Rare disease

Accessory mitral valve without subaortic obstruction of left ventricular outflow tract in a middle-aged male

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Summary

Accessory mitral valve (AMV) is a rare congenital abnormality with a usually early-age clinical onset, being potentially a cause of subvalvular obstruction of the left ventricular outflow tract. This report describes the case of a 60-year-old patient presented with palpitations and chest pain. Primary evaluation revealed a ventricular tachycardia episode while transthoracic echocardiography showed an intracardiac additional structure at the level of the left ventricular outflow tract. After transoesophageal echocardiography and paraclinical investigations this structure was proven to be an AMV tissue which did not provoke left ventricular outflow obstruction. This case presents an unusual late-age clinical onset of AMV without a clinically significant LVOT (left ventricular outflow tract) obstruction and highlights the importance of transthoracic and transoesophageal echocardiography in the diagnosis of this rare cardiological entity.

BACKGROUND

Accessory mitral valve (AMV) belongs to a group of various anatomical lesions that might produce clinical findings of subaortic stenosis. It was first described by MacLean *et al.*¹ AMV is usually associated with other congenital abnormalities, such as ventricular septal defect and transposition of great vessels.^{2–3} Fewer than 100 cases of AMV have been reported in the literature, most of which have been in children.⁴ We report the relatively rare, easily misdiagnosed cardiological entity called AMV in a middle-aged man without significant clinical LVOT (left ventricular outflow tract obstruction) who was presented with retrosternal pain and ventricular tachycardia.

CASE PRESENTATION

A 60-year-old man presented in our institution referring retrosternal chest pain and palpitations. ECG revealed ventricular tachycardia (220 bpm; figure 1). The patient's history included long-standing hypertension, a single episode of supraventricular tachycardia and smoking. On physical examination, blood pressure was normal and cardiac auscultation showed a 2/4 diastolic murmur along the left sternal border and 1/6 systolic murmur at the cardiac apex. Blood and biochemical laboratory tests were between normal limits without any markable electrolytic disturbance. Thus, ventricular tachycardia was successfully cardioverted to sinus rhythm with the use of intravenous lidocaine 60 mg (dosage 1 mg/kg, patient's weight approximately 65 kg; figure 1). The retrosternal pain disappeared after the cardioversion.

Furthermore, transthoracic echocardiography examination was performed and revealed a parachute-like structure attached to the anterior mitral valve leaflet. The maximal velocity between aorta and left ventricle was measured as 1.9 m/s. We observed a mild degree of mitral regurgitation and a mild-to-moderate degree of aortic regurgitation. Pulmonary stenosis was absent. Pulmonary artery systolic

pressure (PASP) and right ventricle systolic pressure (RVSP) were in high-normal values (RVSP=PASP=30 mm Hg). The size of the left atrium was larger than normal (4.4 cm) but the size of the other cardiac chambers were between normal values. Finally, we did not notice any abnormality of the ventricular and interatrial septum and aortic arch.

In addition, we performed transoesophageal echocardiography which revealed a mobile, echogenic membrane-like structure that was moving into LVOT during systole, occupying the subaortic area. The structure was attached to the ventricular side of the distal part of the anterior leaflet of the mitral valve (figure 2A–C). The mitral valve showed mild regurgitation and the aortic valve revealed a moderate degree of regurgitation. The pulmonary and tricuspid valves were normal. We noticed also a mild dilatation of the left atrium and a moderate degree hypertrophy of the left ventricle. Systolic function of the left ventricle was estimated between normal limits (left ventricle end systolic diameter 4 cm, left ventricle end diastolic diameter 5.4 cm, left ventricle ejection fraction 60%) but diastolic was presented dysfunctional (E<A). The function of the right ventricle was normal, as well as the dimensions and morphology of the aortic arch, ascending and descending aorta. The patient underwent coronary angiography imaging test which revealed a non-critical 20–30% stenosis at the left anterior descending branch (LAD). We recommended electrophysiological testing but our patient refused to undergo the procedure. Moreover, stress test was performed and echocardiographic re-evaluation of LVOT gradient revealed no significant increase during stress indicative for LVOT obstruction (resting mean LVOT gradient 20 mm Hg and postexercise mean LVOT gradient 26 mm Hg) as well as he remained symptomless without any arrhythmic events at the ECG recording during the stress test. We considered possible implantation of an implantable cardiac defibrillator (ICD) but since our patient did not belong to high-risk category for sudden cardiac death (left ventricular dysfunction

Figure 1 Patient's ECG at admission (A) and after cardioversion (B). (A) Ventricular tachycardia and (B) sinus rhythm after lidocaine cardioversion.

after myocardial infarction, LVEF<35%, non-ischaemic cardiomyopathy, NYHA class II, III) as well as he had no history of thyroid gland dysfunction or pulmonary function problems we recommended amiodarone which prevents the recurrence of life-threatening ventricular arrhythmias. Total stay in the hospital was 5 days. The patient was consulted endocarditis prophylaxis, prophylactic therapy for recurrent episodes of ventricular tachycardia with peros amiodarone 200 mg two times per day and close follow-up with annual transthoracic echocardiographs.

