Journal of Cardiology Cases 29 (2024) 50–54

Contents lists available at ScienceDirect

Journal of Cardiology Cases

journal homepage: www.elsevier.com/locate/jccase

Case Report

Left sinus of Valsalva aneurysm causing acute myocardial infarction with compression of the left main coronary trunk

Genryu Mizokami (MD) [a], Masatsugu Nozoe (MD, PhD) [a], Seiya Kato (MD, PhD) [b], Daisuke Nagatomo (MD) [a], Nobuhiro Suematsu (MD, PhD) [a], Yuta Sukehiro (MD) [c], Noritoshi Minematsu (MD) [c], Satoko Sakata (MD, PhD) [d], Toru Kubota (MD, PhD) [a,*]

[a] Division of Cardiology, Cardiovascular and Aortic Center, Saiseikai Fukuoka General Hospital, Fukuoka, Japan
[b] Division of Pathology, Saiseikai Fukuoka General Hospital, Fukuoka, Japan
[c] Division of Cardiovascular Surgery, Cardiovascular and Aortic Center, Saiseikai Fukuoka General Hospital, Fukuoka, Japan
[d] Department of Epidemiology and Public Health, Graduate School of Medical Sciences, Kyushu University, Fukuoka, Japan

ARTICLE INFO

Article history:
Received 25 May 2023
Received in revised form 12 September 2023
Accepted 26 September 2023

Keywords:
Sinus of Valsalva aneurysm
Myocardial infarction
Coronary artery bypass grafting
Autopsy

ABSTRACT

Sinus of Valsalva aneurysm (SVA) is a rare cardiac anomaly that commonly originates from the right or noncoronary sinuses and rarely from the left sinus. SVA is usually diagnosed in the setting of clinical sequelae of a rupture. We herein report a case of an unruptured left SVA presenting as acute myocardial infarction. A 54-year-old woman with a history of radical operation for patent ductus arteriosus in childhood was transferred to our hospital. An electrocardiogram indicated extensive myocardial ischemia with ST elevation. Urgent coronary angiography was performed but was unable to identify the left coronary artery. Subsequent aortography and computed tomography revealed a large SVA originating from the left sinus and compressing the left coronary artery. The patient died after approximately one month of intensive care, including mechanical circulatory support and coronary artery bypass grafting. Autopsy confirmed that the left main coronary trunk was stretched and compressed by the SVA and revealed unexpected atherosclerosis in the left anterior descending artery. Although a left SVA is an extremely rare anomaly, it occasionally provokes fatal myocardial infarction. Since an SVA might hinder performing percutaneous coronary intervention, cardiac surgery should be considered when myocardial ischemia is recognized.

Learning objective: We herein report a case of an unruptured left sinus of Valsalva aneurysm (SVA) with acute myocardial infarction. Urgent percutaneous coronary intervention (PCI) was unsuccessful, as the left coronary artery was compressed by the SVA. The patient died after intensive care, including coronary artery bypass grafting (CABG). SVA, especially from the left sinus, is extremely rare but occasionally provokes myocardial infarction by compressing the coronary arteries. Because SVA might hinder PCI, CABG should be considered when myocardial ischemia is recognized.

Introduction

Sinus of Valsalva aneurysm (SVA) is a rare cardiac anomaly that occurs in <0.1 % of the general population [1]. SVA commonly originates from the right sinus (65 %–85 %), less commonly from the noncoronary sinus (10 %–30 %), and rarely from the left sinus (<5 %). A frequent complication of SVA is rupture into an adjacent chamber that occurs mostly between 20 and 40 years old [2]. Unruptured SVA is typically asymptomatic. However, a large unruptured left SVA occasionally causes myocardial ischemia by compressing the left coronary artery (LCA) [3].

We herein report an autopsy case of a middle-aged woman with a left SVA presenting with extensive anterolateral acute myocardial infarction (AMI). The autopsy confirmed that the left main coronary trunk (LMT) was compressed by the SVA and revealed unexpected progression of atherosclerosis in the proximal portion of the left anterior descending artery.

Case report

A 54-year-old woman was transferred to our emergency department with sudden onset of chest discomfort. She had a history of radical

* Corresponding author at: Division of Cardiology, Cardiovascular and Aortic Center, Saiseikai Fukuoka General Hospital, 1-3-46 Tenjin, Chuo-ku, Fukuoka 810-0001, Japan.
E-mail address: kubota-t@saiseikai-hp.chuo.fukuoka.jp (T. Kubota).

https://doi.org/10.1016/j.jccase.2023.10.003

operation for patent ductus arteriosus (PDA) in childhood. She had undergone annual medical checkups in the past five years, and no major health problems had been noted. The time from chest discomfort to hospital arrival was three hours.

