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Case Report

Giant unruptured sinus of Valsalva aneurysm with complete heart block

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ABSTRACT

In this rare case report of giant unruptured sinus of Valsalva aneurysm (SOVA), a 17-year-old male presented with sudden onset syncope due to complete heart block (CHB). An emergency evaluation was done with the help of transthoracic echocardiography, transesophageal echocardiography, and cardiac catheterization with support of temporary pacemaker. The obvious distorting effects of a giant SOVA dissecting into interventricular septum were CHB, significant regurgitation of tricuspid and mitral valve, mild regurgitation aortic valve and biventricular dysfunction. The case was treated by repair of SOVA and posterior mitral ring annuloplasty. CHB improved to sinus rhythm on 11th day after surgery. On follow-up, tricuspid valve regurgitation improved to mild regurgitation and he continued to have mild aortic regurgitation.

<**Learning objective:** Sinus of Valsalva aneurysm (SOVA) may present with cardiac emergency without rupture. A giant unruptured SOVA competes for space with neighboring intra cardiac structures. The distorting effects are erosion into interventricular septum, complete heart block (CHB), valvular and ventricular dysfunction. Transthoracic and transesophageal echocardiography provide enough information for emergency surgery. Sometimes, evaluation may need cardiac catheterization and computed tomography. Immediate surgery saves life. CHB may improve on follow-up.>

Introduction

A congenital sinus of Valsalva aneurysm (SOVA) is usually clinically silent. The presentation may vary from asymptomatic dilatation of coronary cusp detected in routine 2-dimensional echocardiography to symptomatic presentations related to the compression of adjacent structures or intracardiac shunting caused by rupture of the SOVA mostly into the right side of the heart [1]. The compressive features are tricuspid valve regurgitation, right ventricular outflow tract obstruction and rarely complete heart block (CHB). Dissection or erosion into interventricular septum is one of the rarest complications of SOVA [1]. Most of the large or giant SOVA rupture on follow-up [2]. The symptomatic presentation is almost always a surgical emergency. Sometimes catheter-based closure of ruptured SOVA also can be attempted with suitable anatomy.

Case report

A 17-year-old, apparently healthy male presented with sudden onset of syncope. There were no clinical findings of Marfan syndrome or Ehlers–Danlos syndrome. A 12-lead electrocardiogram showed complete heart block (CHB) (Fig. 1A). He was immediately supported by temporary pacing. Routine investigation reports were in the normal limit. Transthoracic (Fig. 1B,C, Video 1) and transesophageal echocardiographic (TTE, TEE) (Fig. 2A–C, Video 2) assessment revealed a giant unruptured SOVA (5.2 cm × 3 cm) of right coronary sinus eroding far into interventricular septum. The apparent protrusion into right ventricular outflow tract (RVOT) and cranial displacement of septal leaflet tricuspid valve (TV) were apparent on TTE and TEE. RVOT gradient was 40 mmHg. There was more than moderate tricuspid valve regurgitation. There was more than grade II aortic valve regurgitation due to poorly coapting right coronary cusp. Mitral valve regurgitation was moderate to severe due to relatively small left ventricular cavity that can be explained by giant aneurysm in the interventricular septum (IVS). The aortic root angiogram in left-anterior oblique 42 degree projection showed unruptured SOVA of right coronary sinus eroding far distal into IVS,

