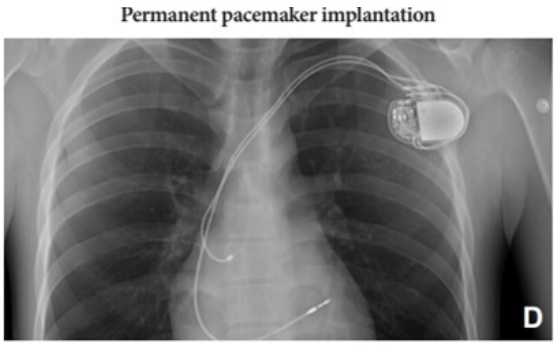

Figure 1C The patient received a permanent pacemaker implantation.

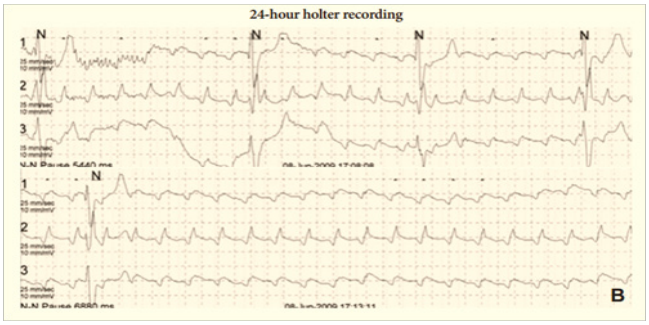

Figure 1D ECG, electrocardiography, RBBB, right bundle branch block, LAFB, left anterior fascicular block, RFCA, Radio frequency catheter ablation.

Discussion

In 1977, Brink et al.[2] pronounced the existence of a new hereditary familial cardiac disease of PFHB with an autosomal dominant inheritance, primarily disturbing the conducting tissue of heart. The ECG pattern of type I PFHB is delineated by confirmation of RBBB, LAFB, and PR interval prolongation or complete heart block with wide QRS complexes. These ECG features distinguish type I PFHB from type II PFHB in which the commencement of third-degree heart block is linked with narrow QRS complexe.[1] Type I PFHB is expressed clinically while third-degree heart block occurs, presenting as dyspnea, syncopal episodes, and sudden death. Etiology and pathophysiology of this condition is not known clearly. Nevertheless, genetic investigations including analysis of linkage and positioning cloning propose a means of identification of some genes which cause this condition. Brink PA et al.,[5–8] by means of linkage analysis, clarified that type I PFHB was naturally mapped to a group of 4 linked loci on 19q13.2-13.3 chromosome. Previous study[5] gathered evidence for KCNA7 exclusion which is located in a region of type I PFHB locus, as the type I PFHB-causative gene. Direct sequencing of KCNA7's coding sequence in type I PFHB-affected ones revealed no pathologic alterations in the sequence, though 2 single nucleotide polymorphisms(SNP) were spotted in exon two yielded amino-acid replacements. Lately, type I PFHB was plotted to chromosome 19q13, and a new causal gene, TRPM4, a member of the potential melastatin family transient ion channel genes receptor, was acknowledged by positional cloning.[6,8]

Treating type I PFHB is accomplished by the appropriate implanting a permanent pacemaker; treating with a prophylactic pacemaker is still debatable.[6] Follow-up with ECG at least for six months is suggested in those with any heart conduction block plus a yearly examination, at least, is also acclaimed among members of an affected families with unremarkable ECG.[9] Although the worldwide incidence of type I PFHB is not clear, this condition might not be limited only to some regions in Africa. Very few reports addressing hereditary tendency of brad arrhythmia among Korean nation;[10] the current article would be the first reported case of documented type I PFHB in Iran. Besides, our current case also indicates an unusual presentation of hyperkalemia-induced trifascicular block which subsided to LAFB with reducing the level of serum potassium. Even though electrophysiological studies at AV node and His-Purkinje system might be required to authorize our theory which hyperkalemia prompted the ECG pattern of trifascicular block in our mentioned patient, the absence of other conditions, such as medication/drug overdose, myopericarditis, rheumatic fever and acute myocardial ischemia suggests that hyperkalemia may be the underlying etiology. The resolution of trifascicular block to LAFB with strict correction of hyperkalemia also confirms that this electrolyte imbalance may produce the higher-grade AV block. The AV node is recognized to be vulnerable to potassium increase, resulting in prolonged PR interval and widening of QRS seen frequently in setting of hyperkalemia. In the current case, we proposed that trifascicular block occurred due to AV node and Purkinje fibers susceptibility to hyperkalemia. Previous report[11] introduced a case of RBBB with left axis deviation which resoluted with adjustment of potassium derangement. Another study[12] reported that 12 patients showing ECG evidence of fascicular block while hyperkalemic state. Hence, hyperkalemia is a regularly occurred electrolyte aberration which may yield life-threatening

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instabilities of heart conduction system. Physicians must be attentive to the different range of dysrhythmias ascribed to hyperkalemia, such as trifascicular block, and must quickly treat hyperkalemia in order to lessen complications.

Conclusion

This interesting but critical condition could be associated to both etiologies, genetically and/or electrolyte disruption discussed above or even synergically augmented by each of them leading to a fatal single outcome. Our current case report highpoints an uncommon reported dysrhythmia allied to genetic abnormality and hyperkalemia of which medical doctors and residents should be aware.

Acknowledgements

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Conflict of interest

Author declares that there is no conflict of interest.

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