On admission, her blood pressure was 73/50 mmHg, and her heart rate was 122 bpm without irregularity. Her oxygen saturation was 95 % under 5 L/min of oxygen with a reservoir mask. A 12-lead electrocardiogram (ECG) showed sinus tachycardia with left-axis deviation (left anterior fascicular block); wide QRS morphology (right bundle branch block); ST elevation in I, aVR, aVL, and V3–6; and ST depression in II, III, and aVF, suggesting AMI with LMT occlusion (Fig. 1A). A chest X-ray revealed marked pulmonary edema (Fig. 2A). A laboratory test indicated positive troponin T (TnT, 0.159 ng/mL >0.014) and elevated creatine phosphokinase-MB isozyme (CK-MB, 36.7 U/L > 20).

Urgent coronary angiography (CAG) was performed with the support of intra-aortic balloon pumping (IABP). CAG showed a normal right coronary artery (RCA). However, the LCA was not visualized. Subsequent aortography revealed a giant left SVA with a maximal diameter of 5 cm (Fig. 2B). Mechanical circulatory support with veno-arterial extracorporeal membrane oxygenation (V-A ECMO) was initiated in conjunction with IABP to maintain systemic circulation, resulting in temporary resolution of ECG abnormalities, including narrowing of the wide QRS morphology and a reduction in the ST segment elevation compared with the initial ECG (Fig. 1B). Revascularization with percutaneous coronary intervention (PCI) was abandoned because the orifice of the LCA failed to be identified, and the LCA was suspected of being partially reperfused based on the ECG changes. The door-to-suspected reperfusion time was two hours.

Multidetector row computed tomography angiography performed after CAG revealed that the LCA originated from the top of the giant left SVA and that the LMT was stretched and compressed by the aneurysm (Fig. 2C and D). The peak values of creatine kinase (CK), CK-MB, and troponin T were 9047 U/L, 968 U/L, and 25.72 ng/mL, respectively, which were compatible with massive AMI.

On the third hospital day, she developed complete atrioventricular block, resulting in cardiogenic shock despite continued mechanical circulatory support with V-A ECMO and IABP. Echocardiography showed no signs of mechanical complications, including ventricular septal perforation and/or cardiac tamponade. Urgent CAG was performed, and the LCA was considered to have become reoccluded although we could not evaluate the LCA at all due to the technical and anatomical limitations. The RCA was patent. Therefore, on-pump beating coronary artery bypass grafting (CABG) was subsequently performed with the left internal thoracic artery to left anterior descending artery and saphenous vein grafts to the diagonal branch and left circumflex artery. Cardiopulmonary bypass was established by cannulation of the ascending

