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Case Report

Aneurysm of Right Sinus of Valsalva: A Particular Case Report and Brief Review of Literature

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Abstract

The aneurysms of sinus of Valsalva are rare cardiac anomalies, which may be either acquired or congenital. A congenital lack of continuity between the aortic media and annulus fibrosus may initiate aneurysm formation. Acquired aneurysms may result from trauma, endocarditis, syphilis, Behcet, atherosclerosis and senile-type dilatation. We report a case of a 38-year-old man who presented with aneurysm of the right sinus of Valsalva with fistula between it and the right ventricular outflow and pulmonary trunk dilatation diagnosed by transthoracic echocardiography (TTE), chest computed tomography (CT) scan and transesophageal ecocardiography (TEE). We describe this case which has been successfully-treated because the concomitant presence of an aneurysm of the right Valsalva sinus, a fistula between it and the right ventricular outflow and pulmonary trunk dilatation represents an extremely rare and poorly described event in the literature.

Keywords: ***Aneurysm, sinus of Valsalva, fistula***

Introduction

The aneurysms of sinus of Valsalva (SVA) are uncommon cardiac anomalies in which there is dilation of one or more of the three aortic sinuses. SVAs may be either congenital or acquired. Congenital SVAs generally arise due to incomplete fusion of the aortopulmonary septum with the interventricular septum, resulting in weakness at the junction of the aortic media and annulus fibrosus which may initiate dilation. Acquired aneurysms, on the other hand, are associated with infections, atherosclerosis, cystic medial necrosis, vasculitic diseases, degenerative diseases, the abuse of drugs or alcoholism. Acquired SVAs may also be secondary to chest trauma or have iatrogenic origin during interventions. Generally, small unruptured SVAs are asymptomatic and are only incidentally found. The expansion of the aneurysm on the adjacent vital structures can cause coronary ischemia, left and right ventricular outflow tract obstruction, and cardiac conduction abnormalities [1]. Rupture of SVA is a potentially fatal complication that should require urgent cardiac surgery. We present a rare case of 38-year-old man with aneurysm of the right sinus of Valsalva with fistula between it and the right ventricular outflow and pulmonary trunk dilatation. A correct and prompt diagnosis in this context may avoid life-threatening complications.

Case presentation

A 38-year-old man was admitted to the emergency unit of our hospital for the sudden onset of chest pain. He was a smoker but had no other cardiovascular risk factors and had no history of relevant diseases. On cardiovascular examination, a rough grade IV/V systolic murmur was present at the mesocardium and blood pressure was 160/90 mmHg. His electrocardiogram showed sinus tachycardia and complete right branch block. On laboratory tests, cardiac biomarkers and d-dimer were negative. A transthoracic echocardiography (TTE) showed left ventricle cavity-sized at the upper limits with preserved systolic function and mild concentric hypertrophy. The aortic root was ectatic with tricuspid valve and mild regurgitation. In addition, there was a strange spheric cavity (1,9x1,8cm) in the region of the right sinus of Valsalva that was communicating with the aortic lumen with turbulent flow inside. The Pulmonary Trunk was dilated and there was a strange turbulent flow within it that extended towards the aortic valve (Figure 1A and 1B). A contrast-enhanced chest computed tomography (CT) scan was performed in emergency which ruled out an aortic dissection and revealed the presence of an aneurysm of the right sinus of Valsalva (Figure 2). Therefore, a transesophageal echocardiography (TEE) was performed which confirmed the aneurysm of the right sinus Valsalva and the presence of systodiastolic flow between it and the right ventricular outflow compatible with a fistula (Figure 3A and 3B). Cardiac Magnetic Resonance (CMR) was not performed because the diagnosis was already evident and would not add further useful information. For this reason, the patient underwent cardiac surgery of resection of aneurysmatic tissue, direct suture of the fistula by the combined transaortic / transpulmonary route and reductive plastic surgery of the pulmonary trunk. At routine outpatient follow-up 6 weeks later, the patient reported resolution of his cardiac murmur and the successful intervention was noted on the transthoracic echocardiogram.

