Hypertrophic Cardiomyopathy with masked Atrioventricular Block. Report of 2 cases

Cardiomyopathie hypertrophique avec bloc auriculo-ventriculaire masqué. Rapport de 2 cas

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ABSTRACT

Background. Hypertrophic cardiomyopathy is the most common genetic cardiac disorder in Algeria, although syncope is often caused by ventricular arrhythmias or left outflow tract obstruction, it may also be related to complete atrioventricular block; the latter is rarely reported in the literature, the diagnosis is not so obvious, and should be thoroughly researched in presence of syncope. We report two cases of patients with hypertrophic cardiomyopathy and atrio ventricular block revealed by syncope. **Cases' presentation.** We reported two patients who presented hypertrophic cardiomyopathy, with recurrent syncope; one patient presented accessory pathway which masked the complete atrio ventricular block, and the other patient presented alternating occurrence of right bundle branch block and left bundle branch block at the ECG monitoring over a 48 hour. Echocardiography showed hypertrophy of left ventricular walls, but no left ventricular outflow tract gradient was detected. Extensive fibrosis especially in the septum was detected by late gadolinium enhancement. The two patients were implanted with dual chamber pacemakers. During follow up, one patient had developed refractory heart failure and died in 2014. **Conclusions.** Accessory pathway may mask complete atrioventricular block, also alternating bundle branch block could be the only proof of complete atrioventricular block. Fibrosis is the principal substrate of reentry phenomenon, but septal fibrosis can also damage atrioventricular conduction system. Non obstructive hypertrophic cardiomyopathy could be related to fibrosis, and/or asynchrony.

Keywords: Convulsive Syncope, Sudden Cardiac Death, Alternating Bundle Branch Block, Accessory Pathway.

BACKGROUND

Hypertrophic cardiomyopathy (HCM) is a genetic cardiac disease ; it is characterized by left ventricular hypertrophy in the absence of another cause for the increased cardiac mass, it's prevalence in the general population is estimated to be at least 1/500. [1]

Hypertrophic cardiomyopathy is also the most common cause of sudden cardiac death (SCD) in young people [2]. Syncope in patient with HCM could be related to different causes: ventricular arrhythmias, atrial arrhythmias, left ventricular outflow obstruction, vasovagal syncope, sinus node dysfunction and also complete atrioventricular block.

RÉSUMÉ

Introduction. La cardiomyopathie hypertrophique est la pathologie génétique la plus fréquente en Algérie, bien que la syncope soit souvent causée par des troubles du rythme ventriculaire ou une obstruction de la chambre de chasse ventriculaire gauche, elle peut être liée à un bloc auriculo ventriculaire complet, ce dernier est rarement rapporté dans la littérature, son diagnostic n'est pas évident, et doit être rechercher minutieusement. Nous rapportons deux cas de patients atteints de cardiomyopathie hypertrophique et de bloc atrio-ventriculaire révélés par une syncope. **Présentation des cas.** Nous rapportons deux patients, une femme et un homme qui présentaient une cardiomyopathie hypertrophique avec des syncopes à répétition, la femme avait une voie accessoire qui masquait le bloc auriculo ventriculaire complet, et l'homme avait un bloc de branche gauche alternant avec un bloc de branche droit durant l'enregistrement continue sur 48 heures. L'échocardiographie doppler, avait montré une hypertrophie ventriculaire gauche importante mais sans obstruction, chez les deux patients. La fibrose diffuse dans le septum a été objectivée grâce à la fixation tardive du gadolinium à l'imagerie par résonance magnétique. Les deux patients ont été implantés par des pacemakers double chambre, durant le suivi, la femme a développé une insuffisance cardiaque réfractaire et décédée en 2014. **Conclusion.** Dans la cardiomyopathie hypertrophique, la voie accessoire peut masquer le bloc auriculo ventriculaire, le bloc de branche alternant peut être la seule preuve du bloc auriculo ventriculaire complet. La fibrose diffuse est le substratum anatomique de la fibrose, mais peut également détruire les voies de conduction. L'absence de l'obstruction de la chambre de chasse peut être en rapport avec la fibrose ou l'asynchronisme.

Mot clés: état de mal syncopal, mort subite cardiaque, bloc de branche alternant, voie accessoire.

The Complete Atrioventricular Block (CAVB) is rarely reported [3-7], the diagnosis is not so obvious, and should be thoroughly researched in presence of syncope.

We reported two cases of patients with hypertrophic cardiomyopathy and atrio ventricular block revealed by syncope.

CASE 1

A 24-year-old female, followed in our consultation since 2005, she presented dyspnoea, with previous medical history of HCM and accessory pathway (figure 1), she was also being followed for recurrent seizures despite anticonvulsants drugs; in 2011 she

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presented several episodes of palpitations, and 48 hours ECG recording repeated several times showed ventricular premature beats, echocardiography Doppler showed biventricular hypertrophy without obstruction, and MRI showed extensive fibrosis everywhere in the left ventricle walls.