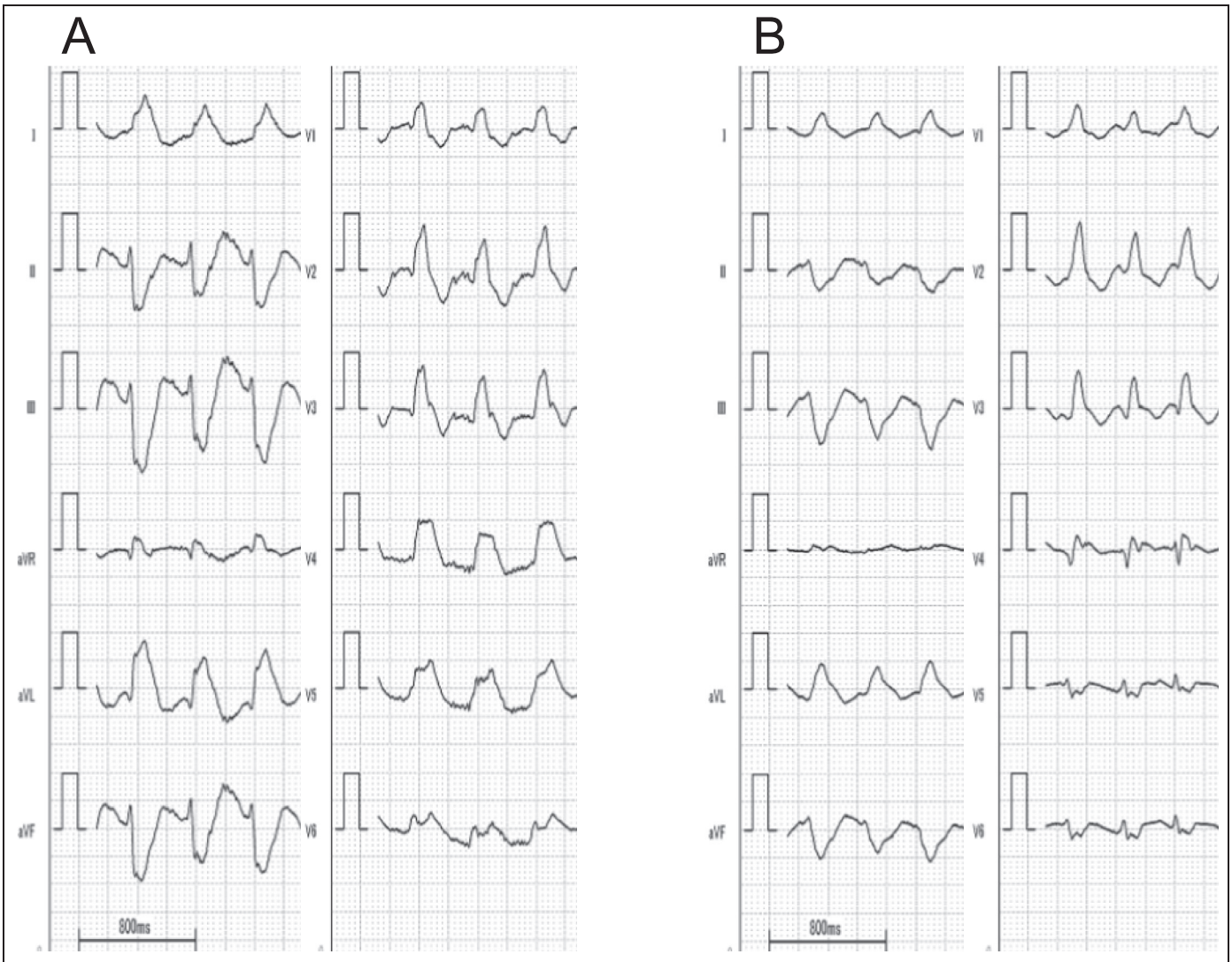

Fig. 1. Twelve-lead electrocardiograms at the beginning (A) and end (B) of cardiac catheterization.

