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Submitral aneurysm in adults: A rare entity with varied presentations

Baban Kumar, Santhosh Satheesh, Raja Selvaraj *

Department of Cardiology, Jawaharlal Institute of Postgraduate Medical Education and Research, Puducherry, India

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ABSTRACT

Submitral aneurysm is a rare cardiac entity, initially described in African black patients but rarely also reported in Indian population. Patients may be entirely asymptomatic for many years or have varied clinical manifestations. We are presenting a case series on 4 cases of submitral aneurysm which came to our institution, each of them having varied presentations.

Introduction

Submitral aneurysm (SMA) is a rare cardiac pathology thought to be caused by a congenital defect in the posterior portion of the mitral annulus, occurring commonly in African black patients. But cases have also been described in patients of all races and also in Indians.[1,3,6] They may be asymptomatic, but can produce morbidity due to mitral regurgitation with or without LV dysfunction. They may exhibit varied clinical manifestations like heart failure, systemic thromboembolism, ventricular wall rupture, myocardial ischemia due to compression of coronary arteries, ventricular arrhythmias or sometimes sudden cardiac death.[1–3]

Case series

Case 1

A 35-year-old male presented with worsening dyspnea on exertion since 6 months and orthopnea for one week. There was no history of acute coronary event, trauma, rheumatic fever, tuberculosis or takayasu aortitis. On examination, he had signs of mitral regurgitation and congestive cardiac failure. Transthoracic echocardiography showed a large submitral aneurysm in the posterolateral left ventricle just below the posterior mitral leaflet with severe eccentric mitral regurgitation. Left atrium and left ventricle were dilated along with mild left ventricular systolic dysfunction (LVEF 50%). He was stabilized with decongestive therapy and referred for surgical resection of submitral aneurysm and mitral valve repair/replacement.

Case 2

A 42-year-old male presented with documented wide QRS tachycardia of left bundle branch block morphology with left axis deviation. He gave a history of recurrent such episodes presenting with palpitations and giddiness. Transthoracic and transoesophageal echocardiography showed a wide necked submitral aneurysm measuring 5×5 cm with severe mitral regurgitation. Coronary angiography showed normal coronary arteries. Cardiac MRI clearly demonstrated the aneurysm below the mitral valve. Ventricular tachycardia (VT) of right and left bundle branch block morphologies were induced with single ventricular extrastimuli during electrophysiological testing. The patient was prescribed oral amiodarone and advised surgical resection of submitral aneurysm with mitral valve repair or replacement.

Case 3

A 60-year-old male patient presented with exertional breathlessness and recurrent syncope preceded by palpitations since 2 months. He was hospitalized twice with tachycardia and cardioverted on both occasions due to hemodynamic instability. Twelve lead electrocardiogram showed a wide complex tachycar-

* Corresponding author at: Department of Cardiology, JIPMER, Dhanvantri Nagar, Puducherry, 605006, India.
E-mail address: rajaj.selvaraj@jipmer.edu.in (R. Selvaraj).