So, we prescribed Amiodarone and after 2 months she had been resuscitated from sudden cardiac death, and a temporary trans venous ventricular pacemaker was inserted as an emergency, because of ventricular asystole. (Figure 2)

Figure 1. ECG 1st patient Accessory pathway masked Complete Atrioventricular Block (CAVB)

Subsequently, a permanent dual chamber pacemaker was implanted to her, after implantation, she no longer had seizures, anticonvulsants drugs were withdrawn, at this time we realized that she suffered from convulsive syncope related

to paroxysmal CAVB, in this case Amiodarone blocked accessory pathway and unmasked CAVB, unfortunately she had developed refractory heart failure and died in 2014.

Figure 2. ECG 1st patient: Complete Atrioventricular Block after blocking accessory pathway with Amiodarone

CASE 2

A 23-year-old men, who was referred to our consultation in 2017 because of several episodes of syncope, surface ECG showed left ventricular hypertrophy (LVH) with left bundle branch block (LBBB), ECG monitoring over a 48 hour,

revealed alternating occurrence of right bundle branch block (RBBB) and left bundle branch block (LBBB) (Figure 3), echocardiography Doppler showed biventricular hypertrophy (figure 4) without obstruction, intra ventricular gradient 17

mm Hg, (at rest or during Valsalva maneuver) and MRI showed septal fibrosis, he was implanted by dual chamber pacemaker, three months after implantation, he became

depend on pacemaker, because paroxysmal CAVB has become permanent.

For our patients, all other causes of left ventricle hypertrophy were ruled out.

Figure 3. Patient 2: Holter monitor: Alternating occurrence of (RBBB) and (LBBB)
RBBB: right bundle brunch block. **LBBB:** left bundle branch block

DISCUSSION

Hypertrophic cardiomyopathy is a genetic cardiac disease; characterized by left ventricular hypertrophy in the absence of another cause for the increased cardiac mass, it's prevalence in the general population is estimated to be at least 1/500. [1]

It is characterized by a heterogeneous clinical presentation, ranging from asymptomatic, mildly symptomatic or very symptomatic forms.

Sudden cardiac death is a devastating and often unpredictable complication; it is thought to be related to ventricular arrhythmias. [2]

Syncope or convulsive syncope is a challenging clinical diagnosis, it could be related to many causes benign or malignant: supra ventricular or ventricular arrhythmias, left outflow ventricular obstruction, conduction disturbances, or vasovagal syncope.

According to the literature, association of AV block and HCM is very rare, Fananapazir et al demonstrated abnormal His-Purkinje conduction in 23 % of HCM patients who survived sudden cardiac death and Barriaes-Villa et al reported 3.5 % of 451 patients with 2nd or 3rd degree AV block. [3-7].

For our first patient, sudden cardiac death is caused by ventricular asystole, due to blocked accessory pathway with Amiodarone; combination of HCM, accessory pathway and CAVB had never been reported in the literature, and also convulsive syncope was misdiagnosed for several years as seizures.

For our second patient, syncope was caused by paroxysmal CAVB, and repeated Holter monitor, revealed alternating bundle branch block.

The physiopathology of CAVB in human HCM is not clear; MARON and colleagues reported a case of congenital CAVB and HCM in 1981.

Figure 4. Patient 2, Echocardiography Doppler showed biventricular hypertrophy

Other histopathologic aspects, such as interstitial fibrosis or myocardial necrosis in the conduction system, abnormal intramural coronary arteries (small with thickened walls), have been reported in some patients with HCM and advanced atrioventricular conduction disorders [8].

Some factors such as thickened walls, ischemia, and fibrosis contribute to arrhythmias and sudden death and may also contribute to conduction disturbances.

The utility of late gadolinium enhancement (LGE) for detecting myocardial fibrosis is well established, its prognostic value has been described and significant relationships with SCD have been demonstrated [9].

It has also been reported that alternating BBB phenomenon is a consequence of an extensive damage of infranodal conduction system with potential high risk of progression to CAVB in ischemic cardiomyopathy but never in HCM [10].

Our patients had extensive fibrosis in late gadolinium enhancement (LGE), especially in the septum, but for the first patient we are not sure about the origin of her CAVB, congenital or fibrosis.

The two patients had non obstructive hypertrophic cardiomyopathy, this form could be related to fibrosis or asynchrony which caused by bundle branch block, or accessory pathway.

To the best of our knowledge, this is the first report of association of CAVB, accessory pathway and non-obstructive HCM. Also, it's the first report of alternating BBB in non-obstructive hypertrophic cardiomyopathy.

CONCLUSION

The literature, suggests that atrioventricular block is rarely observed in patients with hypertrophic cardiomyopathy, but this combination should be considered in presence of syncope.

Accessory pathway may mask CAVB, also alternating BBB could be the only proof of CAVB.

Fibrosis is the principal substrate of reentry phenomenon, but septal fibrosis can also damage atrioventricular conduction system.

Non obstructive hypertrophic cardiomyopathy could be related to fibrosis, and/or asynchrony.

CONFLICT OF INTEREST

The authors declare that they have no conflict of interests.

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