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Large-Vessel Vasculitis Presenting with Syncope and Complete Heart Block

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We present the case of a 55-year-old male with a pseudoaneurysm in the right coronary sinus of Valsalva. He was found to have a complete heart block in the context of large-vessel vasculitis involving the aortic root and basal interventricular septum. This case demonstrates the importance of generating a thorough differential diagnosis of complete heart block and using multimodality imaging to pursue the investigations for aortitis when a septal recess near the aortic root is detected. Considering to proceed to a surgical intervention earlier in the progression of the disease should also be part of the management.
Clinical case

A 55-year-old male with no significant past medical or family history presented to the emergency department following a syncopal episode without prodromal symptoms. He had no cardiac or constitutional symptoms. His physical exam, including orthostatic vitals, was normal. His blood pressure was 132/69 mmHg, temperature 36.6°C and heart rate (HR) 66 beats/min. Serial troponins were negative, but the d-dimer level was >4000 ng/L FEU (normal <500 FEU/L), and the C-reactive protein (CRP) was 78 mg/L (normal 0-10 mg/L). His electrocardiogram (ECG) showed sinus rhythm with a right bundle branch block (RBBB) which was new compared to his ECG 6 years ago.

A computed tomography pulmonary angiogram ruled out pulmonary embolism. A transthoracic echocardiogram (TTE) showed normal cardiac chambers, preserved left ventricular ejection fraction (LVEF), and a recess in the right coronary aortic root with extension into the basal ventricular septum (Figure-1A-D, Video-S1). There was thickening of the aortic root anterior wall without any intimal flap. A computed tomography angiogram (CTA) of the thoracic and abdominal aorta demonstrated no aortic dissection or intramural hematoma but revealed mild near-circumferential wall thickening throughout the aorta extending into its branches (Figure-2A). Shortly afterward, the patient developed a new complete heart block (CHB). The differential diagnoses for CHB with a pseudoaneurysm in the right coronary aortic root included sarcoidosis, aortitis and, endocarditis with a paravalvular abscess. A temporary screw-in pacemaker was inserted pending an infectious work up.
An F-fluorodeoxyglucose positron emission tomography (FDG-PET) revealed diffuse moderate uptake in the aortic root, arch, and ascending aorta, with mild uptake in the descending aorta, the brachiocephalic, subclavian, and common carotid arteries, indicating active large-vessel vasculitis (Figure-2B). A cardiac magnetic resonance imaging (CMR) showed a 16 x 13 mm pseudoaneurysm containing a small thrombus originating from the base of the right sinus of Valsalva and extending to the basal anteroseptal wall (Figure-2E). A later cardiac CTA showed the pseudoaneurysm extending into the basal anteroseptal myocardium with no thrombus (Figure-2C-D). The thrombosis service involved in the care of the patient decided against anticoagulation as the thrombus was no longer visualized on the repeat cardiac CTA.

An autoimmune work up, including antibody testing was negative. An infectious work up including blood cultures, serologies, and tuberculosis testing was negative, except for Q fever serology, which was positive. A left temporal artery biopsy showed no evidence of giant cell arteritis.

A permanent pacemaker was implanted. While awaiting the Q fever PCR results, the patient was started on doxycycline and hydroxychloroquine 200mg twice daily. For the suspected aortitis, he was started on prednisone 1mg/kg daily. The CRP had spontaneously improved to 33 and normalized after prednisone initiation. After the Q fever PCR result was negative, hydroxychloroquine was discontinued and tocilizumab was initiated for vasculitis treatment. Prednisone was tapered over six months. The CRP remained normal upon follow up, recognizing that this could have been confounded by tocilizumab use.
Yearly echocardiograms showed minor increase in the right coronary aortic root pseudoaneurysm, for which no change in the medical treatment was warranted. Since the initial hospitalization, the vascular surgery team monitored him with yearly CTAs of the thoracic and abdominal aorta which showed stability of the diffuse aortic wall thickening and arch branches.

Five years after the initial presentation, the patient presented to the emergency room for chest pain. The repeat TTE showed expansion of the pseudoaneurysm downwards into the basal aspects of the inferoseptal and inferior walls (Figure-1E-H, Video-S2). The troponins were 5 and CRP 11. The FDG-PET showed stable minimal arterial wall FDG, below diagnostic threshold for active vasculitis. Cardiac CTA showed a 90% stenosis of the proximal circumflex artery. No coronary angiogram was performed due to the presumed risks of perforation of the sinus of Valsalva. After a multidisciplinary discussion involving cardiac surgery, cardiology and rheumatology, the patient underwent a successful Bovine patch repair of the sinus of Valsalva (Figure-S1, Video-S3) and a coronary artery bypass graft to the circumflex artery.

