Mechanisms of pseudo-quadricuspid aortic valve perforation with predominant eccentric aortic regurgitation

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Received 25 December 2021; first decision 8 February 2022; accepted 1 April 2022; online publish-ahead-of-print 6 April 2022

Figure 1 In the aortic short-axis view (A), the pseudo-cusp (asterisk) had a defect (arrow) which was determined as the ‘blood sack’ and its flow outlet (arrow) by simultaneous colour Doppler flow imaging. In the aortic long-axis view (B), the pseudo-cusp (asterisk) was recognized as a dissection of the non-coronary cusp belly in the setting of non-coronary cusp perforation (arrow). Biplane images (C) helped visualize the torrential eccentric aortic regurgitation going through the ‘blood sack’ (arrow). A 3D transesophageal echocardiography reconstruction of the aortic long-axis view (D) clearly shows both the inlet (arrow) and outlet (curved arrow) of the ‘blood sack’ (asterisk). A 3D en face view of the aortic valve (E) enables a visualization of the non-coronary cusp perforation, which resembles the exact anatomy in the surgical finding (F).

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Handling Editor: Romain Didier
Peer-reviewers: Giulia Elena Mandoli and Stefano Nistri
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A 57-year-old man with a known 2-year history of aortic insufficiency was referred for surgery for severe aortic regurgitation (AR) and LV enlargement (LVE). Previous transthoracic echocardiography (TTE) reported aortic valve perforation (AVP) or quadricuspid aortic valve. He was on warfarin, and repeated TTE at the first 3-month follow-up showed an LVEDD of 55 mm and an LV ejection fraction of 62%.

Isolated aortic perforation with cusp dissection is an extremely rare aetiology for severe eccentric AR. Given that the aortic valve has sandwich-like three-layered components, such as the fibrosa (aortic side), spongiosa (inner layer), and ventricularis (ventricular side), the dissection of the cusp may result from unbalanced sheer stress on the spongiosa. And the belly of the cusp is more vulnerable owing to its less spongiosa elastin than the hinge and coaptation regions. We presented a predominant anatomical ‘pocket’ that mimicked quadricuspid aortic valves in short-axis view but was finally ruled out by using a ‘region of interest’—derived scanning approach, i.e. by focusing the pseudo-cusp and rotating the probe on simultaneous biplane and CDFI modes. Our experience highlights that, in patients with severe eccentric AR and highly suspected AVP, multiplanar TEE, CDFI, and 3D TEE reconstruction should be applied sufficiently to assist the accurate diagnosis of the aortic anatomy and mechanisms of the regurgitation.

Lead author biography

Dr Zhenzhen Wang is an associate professor in Ultrasound Medicine at the Zhejiang Provincial People’s Hospital. She graduated with an MD in Imaging and Nuclear Medicine from Harbin Medical University. She was a visiting research fellow for Cardiovascular Medicine at the Mayo Clinic from 2018 to 2020. She is currently a specialist in intraoperative transesophageal echocardiography and 3D reconstruction. She is a member of the European Society of Cardiology, the American Heart Association, and the American Society of Echocardiography and is a Craniofacial Committee member of the China Society of Ultrasonic Medical Engineering.

Supplementary material

Supplementary material is available at European Heart Journal—Case Reports online.

Acknowledgements

We really appreciate the technical support from Dr Yi Lin (Radiology Department) and Dr Litao Sun (Ultrasound Department) of Zhejiang Provincial People’s Hospital.

Consent: The authors confirm that written informed consent for the publication of this case report was obtained from the patient in line with Committee on Publication Ethics (COPE) guidelines.

Conflict of interest: None declared.

Funding: None declared.

Data availability

The data underlying this article will be shared on reasonable request to the corresponding author.

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