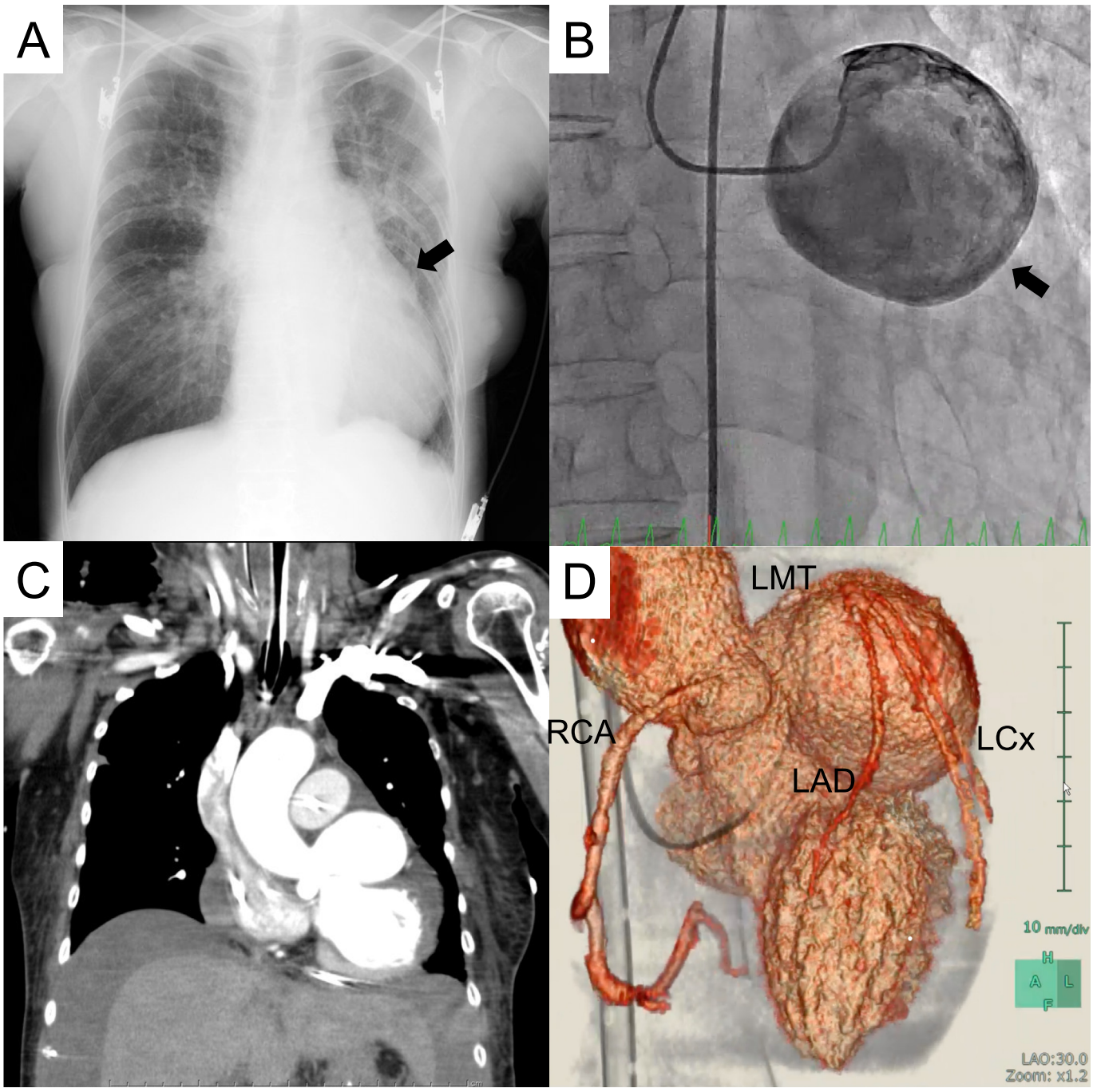

Fig. 2. Radiographical images of the left sinus of Valsalva aneurysm (SVA). (A) Chest X-ray showing marked pulmonary edema with a slight bulge along the left border of the heart (arrow). (B) Aortography showing the giant left SVA (arrow). (C) Computed tomography of the chest (coronal view). (D) Multidetector-row computed tomography angiography of the heart in three-dimensional reconstruction. The left coronary artery originated from the top of the giant left SVA. The left main coronary trunk (LMT) was stretched and compressed by the SVA. The flow of the right coronary artery (RCA) was preserved.
LAD, left anterior descending artery; LCx, left circumflex artery.

aorta and right atrium. Radical operation of the SVA was not performed due to her unstable general condition.

On the fifth hospital day, IABP was converted to Impella CP (Abiomed, Danvers, MA, USA) to ameliorate pulmonary hypertension by unloading the left ventricle. On the eighth hospital day, the Impella CP was removed due to worsening aortic regurgitation, and peripheral V-A ECMO was converted to central V-A ECMO to secure the antegrade arterial flow. However, despite intensive care, the wall motion of the left ventricle did not recover, and mechanical circulatory support with central V-A ECMO was continued. On the 23rd hospital day, she developed hemorrhagic cerebral infarction in her right cerebral hemisphere, and she died on the 26th hospital day.

An autopsy was performed after obtaining informed consent of her family. Macropathology revealed a large unruptured left SVA with extensive hemorrhagic infarction at the antero-lateral myocardium (Fig. 3A). The LMT running along the aneurysm was stretched, but no stenosis or plaque rupture was evident in its lumen (Fig. 3B). Histology showed fibrous thickening of the intima and highly degenerated medial

