MITRAL ANNULAR DISJUNCTION AND ARRHYTHMIC RISK: A CASE REPORT

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Abstract

Mitral annular disjunction (MAD) is a structural abnormality of the mitral annulus, defined by a detachment of the atrial wall-mitral valvular junction from the left ventricular (LV) free wall. This structural abnormality in mitral annulus fibrosus is significantly associated with the presence of mitral valve prolapse (MVP) [1], but it can be also detected in normal cases as an anatomical variation of the mitral annulus fibrosus [2]. The main tools to detect MAD are echocardiography and cardiac magnetic resonance. Prevalence of MAD in the general population ranges from 7% [1] to 9% [2]. Several studies proved an association between MAD and arrhythmic events, independently of concomitant mitral valve abnormalities, suggesting the existence of a novel entity: MAD arrhythmic syndrome [3]. Herein we describe a case report of a middle age man with MAD who experienced ventricular arrhythmias and placement of an implantable cardioverter-defibrillator (ICD).

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Shortened title: MITRAL ANNULAR DISJUNCTION

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This structural abnormality in mitral annulus fibrosus is significantly associated with the presence of mitral valve prolapse (MVP) [1], but it can be also detected in normal cases as an anatomical variation of the mitral annulus fibrosus [2]. The main tools to detect MAD are echocardiography and cardiac magnetic resonance. Prevalence of MAD in the general population ranges from 7% [1] to 9% [2].

Several studies proved an association between MAD and arrhythmic events, independently of concomitant mitral valve abnormalities, suggesting the existence of a novel entity: MAD arrhythmic syndrome [3].

Herein we describe a case report of a middle age man with MAD who experienced ventricular arrhythmias and placement of an implantable cardioverter-defibrillator (ICD).

Keywords : Mitral annular disjunction, ventricular arrhythmias, echocardiography, cardiac magnetic resonance

Case report

A 55-year-old man has been sent, by his trusted cardiologist, to our hospital to implant an ICD.

He was affected by paroxysmal atrial fibrillation, not on anticoagulant therapy, because of a CHADsVASC 0, and not on antiarrhythmic therapy, except for propafenone 'pill in the pocket', with a family history of Arrhythmogenic Right Ventricular Dysplasia (ARVD) and unobstructed arteries at the invasive coronary angiogram performed two years before. In the previous years he also experienced several presyncopal episodes.

His resting echocardiogram showed a normal left ventricular systolic function, with an ejection fraction of 55%, and a mild bi-leaflet mitral valve prolapse with moderate mitral regurgitation. Interestingly, on parasternal long-axis view, MAD was well seen in the posterolateral LV region of approximately 11 mm (figure 1). No other significant structural abnormalities were seen. The trusted cardiologist decided to carry out a cardiac magnetic resonance, that showed no signs of ARVD and proved the presence of MAD. End systolic cine MR sequences in 4-chamber (figure 2) and 3-chamber view (figure 3) demonstrated annular disjunction of the mitral valve and short-axis Phase-sensitive Inversion Recovery (PSIR) sequences, after contrast injection (figure 4, 5), pointed out late gadolinium enhancement of the posterolateral annular ring and of the posterior papillary muscle.

It was therefore decided to implant a Loop Recorder in the patient, that after several months recorded an episode of fast ventricular tachycardia (TV) at 200 bpm for about 5 seconds without symptoms.

In the light of the above findings, we agreed to implant an ICD and the patient was discharged the day after.

Five months later the patient presented to our hospital because of a single shock of ICD, due to an episode of ventricular tachycardia. The patient was then hospitalized and first treated with intravenous amiodarone and than with amiodarone orally. We performed an ICD interrogation, which confirmed the episode of ventricular arrhytmia, and after three days, during which the patient was stable and asymptomatic, he was discharged home with amiodarone.

Discussion

Our case report deals with an obscure anatomic variation, probably a congenital abnormality, of the mitral annulus in which there is still little literature. For the first time, in a study of 900 hearts from adult autopsies,

Hutchins et al. in 1986 reported a strong association between the mitral annular disjunction and the presence of MVP (92% of typical floppy mitral valves showed MAD); they also found out that it could be detected in approximately 6% of patients without MVP [1]. This feature was also clinically confirmed by Konda et al. in a study performed in order to investigate frequencies and characteristics of mitral annular disjunction in patients referred to an echocardiography laboratory. They even found a significantly larger percentage of cases (12%) with mitral annular disjunction but no floppy mitral valve [2]. They also reported meaningfully larger distance of the mitral annular disjunction in patients affected by MVP than in patients without it and was not founded any relationship between mitral annular disjunction and the degree of mitral regurgitation (MR).