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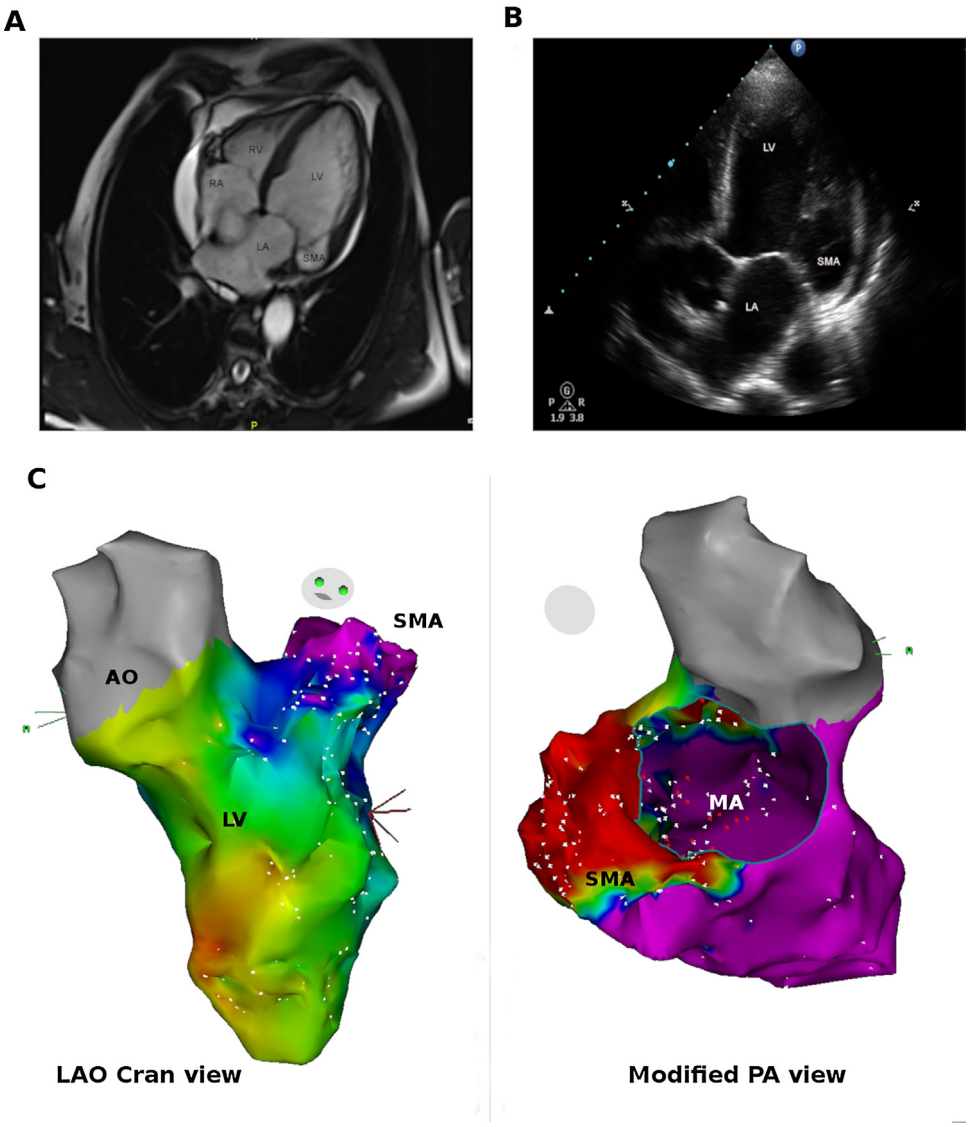

Fig. 1. Submitral aneurysm in patient with ventricular tachycardia.
Panels A and B show four chamber views of the heart by magnetic resonance imaging and echocardiography respectively, demonstrating the submitral aneurysm. Panel C shows the three dimensional voltage map acquired using the CARTO system. An area of low voltage due to scar is seen as a red area at the basal lateral left ventricle.
Ao = Aorta, LV = Left ventricle, MA = Mitral annulus, SMA = Submitral aneurysm

dia at a rate of 160 bpm with right bundle branch morphology, left axis deviation and concordant QRS complexes in precordial leads consistent with ventricular tachycardia. He had a bounding pulse with increased pulse pressure and other signs of aortic regurgitation on physical examination. Echocardiography and cardiac magnetic resonance imaging showed an aneurysm of left ventricle corresponding to submitral position posterolaterally, along with severe aortic regurgitation and moderate mitral regurgitation. Left ventricular systolic function was normal and coronary angiography showed normal coronaries.

During an electrophysiology study, two different VTs were inducible, one of which was similar to the clinically documented VT. Electroanatomic mapping (CARTO, Biosense Webster) showed an area of outpouching with low voltage in the basal lateral left ventricle corresponding to the SMA (Fig. 1). Radiofrequency ablation was performed with an irrigated tip catheter targeting all delayed potentials in the aneurysm. After ablation, no VT was induced with upto three ventricular extrastimuli. During follow-up, he had no recurrence of arrhythmias and is now awaiting aortic valve replacement.

Case 4

A 42-year-old asymptomatic male had undergone mechanical prosthetic mitral valve replacement and tricuspid valve repair 3 years ago for rheumatic heart disease with symptomatic severe mitral and tricuspid regurgitation elsewhere. On examination, he had wide pulse pressure with early diastolic murmur best heard in neoaortic area and well heard prosthetic mitral valve clicks. On 2D transthoracic echocardiography, prosthetic mitral valve was opening well with normal transmitral gradient. A large submitral aneurysm was seen in the posterolateral left ventricle just below the prosthetic mitral valve. Color Doppler showed severe aortic regurgitation with turbulent jet from the right coronary cusp continuing into cystic echolucent cavities in the interventricular septum. Flow could be seen in and out of the aneurysm with sys-