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Case Report

Tumor-like appearance in ruptured sinus of Valsalva with quadricuspid aortic valve

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ABSTRACT

Sinus of Valsalva rarely ruptures due to congenital causes. When it comes to ruptured sinus of Valsalva combined with quadricuspid aortic valve, no cases have so far been reported in Japan. Here, we describe the case of 32-year-old female who developed ruptured sinus of Valsalva with tumor-like appearance. Some of the cases of ruptured sinus of Valsalva show aorta to right atrial tunnel with a windsock aneurysm looking like a tumor by echocardiography.

<**Learning objective:** Sinus of Valsalva rarely ruptures due to congenital causes such as aortic valve anomalies such as quadricuspid aortic valve. Ruptured sinus of Valsalva sometimes shows tumor-like appearance by echocardiography. In such a case, we need to keep in mind that the sinus of Valsalva with aortic valve anomalies rarely ruptured showing a fistulous pouch-like "windsock" in the edge of aneurysm by echocardiography.>

Background

Sinus of Valsalva rarely ruptures due to congenital causes. The incidence of ruptured sinus of Valsalva has been reported to range between 0.1% and 3.5% of all congenital cardiac anomalies [1]. In addition, quadricuspid aortic valve (QAV) is a rare congenital cardiac anomaly, and has been reported in 0.043% and 1% in 13,805 two-dimensional transthoracic echocardiography and in 225 aortic valve surgeries, respectively [2,3]. Ruptured sinus of Valsalva combined with aortic valve anomalies such as QAV rarely occurs. Here, we report a case with ruptured site of sinus of Valsalva looking like a tumor, which was difficult to differentiate by echocardiographic finding alone.

Case report

A 32-year-old female with no medical history was admitted to our hospital due to progressive dyspnea on exertion and leg edema lasting for 6 months despite treatment with medication, following an abrupt onset of chest pain. On admission, blood pressure was 136/74 mmHg, pulse rate was 98/min, and body temperature was 36.5 °C. Cardiac murmur (systolic, grade IV/VI; diastolic, grade II/VI) was heard at the 2nd and 3rd intercostal spaces along the left sternal border. Other physical examination was normal except for three fingerbreadth palpable liver and lower leg edema. Routine laboratory tests showed an increase in B-type natriuretic peptide (160 pg/mL), total bilirubin (1.9 mg/dL), and aspartate aminotransferase (35 U/L). Blood cultures were negative. Electrocardiogram demonstrated normal sinus rhythm, and chest radiography showed a finding of cardiomegaly with a largely bulging right cardiac border. Transthoracic echocardiography (Fig. 1A–D) showed ruptured sinus of Valsalva with QAV, which made communication between coronary cusp and right atrium (RA) (presenting uninterrupted flow increasing up to a peak gradient 145 mmHg estimated by Doppler echocardiography) with something tumor-like in appearance, and which caused mild to moderate aortic regurgitation and moderate tricuspid regurgitation. The preoperative catheter examination showed the following results: pulmonary capillary wedge pressure 20 mmHg, pulmonary artery pressure 43/15/31 mmHg, right ventricular pressure 40/9 mmHg, right atrial pressure 21/19 mmHg, central venous

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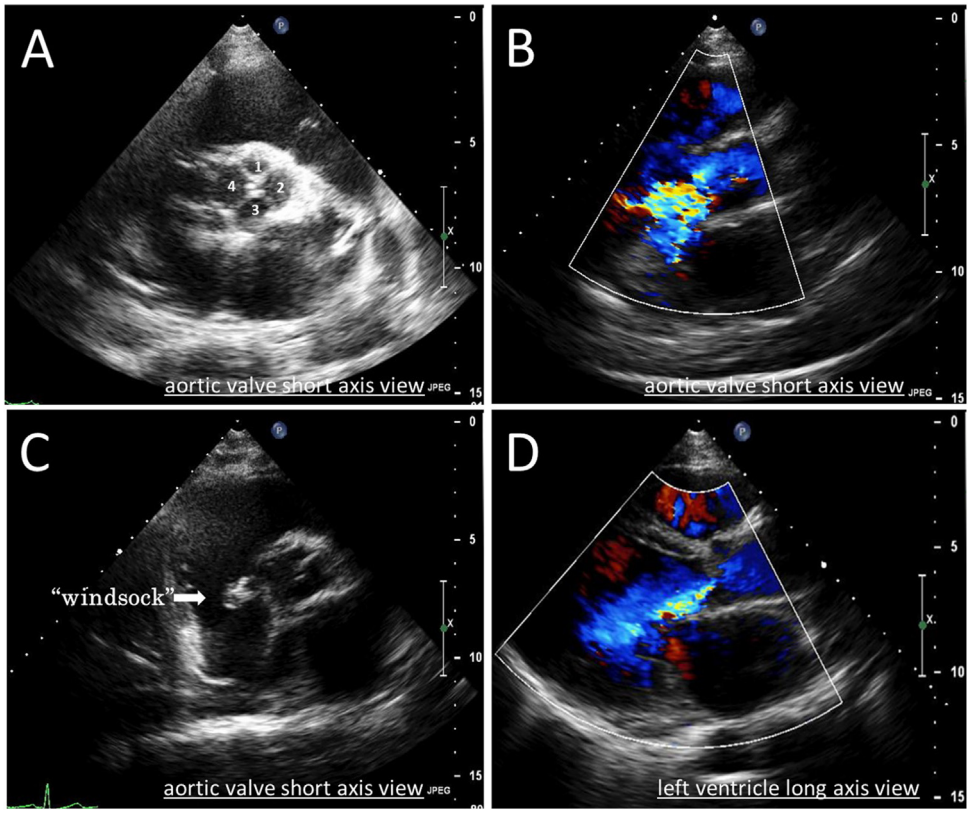