An increased frequency of premature ventricular beats, chest pain and non-sustained ventricular tachycardia was firstly demonstrated by Carmo et al. in patients with MAD in the setting of MVP [4]; this was subsequently proved by the examination of 116 patients with MAD by Dejgaard et al. This is the first large clinical trial on patients with MAD where the authors examined patients with MAD clinically and by echocardiography and cardiac magnetic resonance (CMR), in order to describe the clinical presentation, MAD morphology, association with MVP and ventricular arrhythmias. The most important finding was, however, the high occurrence of ventricular arrhythmias in patients with MAD, independently of concomitant MVP, suggesting the existence of a novel clinical syndrome: MAD arrhythmic syndrome. They also identified young age, lower ejection fraction and papillary muscle fibrosis as markers for severe arrhythmic events. Interestingly, MAD patients with MVP were less likely to have experienced severe arrhythmic events, unlike other papers [3].

The association between MAD and arrhythmic MVP was provided by Basso and Perazzolo Marra. They postulated a cascade of events caused by MAD and leading to electrical instability and also hypothesized papillary muscle, inferobasal wall stretch and fibrosis as arrhythmogenic areas in MVP. The reason of this postulation was the observation of premature ventricular beats originating from these LV sites [5] [6]. Posterior systolic curling caused by MAD might create a mechanical stretch of the inferobasal wall and papillary muscle (PM), leading to myocardial hypertrophy and scarring [7]. The association between LV fibrosis and ventricular arrhythmias in MAD was than confirmed by Dejgaard et al. [3]. MAD itself, even independently from MVP, can therefore account for mechanical stretch of the myocardium and arrhythmogenesis [8].

The main instruments to detect MAD are echocardiography and cardiac magnetic resonance. Absence of myocardium during systole between the posterior mitral valve annulus and adjacent basal segments of the ventricular wall is representative of MAD. On echocardiography, this is most commonly seen in the parasternal long-axis view [9].

Studies focused on MAD, using CMR, confirmed that in patients with late gadolinium enhancement (LGE), presence of LV fibrosis at level of papillary muscle was more frequent in patient with MAD and, according to other studies, showed that patients with MAD have the tendency to be younger and with lower ejection fraction (LVEF), irrespectively of MR severity [10].

A review of several studies showed up that the occurrence of ventricular arrhythmias was higher with a greater extent of MAD distance and circumferential area, highlighting the link between electrical instability and extent of MAD [9]. A MAD with disjunction of >8.5 mm was revealed to be prognostic of an increased risk of ventricular arrhythmias [4].

Recently, since MAD has been suggested as a possible cause of ventricular arrhythmias and sudden cardiac death, Scheirlynck et al. carried out a study to explore the presence of soluble suppression of tumorigenicity-2 (sST2) and transforming growth factor-β1 (TGFβ1) as stretch-related and fibrosis-related biomarkers in MAD patients. They found that circulating sST2 levels were higher in patients with MAD and ventricular arrhythmias, compared with arrhythmia-free patients, while TGFβ1 levels did not differ. Interestingly, they hypothesized that the combination of sST2, LVEF and LGE assessment might detect high-risk individuals [11].

Conclusions

MAD could be associated to LV morphological and functional remodeling, which could be the substrate of ventricular arrhythmias and sudden cardiac death. MAD itself should be considered as an arrhythmogenic entity, irrespective of the presence of MR or MVP, and implantation of an ICD, as for our patient, can be life saving.

In conclusion, we can not consider MAD as a well-known entity and more research and further studies are warranted.

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Figure legends

Figure 1: MAD in parasternal long-axes view (transthoracic echocardiography)

Figure 2: Annular disjunction of the mitral valve in end systolic cine MR sequences in 4-chamber (white arrows).

Figure 3: Annular disjunction of the mitral valve in end systolic cine MR sequences in 3-chamber view (white arrows).

Figure 4 and 5: Short-axis PSIR sequences after contrast injection show late gadolinium enhancement of the posterolateral annular ring (black arrows) and of the posterior papillary muscle (arrowhead).