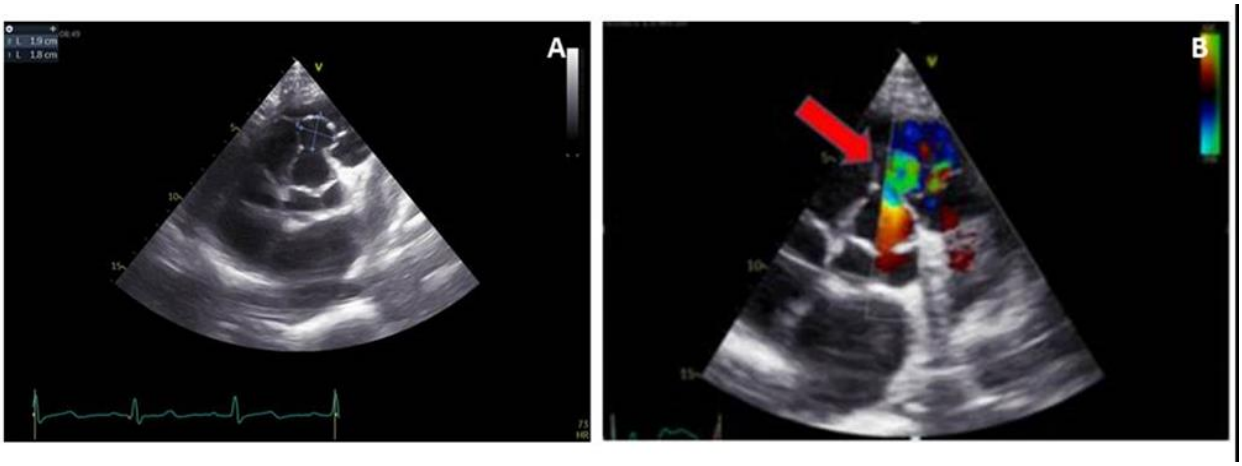

Fig: 1A and 1B Transthoracic echocardiogram showing a spheric cavity (1,9x1,8cm) in the region of the right sinus of Valsalva (A) and an abnormal flow inside it and in the dilated pulmonary trunk (B).

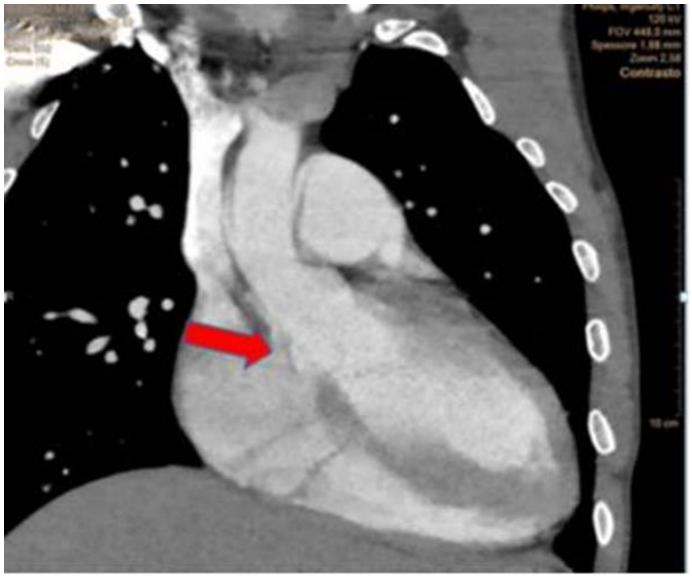

Fig. 2: Computed tomography scan of the chest demonstrating the right sinus of Valsalva aneurysm.