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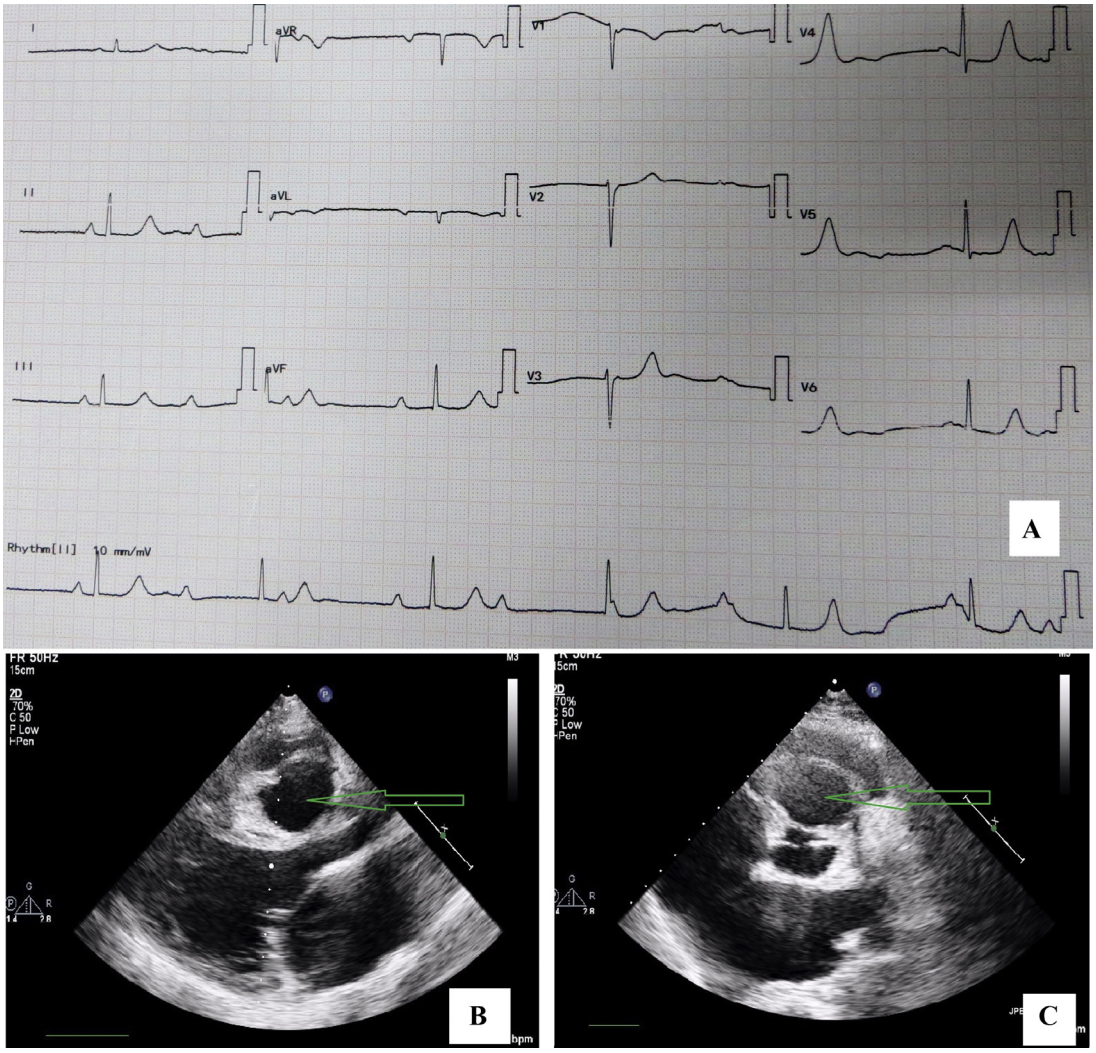

Fig. 1. (A) 12-lead electrocardiogram showing complete heart block at presentation with syncope; (B) Transthoracic echocardiography (TTE) of the left ventricular outflow tract in parasternal long axis (PLAX) which is marked by green arrow demonstrates the unruptured right sinus of Valsalva aneurysm (SOVA); (C) TTE (parasternal short-axis, yellow arrow) shows the large SOVA bulging into the right ventricular outflow tract.

with diastolic opacification (Fig. 2D, Video 3). There was no step-up of oxygen during hemodynamic study. Surgical repair of SOVA and posterior ring annuloplasty of mitral valve (Fig. 3A) were done successfully in emergency. The case was followed with temporary epicardial pacing wire. CHB improved to sinus rhythm (Fig. 3B) on the 11th post-operative day. Tricuspid regurgitation improved after surgery. There was mild residual aortic regurgitation on follow-up.

Discussion

SOVA is a consequence of weakness of the elastic lamina at the junction of the aortic media and aortic annulus fibrosis in either congenital or acquired groups. It is associated with ventricular septal defect in approximately 40% of patients. The incidence of isolated SOVA is rare. It is located to right coronary sinus in 60–85%, noncoronary sinus in 10–30%, and left coronary sinus in 5% of cases with male preponderance (4:1). It accounts for 1% of congenital cardiac defect in grown-up children. The average age of presentation is 30 years and age range is 11–64 years. The one-year follow-up result with medical management is dismal in symptomatic patients with or without rupture. The rupture of SOVA is more frequent into right ventricular outflow tract (60–90%) [1]. The symptomatic aneurysmal dilatation of sinuses is rare in children [3,4]. In general, the rough incidence is 0.09% in the general population. It may be congenital or acquired. The clinical presentation depends upon degree of compression of neighboring structures, competition for intracardiac space, rupture, and dissection into IVS. Therefore, the spectrum of presentation in the descending order of frequency includes rupture, incidental detection without rupture, infective endocarditis, thromboembolism, valvular regurgitation, CHB, and ventricular dysfunction. The associated cardiac defects sometimes include bicuspid aortic valve (9%), ventricular septal defect (34%), and aortic regurgitation (44%) [2,5]. It is sometimes associated with RVOT obstruction or tricuspid valve regurgitation due to encroachment into ROVT and inlet of right ventricle. Unruptured aneurysm may cause distortion and obstruction in the immediate neighboring structures leading to asymptomatic to acute presentation such as severe aortic valve regurgitation, CHB, and acute coronary syndrome. The possible explanation for CHB associated with large aneurysm eroding IVS is compression of bundle of His when aneurysm of right coronary sinus grows caudally distorting or