Fig. 1. Transthoracic echocardiography showed (A) quadricuspid aortic valve, (B) ruptured sinus of Valsalva communicating any coronary cusp and right atrium, (C) a "windsock" aneurysm located around tricuspid valve, and (D) mild to moderate aortic regurgitation without prolapse of aortic valve. 1: right-coronary cusp, 2: left-coronary cusp, 3: non-coronary cusp, 4: accessory cusp.

pressure 16 mmHg, pulmonary-to-systemic flow ratio 2.6. Coronary angiography showed no significant stenosis. Aortography (Fig. 2A) demonstrated QAV and shunt from sinus of Valsalva to RA, and Seller's grade I aortic valve regurgitation. Cardiac magnetic resonance imaging (MRI) was performed to identify the tumor-like finding and exclude other cardiac abnormalities including the aortic valve and sinus of Valsalva. However, cardiac MRI also showed QAV and shunt from sinus of Valsalva to RA, similar to that of aortic angiography (Fig. 2B) and showed no other abnormalities. At this point, although we could not conclude final diagnosis, surgery was indicated for the significant left to right shunt and symptomatic heart failure. Surgical inspection confirmed a fistulous pouch-like "windsock" in RA, and QAV with accessory cusp. The windsock originated in accessory cusp of aortic valve (Fig. 2C and D) and terminated in the RA just below the tricuspid valve, probably through congenital LV-RA shunt located around coronary aortic sinus. Accessory cusp was smaller than the other 3 cusps and positioned between the non-coronary cusp and original right-coronary cusp. After severing "windsock", ruptured sight of Valsalva and septal hole communicating Valsalva and RA were patched. Since the aortic valve tissue was fragile and non-coronary cusp ruptured and was deviated, bioprosthetic aortic valve replacement was performed with a 23-mm Carpentier-Edwards PERIMOUNT MAGNA EASE 23 mm (Edwards Lifesciences, Irvine, CA, USA) considering her desire to be able to become pregnant. In addition, since there was a deformation between the anterior and the septal cusps of the tricuspid valve, edge to edge of the cusps were sutured. Pathological examination revealed sparse connective tissue in the "windsock" without infective findings. The patient recovered well and was discharged on 19th postoperative day without any complications.

Discussion

This is a rare case of ruptured sinus of Valsalva with QAV that formed aorta–right atrial tunnel with a "windsock" aneurysm. To the best of our knowledge, this is the first report of ruptured sinus of Valsalva with QAV in a Japanese patient.

Sinus of Valsalva rarely ruptures due to congenital or acquired causes. Congenital causes are vulnerability or deficiency of the elastic lamina as shown in this case [4]. Acquired causes include infection, atherosclerosis, and deceleration trauma [5]. The right sinus of Valsalva is most commonly involved (76.8%) [1], and usually ruptures into the right heart chambers through congenital or acquired septal shunt [6,7]. Early and aggressive treatment of sinus of Valsalva aneurysms or fistulas is controversial, however, surgical treatment was performed due to refractory heart failure. It was reported that QAV and its associated anomalies are mostly diagnosed by echocardiography (51%), during surgery (22.6%), autopsy (15.6%), and aortography (6.5%) [8]. Since some of the ruptured sinus of Valsalva showed tumor-like appearance by echocardiography, it seems to be difficult to determine whether tumor-like appearance was vegetation, tumor, or "windsock" with the ruptured hole in the edge of the aneurysm [9,10]. In such a case, we need to keep in mind that the sinus of Valsalva with aortic valve anomalies rarely ruptured showing "windsock" aneurysm by echocardiography.