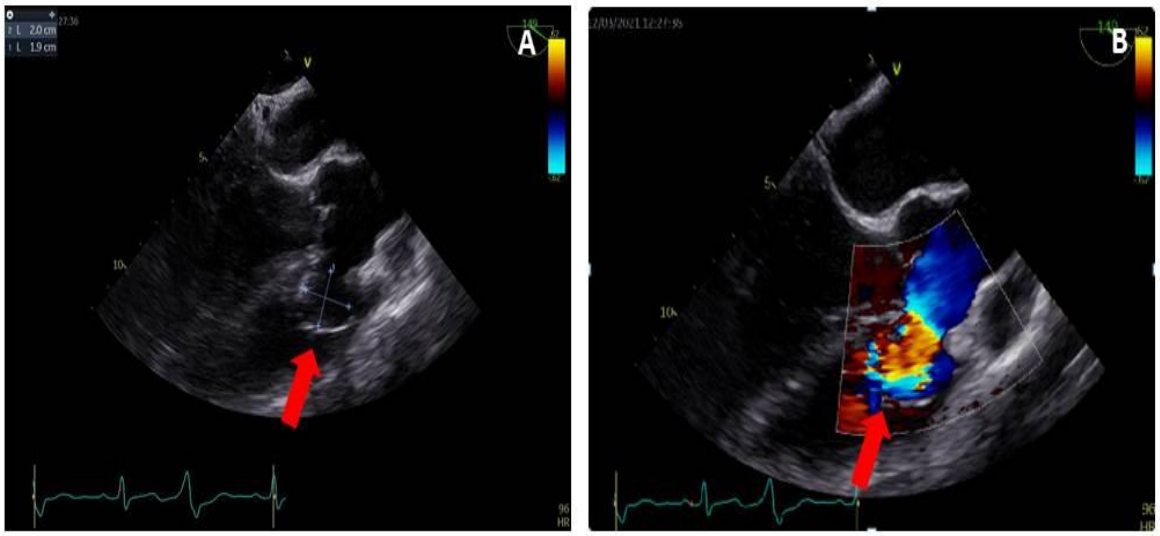

Fig. 3A and 3B Transesophageal echocardiogram showing the presence of aneurysm of the right sinus Valsalva (A) and of the systodiastolic flow between it and the right ventricle compatible with a fistula (B).

Discussion

The sinuses of Valsalva are the three outpouchings of the aortic root located between the aortic annulus and the sinotubular junction. The main function of the normal sinuses is to prevent occlusion of the coronary artery ostia during systole when the aortic valve opens. The aneurysms of sinus of Valsalva (SVA) are rare cardiac anomalies in which there is dilation of one or more of the three aortic sinuses [1]. The etiology of SVAs can be either congenital or acquired as we have already described above. Transthoracic Echocardiography (TTE) plays a key first-line role in the diagnostic evaluation of suspected SVAs, if further assessment is needed TEE may be

reasonable to provide incremental diagnostic information. Imaging with Multidetector Cardiac Computed Tomography (MDCT) and Cardiac Magnetic Resonance (CMR) is helpful for accurate assessment of the entire thoracoabdominal aorta [2]. Established guidelines on the diagnosis and management of sinus of Valsalva aneurysms are lacking. It is clear that in the case of ruptured or unruptured, symptomatic aneurysms, definitive intervention is indicated. In general, it is accepted practice to follow guidelines for repair of aortic root aneurysms [3]. Several clinical cases of sinus of Valsalva aneurysms have been reported in the literature. Some of these are very interesting especially for the clinical presentation and complications described [4-8]. However, there are few well-described cases of right sinus Valsalva aneurysm with fistula in the outflow tract of the right ventricle and simultaneous presence of pulmonary trunk dilation. We describe this rare case precisely to emphasize the importance of early diagnosis in this context and to avoid life-threatening complications. In our case, the transthoracic echocardiogram permitted to identify a strange spherical cavity in the region of the right Valsalva sinus in addition to identifying an abnormal flow inside it and in the dilated pulmonary trunk; a contrast-enhanced chest CT scan enabled the diagnosis of right Valsalva sinus aneurysm. The execution of the transesophageal echocardiogram allowed to better visualize the presence of a turbulent Doppler flow systo-diastolic between the aneurysm of the right Valsalva sinus and the outflow tract of the right ventricle and hypothesize a solution of continuity between these neighboring structures compatible with a fistula. In our case, the etiology of the aneurysm is not very clear. We believe that it could be a congenital aneurysm that has fissured over the years creating a fistula with the right ventricular outflow. This fistula caused a significant left-to-right shunt resulting in haemodynamic changes such as pulmonary trunk dilation. This would explain the situation that we have found with the simultaneous presence of aneurysm of right sinus Valsalva with fistula in the outflow tract of the right ventricle and pulmonary trunk dilation, an extremely rare event that has been poorly described in the literature. The present case report demonstrates how the diagnosis of this pathology is not difficult and above all it can be performed with diagnostic tools available to most hospitals, such as transthoracic and transesophageal echocardiography and CT scan.

Ethics approval and consent to participate

The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with Committee on Publication Ethics (COPE) guidance.

List of abbreviations

TTE: Transthoracic echocardiography;
CT: Chest computed tomography;
TEE: Transesophageal echocardiography;
SVA: Aneurysms of sinus of Valsalva;
CMR: Cardiac Magnetic Resonance;
MDCT: Multidetector Cardiac Computed Tomography.

Data Availability

The case history and reports used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Conflict of interest

We have no conflicts of interest to disclose.

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Author contribution

Luigi Nunziata wrote the draft and collected data; Saverio Ambrosino and Mario Volpicelli provided expertise and feedback; Michele Capasso and Luigi Caliendo revised the manuscript and provided final approval

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Not applicable.

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