

Aortic regurgitation due to aortic valve perforation and aortitis in Behçet's disease

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Received 13 February 2021; first decision 10 March 2021; accepted 12 May 2021

A 43-year-old man was hospitalized with paroxysmal nocturnal dyspnoea, when his vital parameters were stable. Physical examination was unremarkable except diastolic murmur. He had recurrent aphthous stomatitis and folliculitis-like eruptions on his lower extremities. The blood examinations were unremarkable, including serological tests. Several blood cultures were negative. Transthoracic echocardiography demonstrated diffuse hypokinesis of enlarged left ventricle with ejection fraction of 45% and severe aortic regurgitation (AR). Transoesophageal echocardiography (TOE) showed the sinus

of Valsalva aneurysms of non-coronary cusp (NCC) and a mobile tissue of NCC (Figure 1, Video 1). Doppler TOE showed prominent jet flow through NCC. The patient had no history of any diseases or drug use included immunosuppression. Aortic valve and root replacement (Bentall) operation was undertaken with bioprosthetic valves. The macroscopic specimens showed shortening of left coronary cusp, perforation of NCC and true Valsalva sinus aneurysms of left and non-coronary. Histopathology revealed fibrotic thickening of aorta and aortic valve, in which lymphocytic infiltration forming follicles and occasional neutrophils were observed in the adventitia and media. The lymphocytic infiltration was found in surroundings of perforation of NCC^{1,2} (Figure 2). Postoperatively, colchicine and steroid treatments were started for pericarditis and transient cardiac

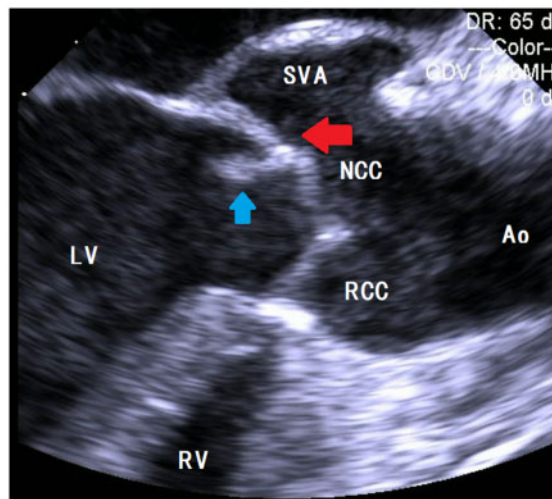


Figure 1 Transoesophageal echocardiography demonstrating the sinus of Valsalva aneurysms of non-coronary cusp, perforation of non-coronary cusp (red arrow), and a mobile tissue (blue arrow). Ao, aorta; LV, left ventricle; NCC, non-coronary cusp; RCC, right coronary cusp; RV, right ventricle; SVA, sinus of Valsalva aneurysms.

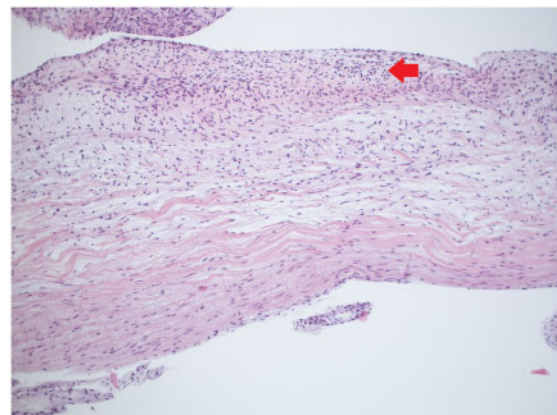


Figure 2 Histopathological specimen of non-coronary cusp presenting fibrotic thickening and lymphocytic infiltration (red arrow) in surrounding of perforation.

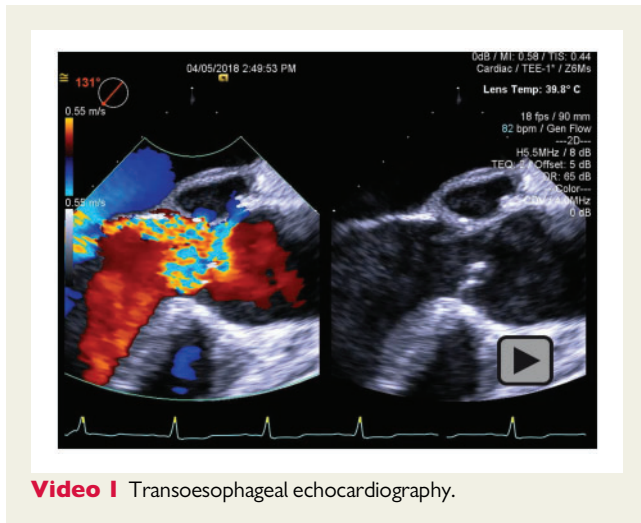
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Handling Editor: Aref Bin Abdulhak

Peer-reviewers: Rizwan Ahmed

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dysfunction. Inflammatory response rapidly subsided with improvements of pericardial effusion and cardiac function. Intriguingly, the patient followed an uneventful course without developing any complications. Collectively, the patient had oral, skin, and cardiovascular lesions, although other ocular, urogenital, and arthritic abnormalities were not identified. The diagnosis was suggested as Behçet's disease (BD) by revised international criteria for BD (2006).³ Although the mobile tissue looked like vegetation, the infectious

endocarditis could be deniable microbiologically, serologically, and pathologically. The case was non-infectious perforation, arising from valvulitis, contributed to AR. In conclusion, the case highlights the importance of perioperative diagnosis of BD for immunosuppressive treatment and the close follow-up.

Acknowledgements

We would like to greatly thank Mika Fujimoto for technical assistance of echocardiography, and Yoshihisa Morimoto MD and Yutaka Okita MD, who are cardiovascular surgeons, for the exquisite operation. We also thank Eisuke Enoki MD, who is pathologist, for the histopathological findings.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared.

Funding: None declared.

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