Discussion

Large-vessel vasculitides are multi-system blood vessel disorders characterized by inflammation of primarily large vessels related to the aorta and its major branches. Because of its highly variable clinical presentation, the diagnosis is challenging and requires a high index of suspicion.(1) Our case had an unusual presentation with new onset CHB resulting in syncope. The aortic inflammation spread to the aortic root and the basal anteroseptal
wall, causing an anatomical impairment of the conduction system. The elevated CRP, the deep recess originating from the base of the right sinus of Valsalva and the wall thickening throughout the length of the aorta suggested either an infectious or inflammatory process. A similar presentation of aortitis was found in two other case reports, where the inflammation extended into the basal septum, causing a pseudoaneurysm and CHB.(2,3)

This case highlights the importance of TTE and multimodality imaging. As per the ACC/AHA/HRS Guideline, a comprehensive approach to syncope is essential. In this case, as the initial evaluation with the abnormal ECG could not rule out cardiovascular abnormalities, the TTE was a useful tool as it was the first investigation demonstrating structural abnormalities, leading to complementary imaging and the diagnosis of large-vessel vasculitis.(4)

The American College of Rheumatology guideline for large-vessel vasculitis recommends medical treatment as the first line over surgical intervention. When there is a decision to perform a surgery for example in the context of limb or organ ischemia, a delayed procedure until the disease is quiescent is recommended. (5) There are no guidelines about the optimal timing to proceed to a surgical intervention. However, this case prompts us to consider whether intervention should occur as soon as inflammation is under control to prevent further complications from the pseudoaneurysm, such as rupture or fistula formation. In our case, the pseudoaneurysm significantly increased in size despite controlled inflammation, leading us to suspect a major role played by the pressure within the pseudoaneurysm exerting force downward on the septum.
In conclusion, this case highlights the importance of considering large-vessel vasculitis in patients with syncope and new-onset CHB, especially with basal anteroseptal wall abnormalities on echocardiogram. An FDG-PET scan is crucial when a septal recess is detected after ruling out dissection and infection. Prompt immunosuppressive treatment can improve outcomes. Multidisciplinary follow-up with cardiology, rheumatology and vascular surgeons is needed to determine the optimal timing and nature of intervention.
References


Figure legends

Figure-1. (A-D) Echocardiogram at the initial presentation. (E-H) Echocardiogram five years later. A, E) Parasternal long-axis, B, F) parasternal short-axis, C, G) apical four-chamber, and D, H) apical three-chamber views of a TTE revealing the pseudoaneurysm in the right coronary aortic root extending into the septum (white arrows).

Figure-2. (A-D) CTA, FDG-PET and CMR. A) CTA aorta demonstrating near-circumferential mural thickening of the ascending and descending aorta (white arrows). B) FDG-PET: Uptake throughout the aortic root (black arrows), ascending aorta, aortic arch and its major branches, with milder uptake in the descending aorta. C) Cardiac CTA: short-axis view demonstrating the pseudoaneurysm extending into the basal anteroseptal segment (yellow arrow). D) Cardiac CTA: orthogonal long axis view of the pseudoaneurysm (yellow arrow). E) CMR: Three-chamber image from LGE sequence demonstrating the pseudoaneurysm arising from the inferior aspect of the right coronary cusp. A small thrombus is seen within the pseudoaneurysm (blue arrow).
Supplementary material

Supplemental Figure-S1. Surgery images. A) Baseline anatomy with visualization of the pseudoaneurysm of the sinus of Valsalva from the right coronary sinus (white arrow). B) Bovine patch. C) Bovine pericardial patch descended in the proper position (black arrow). D) Final result.

Video-S1. Echocardiogram at the initial presentation. Parasternal long-axis, parasternal short-axis, apical four-chamber, and apical three-chamber views of a TTE revealing the deep recess in the right coronary aortic root extending into the septum.

Video-S2. Echocardiogram before the surgery. Parasternal long-axis, parasternal short-axis, apical four-chamber, and apical three-chamber views of a TTE revealing the deep recess in the right coronary aortic root extending into the septum.

Video-S3. Echocardiogram after the surgery. Parasternal long-axis, parasternal short-axis, apical four-chamber, and apical three-chamber views of a TTE post repair of the right coronary sinus pseudoaneurysm.