DISCUSSION

AMV is a rare cause of subvalvular IVOT obstruction usually seen in children.⁵ AMV tissue has been variably described as parachute-like, sail-shaped, sac-like, leaflet-like or as a sheet.⁵ During early childhood, systolic ballooning of the AMV into the outflow tract results in a mass effect and subaortic obstruction. Within a few years, the continuing

turbulence produced by an AMV into the IVOT can result in permanent deposits of fibrous tissue and subaortic obstruction.⁶ There have been few reports concerning the progression of IVOT obstruction in adult patients with AMV. In fact, the incidence, natural history and embryological development of this malformation are not fully understood. The embryological development seems to involve an incomplete, an abnormal separation of the mitral valve from the endocardium during cardiac development.⁴

The condition is usually presented as an asymptomatic heart murmur although the youngest patients have low cardiac output and congestive heart failure at 14 months. Furthermore, the condition usually presents signs of IVOT obstruction, which generally develop during the first decade with symptoms such as exercise intolerance, chest pain on exertion and syncope. Our middle-aged patient described none of these symptoms during his life as well as he did not develop IVOT obstruction. Usually the

Figure 2 (A–C) Transoesophageal echocardiogram in different views showing accessory mitral valve (AMV) tissue attached to ventricular aspect of anterior mitral valve leaflet prolapsing into left ventricular outflow tract in systole (arrows).

symptoms are presented when the mean gradient across the IVOT is higher than 50 mm Hg.⁷ In our case, the patient did not present any echocardiographic evidence (gradient >50 mm Hg, systolic anterior motion of the mitral valve or dagger-shaped appearance at continuous Doppler of IVOT) of hypertrophic obstructive cardiomyopathy (HOCM). Thus, we considered MRI an exceeding test for further investigation. Moreover, the condition is classified at the basis of intraoperative descriptions and anatomic presentation into two types. Type I AMV which is defined as a fixed mass presenting with a nodular or membranous appearance and type II AMV which occurs as a mobile mass and is classified as pedunculated or leaflet type.^{3 4} The last one is further divided into those with rudimentary chordae tendinae or well-developed chordae tendinae which is the most frequent presentation reported.^{2 3} In the literature there are statistics showing six different locations of the

Video 1 Transoesophageal echocardiogram with an accessory mitral valve (AMV) tissue attached to the ventricular aspect of the anterior mitral valve leaflet prolapsing into left ventricular outflow tract in systole.

insertion of the chordae tendinae of the AMV: the left ventricular wall, accessory papillary muscle, anterolateral papillary muscle, interventricular septum, anterior mitral valve leaflet and the anterior mitral valve chordate.⁴ In our patient the AMV tissue had a parachute-like structure (leaflet-type) that was attached to the ventricular part of the anterior mitral valve (video 1). The VT episode and AMV co-existence described in our case could be considered either a spontaneous or a related to each other event. In the literature we find no evidence for any connection between VT and AMV. Contrary, there is little evidence regarding the correlation of ventricular tachycardia and even supraventricular tachycardia with in general morphological anomalies of the atrioventricular valves. In particular, biopsies and pathologic anatomical studies of these cases revealed structural anomalies of the conduction system without mentioning specifically AMV lesion¹

Owing to the existence of other types of left ventricular masses such as tumours (myxomas and papillary fibroelastomas) or vegetations that can produce similar echocardiographic appearances, AMV should be considered in the differential diagnosis of a cardiac mass.^{3 4 8} Echocardiography especially TEE, is a valuable diagnostic tool for recognising the nature, morphology and attachment point of a mass in the LVOT in a patient with an asymptomatic murmur.^{3 4 8 9} It provides us with significant information to make diagnosis without the use of an invasive approach such as cardiac catheterisation.⁴ During the past decade, the number of reports of AMV has significantly increased as a result of the availability of echocardiography.^{3 4 8} In addition to amiodarone prophylaxis for recurrent episodes of ventricular tachycardia, we recommended a conservative approach including periodic echocardiographic follow-up for early identification of any changes that might warrant surgery. Amiodarone has been associated with toxicity involving the lungs, thyroid gland, liver, eyes, skin and nerves. The frequency of most adverse effects is related to the total amiodarone exposure (ie, dosage and duration of treatment). We considered potential drug toxicity due to long-term amiodarone intake since our patient was 60 years old. Therefore, we recommended the lowest possible dosage of amiodarone, periodical laboratory (liver, thyroid gland and lung

function) tests during his follow-up and if necessary discontinue treatment if adverse effects occur.

Learning points

- ▶ Accessory mitral valve (AMV) may depute without symptoms of left ventricle outflow (LVOT) obstruction although it is a rare cause of LVOT obstruction.
- ▶ Although an early-age—congenital valvular anomaly it could be clinically presented with middle-age onset.
- ▶ If AMV causes left ventricle outflow obstruction it requires surgical repair from the early ages.
- ▶ The most reliable method of AMV diagnosis is transoesophageal echocardiography.

Competing interests None.

Patient consent Obtained.

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