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A Case of Hypertrophic Cardiomyopathy Associated with Wolf Parkison White Syndrome

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Abstract

Introduction: There cardiomyopathy hypertrophic (CMH) is a structural cell disease cardiac , provider of dead sudden at the house of THE youth of less 40 years old . The association with A syndrome of Wolff - Parkinson -W hite (WPW) was describe but stay However rare . We let's report A case of a cardiomyopathy hypertrophic associated has A syndrome of Wolf parkinson white .

Observation: This is a 35-year-old woman residing in Mauritania who consulted for palpitations that had been going on for more than three months, appearing and disappearing suddenly. We found a notion of sudden death in the mother at 54 years old. Physical examination was normal. The electrocardiogram showed ventricular preexcitation with left ventricular hypertrophy. Transthoracic cardiac echocardiography and cardiac MRI concluded that there was a quasi-symmetrical non-obstructive hypertrophic cardiomyopathy, with a mitral leak without alteration of left ventricular function. The European Society of Cardiology CMH rhythm score is 5.05. Electrophysiological exploration found a patent accessory pathway with a refractory period of 220 ms, thus indicating an ablation. The ablation was successfully performed. The control electrocardiogram showed disappearance of preexcitation with persistence of a short PR. The patient is currently on a beta blocker. The patient refuses the ICD. Genetic research is underway.

Conclusion: The association of MHC and WPW syndrome is possible and a mutation in the PRKAG2 gene is often described in this association. Rhythm risk stratification is necessary for better management.

Keywords: Hypertrophic Cardiomyopathy; Wolf Parkinson; White -PRKAG2; Dakar (Senegal)

F-Parkinson-White Syndrome

Introduction

There cardiomyopathy hypertrophic (CMH) is a structural cell disease cardiac conditions characterized by segmental or diffuse hypertrophy of the left ventricle [1] . It is a genetic disease with autosomal dominant transmission, very heterogeneous clinically and genetically. Mutations in the PRKAG2 gene have been associated with the existence of an atrioventricular accessory pathway [2]. The association of MHC and syndrome of Wolff - Parkinson - White (WPW) was describe but stay however rare. These patients may potentially be misdiagnosed as HCM or WPW syndrome due to electrocardiographic changes. Early recognition of its entities allows adapted and adequate care. These two conditions cause sudden death in young people. We let's report A case of a cardiomyopathy hypertrophic associated has A syndrome of Wolff - Parkinson - White.

Observation

This is a 35-year-old woman, an accounting assistant, residing in Mauritania who consulted for palpitations with a sudden onset and end. She was referred to our cardiology center for exploration and management of palpitations with an episode of unexplained syncope.

She has been complaining of palpitations for more than a year which have become more and more frequent during the last three months, although no episode of palpitation could be recorded on the surface electrocardiogram. A notion of sudden death in the mother at 54 years old was found during the interrogation. Physical examination was normal.

The electrocardiogram showed a regular sinus rhythm with a heart rate of 64 cycles per minute with a normal QRS axis. The PR interval was around 85msec and there was a delta wave with wide QRS complexes. The delta wave is positive in leads V1V2V3, negative in leads D1 and aVL with a positive concordance of the QRS complex in the precordial leads. Calculation of ventricular overload indices showed left ventricular hypertrophy of the systolic type with a Sokolow -Lyon index of 58 mm. The corrected QT interval was normal. Overall, the electrocardiogram showed Rosenbaum type A ventricular preexcitation. [3] with left ventricular hypertrophy of the systolic type (Figure 1).

Faced with these anomalies, a cardiac ultrasound was requested and revealed hypertrophy of the walls of the left ventricles, predominantly septal, with a thickness measured at 20mm at the septal level and 17mm on the posterior wall of the left ventricle (Figure 2 and 3). The intraventricular gradient was calculated at 27 mmHg (Figure 4). Left ventricular systolic function was normal with ejection fraction calculated at 65%. Cardiac ultrasound revealed a hypertrophic cardiomyopathy with a septal predominance.

