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VASCULAR MEDICINE

IMAGING VIGNETTE: CLINICAL VIGNETTE

Right Coronary Sinus of Valsalva Aneurysm Rupture Precipitated by Nonsyndromic Aortopathy

Daniel Akrawi, MBBS,[a,b] Giuseppe Femia, MBBS, PhD,[b] Matle Fung, MBBS, PhD,[a,b] Kenneth Cho, MBBS, MPhil,[a,b] Ali Toufaili, MBBS[a,b]

ABSTRACT

We present a case highlighting the diagnostic challenges of identifying a ruptured right coronary sinus of Valsalva aneurysm in a patient with nonsyndromic aortopathy. Timely assessment with transthoracic and transesophageal echocardiography is vital for prompt diagnosis and successful treatment. Genetic panel testing should be offered to probands and first-degree family members. (J Am Coll Cardiol Case Rep 2024;29:102250) Crown Copyright © 2024 Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

A 34-year-old obese (body mass index, 36.5 kg/m^2) man presented overnight with sudden onset retrosternal chest pain precipitated by exertion. He was an active smoker with no other significant cardiovascular risk factors.

On examination, his blood pressure and heart rate were 114/59 mm Hg and 120 beats/min, respectively. Oxygen saturation was 95% on room air. There was a soft continuous murmur at the left sternal edge. He lacked phenotypic features of Marfan and Loeys-Dietz syndromes.

A 12-lead electrocardiogram demonstrated sinus tachycardia with no evidence of ST-segment abnormalities or T-wave inversion. His initial high-sensitivity troponin T (hsTropT) value was 790 ng/L (normal values ≤14 ng/L). A nongated computed tomography (CT) aortogram revealed no evidence of aortic root dilation or dissection (**Figure 1A**). Unfortunately, because of the presence of simultaneous contrast media in the right atrium and aortic root, coupled with motion artifact, a ruptured sinus of Valsalva aneurysm (SOVA) was not identified. Compare this with the **Figure 1B** schematic, which represents an unruptured right coronary SOVA into the right atrium.

Given the clinical picture and his raised hsTropT value, he received a provisional diagnosis of and treatment for acute coronary syndrome (ACS). A transthoracic echocardiogram (TTE) demonstrated hyperdynamic left ventricular function (Video 1) with a right coronary SOVA in the parasternal short-axis view (**Figure 1C**, Video 2). There was continuous turbulent color Doppler flow from the aortic root into the right atrium, consistent with a ruptured SOVA (Video 3). A transesophageal echocardiogram (TEE) confirmed these findings (**Figures 1D and 1E**, Videos 4 and 5). The aortic valve was tricuspid, with no evidence of infective endocarditis. There were no atrial or ventricular septal defects. Invasive coronary angiography revealed minor coronary artery disease.

From the [a]Department of Cardiology, Campbelltown Hospital, Sydney, New South Wales, Australia; and the [b]Western Sydney University, Sydney, New South Wales, Australia.

