Isolated sinus of Valsalva aneurism presenting with dyspnea revealed by multimodality imaging

Authors: Djordje Ilić, Snežana Stojšić, Nikola Komazec, Milenko Rosić, Ranko Zdravković, Golub Samardžija, Aleksandra Milovančev, Aleksandra Ilić

Article type: Clinical vignette

Received: April 17, 2024

Accepted: June 17, 2024

Early publication date: June 19, 2024
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Short title: SOVA

Djordje Ilić¹, Snežana Stojšić², Nikola Komazec², Milenko Rosić¹,², Ranko Zdravković¹,², Golub Samardžija¹,², Aleksandra Milovančev¹,², Aleksandra Ilić¹,²

¹Faculty of Medicine, University of Novi Sad, Novi Sad, Serbia
²Institute of Cardiovascular Diseases of Vojvodina, Sremska Kamenica, Serbia

Correspondence to:
Ass. Prof. Aleksandra Milovančev, MD, PhD,
Faculty of Medicine, University of Novi Sad,
Hajduk Veljkova 3, 21000 Novi Sad, Serbia,
phone: +381 64 27 46 046,
e-mail: aleksandra.milovancev@mf.uns.ac.rs

Isolated sinus of Valsalva aneurysm (SOVA) is a very rare anomaly. Separation between the media and annulus fibrosus causes weakness in the wall leading to the dilatation in one of the aortic sinuses, typically right [1]. Symptoms can vary depending on the compression of immediate structures. Nonetheless undetected and ruptured it can be associated with high mortality [2, 3]. Surgery is a recommended treatment, but it can also be associated with complications [4].

After complaining of exertional dyspnea during a routine gynecological examination a 59-year-old woman was referred to a cardiologist. An ambulatory out-of-hospital transthoracic echocardiogram was suggestive of a large pericardial cyst and the patient was referred to our tertiary care center. In the hospital, transthoracic echocardiogram in parasternal long axis view revealed a circular thin-walled structure located above the Aorta in the projection of the right ventricle (Figure 1A). The 4-chamber view showed a similar structure between the right atrium and ventricle, but its connection with neighboring structures remained unclear.
Upon administering agitated saline, the microbubbles remained confined solely to the right atrium and ventricle. Notably, the thin-walled structure displayed no observable changes (Figure 1B). To obtain left heart chamber opacifications, intravenous transpulmonary echocardiographic contrast agent was administered. Supplementary material, Video S1 shows how contrast agent fills left atrium, ventricle, and ascending aorta revealing a huge dilation of the sinus of Valsalva. Transesophageal echocardiogram on the mid-esophageal short axis view revealed an abnormal dilatation of the sinus of Valsalva aneurism of the right coronary cusp (Figure 1C). A cardiac computed tomography confirmed a giant dilatation of the sinus Valsalva (Figure 1D) with dimensions of $66 \times 47 \times 42$ mm and computed tomography coronary angiography showed normal coronary arteries. Cardiac surgery was performed using the Bentall procedure (Figure 1E). The aortic root and ascending aorta were reconstructed, and a prosthetic valve was implanted. The histopathology of the resected part of the aorta showed thinned media with mucoid degeneration and disrupted elastic tissue (Figure 1F). The postoperative course was uneventful. On one month follow up she was asymptomatic and feeling well.

In our reported case multimodality imaging including contrast agents enhanced timely diagnosis of SOVA, and elective cardiac surgery using the Bentall procedure resulted in a favorable outcome. Raising awareness of SOVA, a rare anomaly and an unusual cause of dyspnea, could mitigate the risk of rupture and prevent adverse outcomes.

Article information

**Conflict of interest:** None declared.

**Funding:** None.

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Figure 1. Images of SOVA. A. Transthoracic echocardiography (TTE), parasternal long axis (PLAX) view. B. TTE 4 chamber view with agitated saline opacification right heart chambers. C. Transesophageal echocardiography midesophageal short axis view (ME SAX). D. Computed tomography arrow depicting SOVA. E. Cardiac surgery: Big arrow depicting SOVA. Small arrow depicting RCA. F. Histopathology: Arrows depicting mucoid degeneration
Abbreviations: Ao, ascending aorta; LA, left atrium; LCC, left coronary cusp; LV, left ventricle; NCC, non-coronary cusp; RCA, right coronary artery; RCC, right coronary cusp; RV, right ventricle; SOVA, sinus of Valsalva aneurism; TA, truncus pulmonalis