Cardiac magnetic resonance imaging (MRI) confirmed the hypertrophic cardiomyopathy, concluding in a quasi-symmetrical non-obstructive hypertrophic cardiomyopathy, with a small mitral leak without alteration of left ventricular function (Figure 5).

Given the electrocardiographic appearance of ventricular preexcitation and hypertrophic cardiomyopathy confirmed by ultrasound and cardiac MRI, the diagnosis of hypertrophic cardiomyopathy associated with Wolff-Parkinson-White syndrome was made. In this context, rhythmic risk stratification is necessary with the performance of a 48-hour rhythmic Holter, the calculation of the CMH rhythm score of the European Society of Cardiology and the electrophysiological study of the accessory pathway.

Rhythmic Holter showed permanent ventricular preexcitation without rhythmic events throughout the 48 hours.

The online CMH rhythm score of the European Society of Cardiology [4] was calculated at 5.05. We proposed implantation of an automatic cardiac defibrillator (ICD) which the patient refused. Faced with the rhythmic risk linked to HCM, the patient was placed on antiarrhythmic treatment based on beta blockers.

Electrophysiological exploration found a patent left anterolateral accessory pathway with a short refractory period of 220 ms. No arrhythmias were triggered during increasing atrial pacing maneuvers. Ablation was proposed to the patient and carried out successfully. The control electrocardiogram showed a disappearance of the preexcitation with persistence of a short PR at 110ms (Figure 6).

Genetic research and an ICD were offered to the patient.

After a 06-month follow-up, no palpitation was reported by the patient.

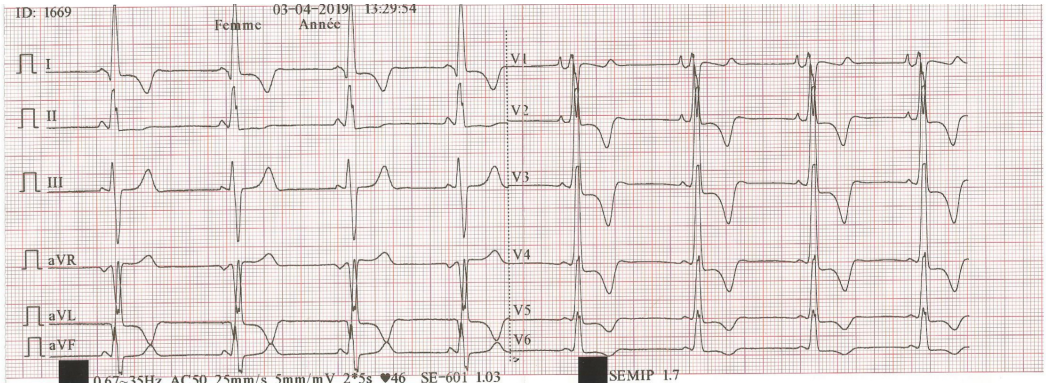

Figure 1: Appearance of ventricular preexcitation with left ventricular hypertrophy of the systolic type

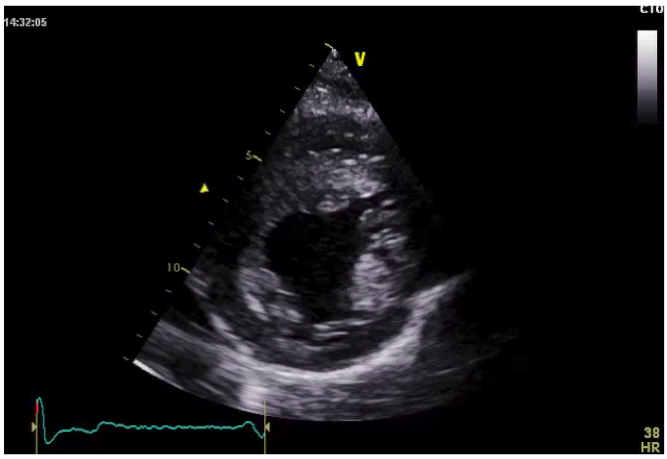

Figure 2: Transventricular short-axis ultrasound section showing hypertrophy of the walls of the left ventricle