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ABBREVIATIONS AND ACRONYMS

ACS = acute coronary syndrome

CT = computed tomography

hsTropT = high-sensitivity troponin T

SOVA = sinus of Valsalva aneurysm

TEE = transesophageal echocardiogram

TTE = transthoracic echocardiogram

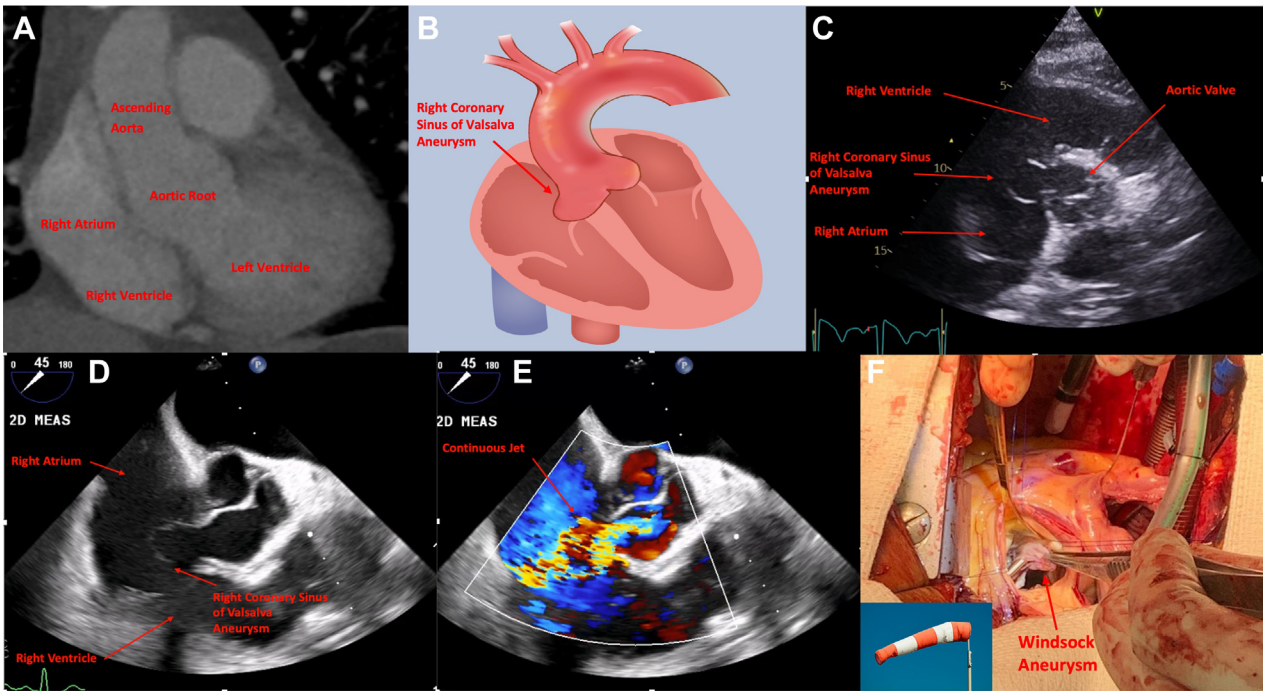
An urgent sternotomy confirmed a ruptured windsock aneurysm (Figure 1F) that was successfully repaired with a pericardial patch. Histologic analysis identified disorganized and fractured elastic fibers, with negative infective, autoimmune, and vasculitic screening results. The patient had an uncomplicated postoperative course and has returned to full-time employment. Subsequent familial screening revealed significant aortic sinus dilation in 2 first-degree relatives, consistent with an inherited nonsyndromic aortopathy.

This case highlights the challenges in diagnosing a SOVA rupture, particularly when patients present with clinical symptoms and investigations that mimic ACS.

Inherited aortopathies denote a group of genetic conditions related to impaired proliferation of vascular smooth muscle cells, destructive matrix remodeling with elastin fragmentation, and proteoglycan deposition.[1,2] These conditions can be classified into 2 broad groups: syndromic and nonsyndromic. Syndromic aortopathy refers to genetically mediated syndromes with systemic features, aortic dilation, and acute aortic events; examples include Marfan syndrome, Turner syndrome, Loeys-Dietz syndrome, and vascular Ehlers-Danlos syndrome. Nonsyndromic aortopathy refers to various genetic predispositions without systemic features.

A lack of syndromic features and an apparently normal CT aortogram do not preclude the diagnosis of an aortopathy or a SOVA rupture. Despite diagnostic challenges, prompt investigation with good-quality TTE and TEE is vital to timely diagnosis and successful treatment. Probands and family members should be offered genetic testing.[3] Positive genetic testing results should trigger gene-based management and cascade testing of first-degree family members.[3]

FIGURE 1 Multimodal Imaging



(A) Nongated computed tomography aortogram. Coronal plane with simultaneous contrast in the right atrium and aortic root revealing no evidence of aortic root dilation, aneurysm, or dissection. (B) Coronal plane representing an unruptured right coronary sinus of Valsalva aneurysm (SOVA) into the right atrium. (C) Transthoracic echocardiogram. Parasternal short-axis view at the aortic valve level revealing a right coronary sinus of Valsalva. (D) Transesophageal echocardiogram. Midesophageal aortic valve short-axis view at 45° revealing a right coronary sinus of Valsalva aneurysm. (E) Transesophageal echocardiogram. Midesophageal aortic valve short-axis view at 45° revealing a continuous color Doppler jet from the right coronary sinus of Valsalva into the right atrium. (F) Intraoperative finding of a ruptured windsock aneurysm. 2D MEAS = 2-dimensional measurement.

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ADDRESS FOR CORRESPONDENCE: Dr Daniel Akrawi, Department of Cardiology, Campbelltown Hospital, Therry Road, PO Box 149, Campbelltown, Sydney, New South Wales, Australia. E-mail: daniel.akrawi@health.nsw.gov.au.

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KEY WORDS aorta, computed tomography, echocardiography, vascular disease

APPENDIX For supplemental videos, please see the online version of this paper.