Submitral Aneurysm of the Left Ventricle

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Introduction

Submitral aneurysms of the left ventricle were first reported from Nigeria and other African nations [1]. Several reports from Indian centers of small numbers of cases have highlighted their presentation with left ventricular dysfunction, heart failure and mitral regurgitation with poor outcome both on medical and surgical management [2,3,4]. We report two cases who had presented to our centre in the last six months, highlighting the wide spectrum in the presentation and outcome in this rare disease.

Case Report 1

A 39 year old male presented with recurrent episodes of palpitation. An electrocardiogram (ECG) recorded during palpitations showed sustained ventricular tachycardia (VT) of right bundle branch block (RBBB) morphology with left axis deviation (Fig 1) which was terminated by DC shock. Physical examination, resting ECG, chest radiograph, hematological and biochemical parameters were normal. Echocardiography did not reveal any abnormality with normal mitral flow and left ventricular function. Treadmill test was negative for inducible ischemia. In view of the recurrent episodes of ventricular tachycardia the patient was taken up for electrophysiological studies. The VT however could not be induced on table either by multiple atrial or ventricular extrastimuli with or without isoprenaline infusion. A left ventricular angiogram done showed a submitral aneurysm. Coronary angiography showed right dominant circulation with normal coronaries. The patient has been exhibited to Tablet Amiodarone 200 mg OD. He is asymptomatic on follow up.

Case Report 2

A 14 year old boy presented with progressively worsening dyspnea of one year duration with mitral regurgitation and congestive heart failure (NYHA class IV). He was hospitalised

for stabilisation and evaluation. There was no history suggestive of rheumatic fever. Clinical examination revealed heart rate of 90 per minute, sinus rhythm and blood pressure 100/70 mm Hg. The jugular venous pressure (JVP) was 8 cm above the angle of Louis. He had cardiomegaly and the cardiac apex was left ventricular and sustained. The first sound was muffled and left ventricular third sound was present. A grade 3/6 pansystolic murmur and short mid diastolic murmur were heard at the apex. The ECG showed left ventricular hypertrophy with diastolic overload pattern. Chest radiograph showed cardiomegaly. Transthoracic (Fig 2) and transesophageal echocardiogram revealed left ventricular dilatation and systolic dysfunction with ejection fraction of 40%, severe mitral regurgitation and mild aortic regurgitation. A large chamber was seen lying posterior to the left atrium opening into the posterobasal submitral left ventricle. The neck of the aneurysm was defined in both the parasternal long axis and the cross sectional views. Regurgitation into the chamber was seen on colour imaging in systole.

The patient underwent cardiac catheterisation which showed mild pulmonary hypertension (40/23 mean 30 mm Hg), elevated pulmonary wedge pressure (mean 22 mm Hg) and elevated left ventricle end diastolic pressure (20 mm Hg). Left ventricular angiogram was done which revealed a submitral aneurysm with left ventricular ejection fraction of 40%. The patient was stabilised on diuretics, digoxin, vasodilators and taken up for surgery. The patient however did not come off pump and died after undergoing a mitral valve repair with resection of the aneurysm.

Discussion

Submitral left ventricular aneurysms are rare and have been described in black and negroid people [1]. Sporadic case reports in the last two decades have documented their existence in the Indian population [2,3,4]. Our first case presented with VT with RBBB and left axis deviation which was thought to be a intrafascicular

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Fig. 1 : ECG showing wide QRS tachycardia with left axis deviation and RBBB morphology

Fig. 2 : Transthoracic echocardiogram showing the submitral aneurysm with regurgitant flow from left ventricle in systole

tachycardia in a structurally normal heart. This form of tachycardia is characterized by (1) induction with atrial pacing (2) RBBB, left axis configuration (3) characteristically the QRS is not more than 140 msec in duration (4) the VT is Verapamil sensitive and (5) there is no structural heart disease. It typically manifests in individuals in the age group of 15 to 40 years of age. Sixty to eighty percent of them are males. It is amenable to radio-frequency ablation. It was only

when the tachycardia could not be induced by multiple extrastimuli and isoprenaline infusion that a left ventricular angiogram was done revealing a small submitral aneurysm which was not detected on echocardiography. This highlights the fact that even small aneurysms with normal mitral valve, ventricular dimensions and ejection fraction can be symptomatic.

The second case highlights the importance of considering submitral aneurysm in the differential diagnosis of mitral regurgitation with left ventricular dysfunction and heart failure in the young.

The aetiology of this entity is unknown though a congenital defect in the posterior mitral annulus has been postulated as a probable cause of these aneurysms [3]. Submitral aneurysms coexisting with Takayasu's arteritis [5] and tubercular pericarditis [2] have been reported. The aneurysms are well outlined on magnetic resonance imaging [4]. The clinical presentation can be protean. They can be asymptomatic or associated with ischemia due to compression of the left main and left circumflex coronary arteries, thromboembolism, ventricular arrhythmias, mitral regurgitation and left ventricular dysfunction [2]. The aneurysm neck, arising from below the leaflet of the mitral valve communicating with the left ventricle is usually identified on echocardiography.

Fig. 3 : Left ventriculogram showing the submitral aneurysm

Coronary angiography is required for delineation of coronaries. In our first case the submitral aneurysm was small and detected only on angiography.

Surgery of submitral aneurysms is associated with poor outcome with over 50% mortality in the small number of cases reported and is particularly grave for those with large aneurysms and left ventricular dysfunction.

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