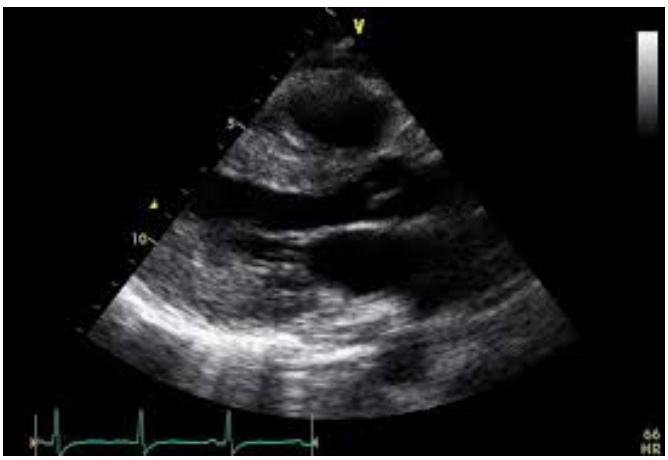

Figure 3: Long axis parasternal section showing thickening of the walls of the left ventricle

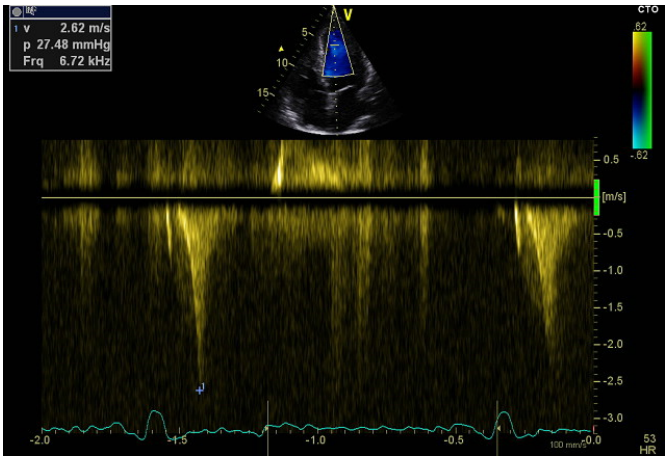

Figure 4: Continuous Doppler with demonstration of a transventricular gradient

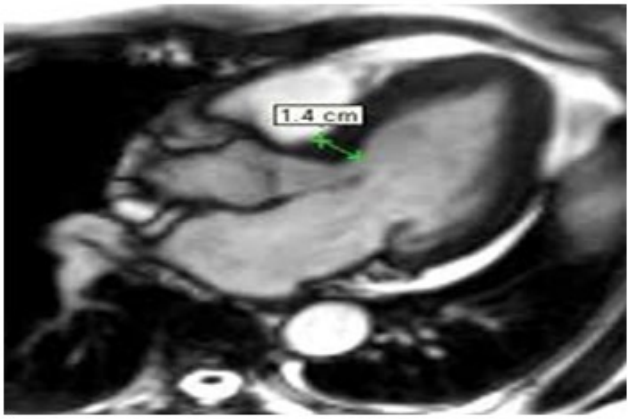

Figure 5: Demonstration of wall hypertrophy on MRI

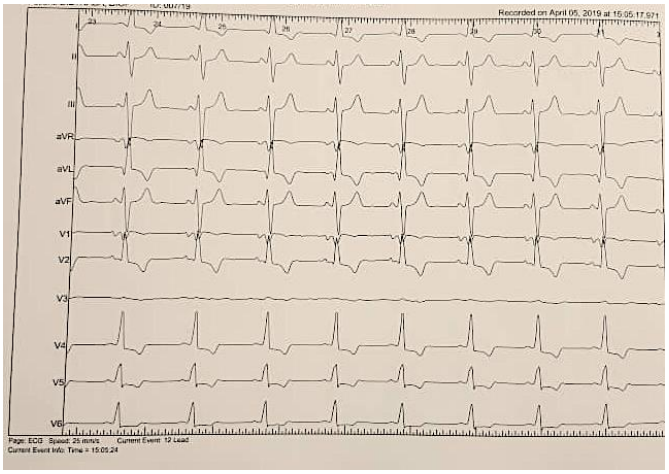

Figure 6: Post ablation ECG showing the persistence of a short PR at 110msec.