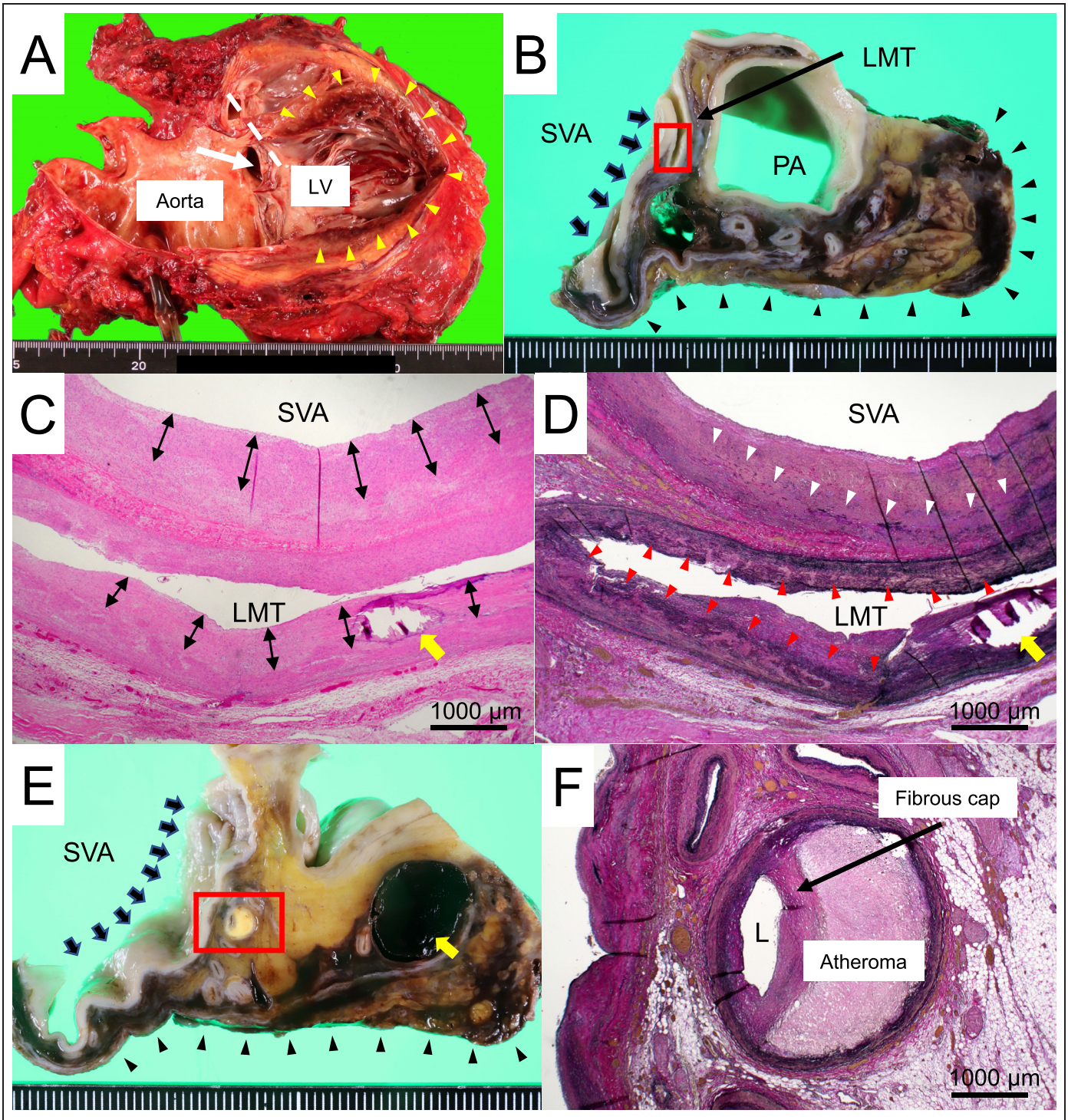

Fig. 3. Autopsy findings of the heart. (A) Gross finding of the prefixed heart. The heart weighed 625 g. The orifice of the sinus of Valsalva aneurysm (SVA) was observed on the cut surface of the long axis (white arrow), with extensive hemorrhagic infarction (yellow triangles) of the left ventricle (LV). (B) Cut surface indicated by a dotted line in (A) (post-fixation). The stretched left main coronary trunk (LMT) ran along with the SVA wall (black arrows) without luminal stenosis. The main trunk of the pulmonary artery (PA) was also observed in this slice. Pericardial side was indicated by black triangles. (C) Histology of the SVA wall and the LMT by hematoxylin and eosin staining. The area was indicated by the square in (B). Approximate zones of fibrous thickening of the intima in both SVA and LMT are indicated by double arrows. (D) Histology by Elastica van Gieson staining. The area was almost the same as in (C). Stretched and damaged SVA and LMT walls were observed with disrupted elastic fibers (white triangles in the SVA wall and red triangles in the LMT side). Small calcification was seen in the LMT wall (yellow arrow), but the luminal surface was smooth without any thrombosis. (E) The cut surface just on the foot side of (B) (postfixation). Progression of atherosclerosis was seen at the proximal portion of the left anterior descending artery (LAD) (square). The SVA wall (black arrows) did not directly compress the LAD. Subpericardial hematoma (yellow arrow) after coronary artery bypass grafting was also observed. Pericardial side was indicated by black triangles. (F) Histology of LAD by Elastica van Gieson staining. The area was indicated by the square in (E). The lumen (L) of the LAD was constricted by a large atheroma, which was covered by a thickened fibrous cap.

tissue in the aneurysmal wall and LMT in association with disrupted elastic fibers (Fig. 3C and D). Unexpectedly, severe stenosis with a large atheromatous plaque was observed in the proximal portion of the left anterior descending artery (Fig. 3E). In contrast, few atherosclerotic changes were observed in the other parts of coronary arteries or in the systemic arteries.