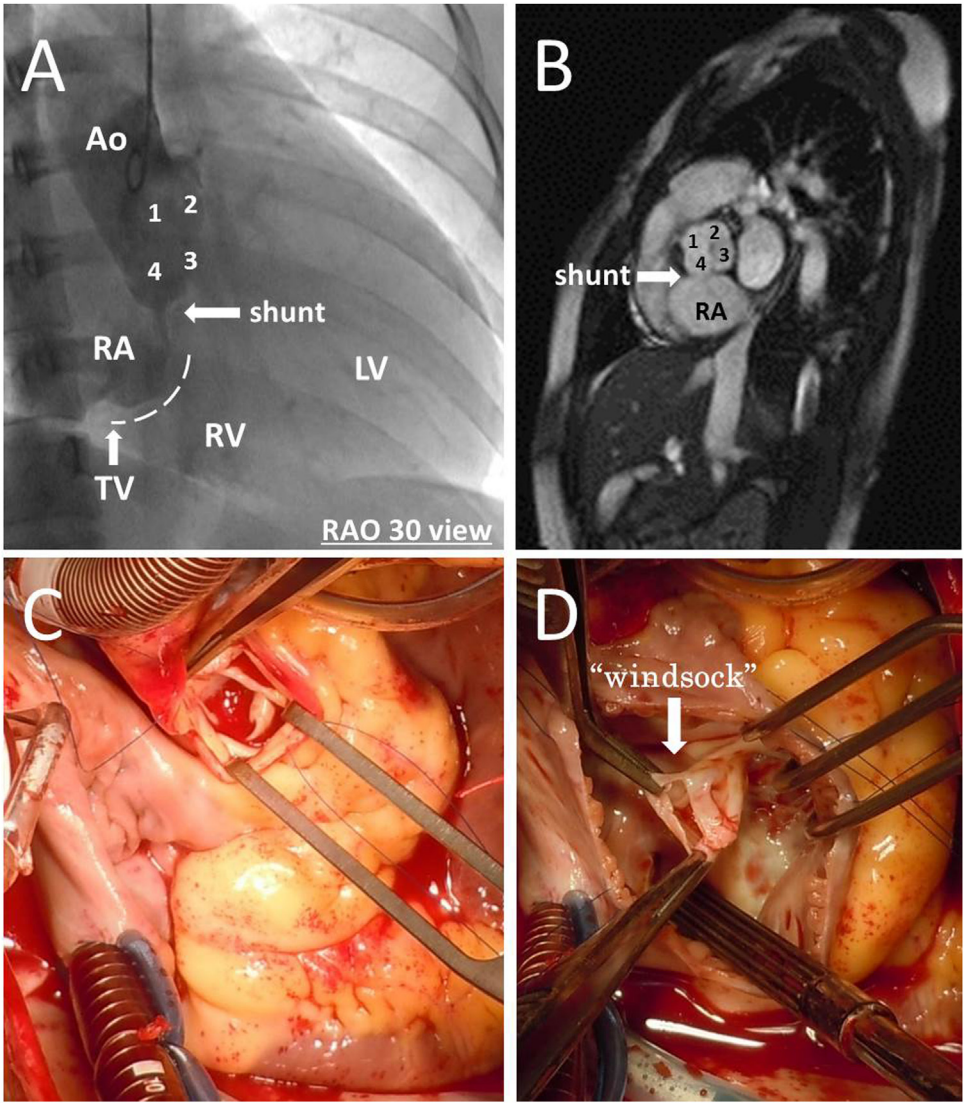

Fig. 2. Aortic angiography and cardiac magnetic resonance imaging demonstrated quadricuspid aortic valve and shunt from sinus of Valsalva to RA (A) (B). Note the perforating shunt of sinus of Valsalva (arrow). Surgical inspection showed (C) the ruptured sinus of Valsalva and (D) the ruptured fistulous pouch resembling a "windsock" tumor just around the tricuspid valve. 1: right-coronary cusp, 2: left-coronary cusp, 3: non-coronary cusp, 4: accessory cusp. RA, right atrium; RV, right ventricle; LV, left ventricle; Ao, aorta; TV, tricuspid valve.

Disclosures

None.

Appendix A. Supplementary data

Supplementary material related to this article can be found, in the online version, at doi:https://doi.org/10.1016/j.jccase.2019.06.009.

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