Discussion

Wolff-Parkinson-White syndrome (WPW), characterized by an electrocardiographic appearance of abnormal ventricular preexcitation which is manifested on the surface electrocardiogram by a short PR interval less than 120msec, the presence of a delta wave with a wide QRS and paroxysmal tachycardia attacks with abrupt onset and end [5,6]. In our patient, we found the same electrocardiographic aspects suggesting the existence of an accessory pathway. WPW syndrome is characterized by attacks of paroxysmal supraventricular tachycardias as is the case in our patient. It represents a rhythmological entity usually benign but often annoying,

sometimes disabling and dangerous which can lead to sudden death [7]. The action to be taken when faced with a WPW syndrome depends on the type of arrhythmia, its tolerance, the terrain and the electrophysiological characteristics of the accessory pathway(s) [8]. There are new therapeutic approaches and treatment must be early and appropriate as it is a common cause of sudden death [9]. In the case of our patient, an electrophysiological exploration was carried out confirming the existence of the patent accessory pathway with a short refractory period.

Hypertrophic cardiomyopathy is one of the most common inherited cardiac disorders with a prevalence of 1/500 [10] . It is diagnosed by the presence of unexplained left ventricular hypertrophy on cardiac imaging (transthoracic echocardiography, magnetic resonance imaging); myocyte hypertrophy and myofiber disorder during histopathological analysis [11]. In our case, the diagnosis of HCM was made based on hypertrophy of the left ventricular walls confirmed by cardiac MRI. It is a genetic disease that can be responsible for sudden death in young people [12]. Its clinical manifestations are very heterogeneous [13].

The association of CMH and WPW is independently associated with various forms of arrhythmias and can be responsible for sudden death in young subjects [14]. Palpitations in WPW syndrome result from supraventricular arrhythmia such as atrial fibrillation and in HCM, they result from ventricular arrhythmia such as non-sustained ventricular tachycardia [15,16]. In certain situations, atrial fibrillation may be a form of rhythmic complications of HCM [17]. In our patient's case, palpitations were the only symptom but they were not documented by electrocardiographic recording. It was difficult to determine the true culprit of palpitations between HCM and WPW syndrome.

WPW syndrome like CMH can cause sudden death, it is necessary to stratify the rhythmic risk by the CMH rhythm score of the European Society of Cardiology, the rhythmic Holter, the search for fibrosis on MRI and electrophysiological exploration of the accessory pathway [9,18]. The demonstration of a patent accessory pathway with a short refractory period in our patient indicated the indication for successful ablation of the accessory pathway [19]. Regarding HCM, the rhythm risk was intermediate and the insertion of an ICD was recommended but the patient refused the ICD [20, 21].

Gollob et al. identified the PRKAG2 gene, a potentially causative gene in patients with HCM and WPW [22]. In certain familial forms, hypertrophic cardiomyopathy is associated with an aspect of ventricular preexcitation, the PRKAG2 mutation is often described in this association [23]. The presence of ventricular preexcitation is also the result of cellular hypertrophy due to excessive glycogen content, leading to disruption of normal development of the annulus fibrosus, as observed by Wolf et al [24]. We proposed a genetic investigation for our patient.

Conclusion

We described the clinical case of a rare association of hypertrophic cardiomyopathy and WPW syndrome in a patient who underwent a complete screening with ablation of the left anterolateral accessory pathway. This association is possible and a mutation in the PRKAG2 gene is often described. We emphasize the importance of early recognition of this entity, because these patients have a progressive disease with a significant risk of sudden death. Rhythm risk stratification is necessary for better management. Ablation is a radical therapeutic alternative for the accessory pathway. The installation of an automatic cardiac defibrillator is an alternative to prevent the occurrence of sudden death. Future possibilities for gene therapy could revolutionize the management of this disease.

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