Discussion

We experienced a case of AMI accompanied by a large unruptured left SVA. The initial ECG findings, including left anterior fascicular block, right bundle branch block, and ST elevation in I, aVR, aVL, and V3–6 and depression in II, III, and aVF, suggested that AMI was provoked by occlusion of the LMT [4]. The absence of plaque rupture or stenosis in the LMT observed by an autopsy and temporal resolution of ST elevation and depression by initiation of mechanical circulatory support suggested that the LMT had become occluded by mechanical compression due to enlargement of the left SVA.

SVA is a rare cardiac anomaly that may be acquired or congenital, most commonly involving the right or noncoronary sinuses [1]. SVA originating from the left coronary sinus, as in the present case, is extremely rare. Congenital aneurysms are more common than acquired ones and may be associated with other heart anomalies, such as ventricular septal defects (30 %–60 %), bicuspid aortic valves (10 %), aortic insufficiency, pulmonary stenosis, coarctation of the aorta, atrial septal defects, and subvalvular aneurysms [1]. However, an association with PDA, as in the present case, is uncommon. Acquired aneurysms are caused by conditions affecting the aortic wall, such as infections (syphilis, bacterial endocarditis, or tuberculosis), trauma, or connective tissue disorders [1]. However, no specific findings suggestive of such disorders were noted in the present case. Therefore, this case appeared to be congenital, although the details of the PDA operation were undetermined.

SVA is usually diagnosed in the setting of clinical sequelae of a rupture that occurs mostly between 20 and 40 years old [2]. Although unruptured SVAs are typically asymptomatic, a large unruptured left SVA occasionally causes myocardial ischemia by compressing the LCA [3]. Lijoi et al. summarized 19 patients with unruptured left SVA who presented with angina or myocardial infarction [8 men and 11 women 51 ± 16 (SD) years old] [5]. The aneurysm was congenital in 10 patients and acquired in 9. The diagnosis was made by angiography in 14 patients, an autopsy in 4, and echocardiography in 1. Only 9 of these 19 patients underwent surgery, with successful surgical repair achieved in 6, while 3 died postoperatively. Associated aortic valve replacement was necessary in six patients, and CABG was performed in five patients. The present case was a 54-year-old woman who presented with myocardial infarction and underwent CABG on the 3rd hospital day but died of cardiogenic shock and hemorrhagic cerebral infarction on the 26th hospital day. Considering the high mortality rate (33 %) of surgical repair in these patients, whether she might have survived if she had undergone CABG on the first hospital day is unclear. However, considering the difficulty of performing PCI in these patients with SVA, we recommend that early revascularization with CABG should thus be considered when myocardial ischemia is recognized.

In the present case, the autopsy revealed the progression of atherosclerosis in the proximal portion of the left anterior descending artery. She had no coronary risk factors in at least the past five years. It has been reported that hemodynamic stress is deeply involved in the pathogenesis of intracoronary atheromatous plaques [6]. Therefore, we suspect that repeated high or low shear stress due to compression by the left SVA in the proximal portion of the left anterior descending artery might have exacerbated endothelial injury and advanced the atherosclerosis in the present case. Because patients with an unruptured left SVA who present with either angina or myocardial infarction are relatively older than those with a ruptured right or noncoronary SVA [1–3,5], the possible coexistence of atherosclerotic lesions in the LCA should therefore be investigated before performing CABG in these patients.

Conclusion

A left SVA is rare but provokes fatal AMI by compressing the LMT. Because PCI with SVA is too demanding to achieve complete revascularization, urgent CABG should be considered when myocardial ischemia is recognized.

Funding

None.

Consent statement

Consent was obtained from the bereaved family for the purpose of anonymized publication.

Declaration of competing interest

None.

Acknowledgments

Part of this work was orally presented at the 27th Annual Meeting of the Japanese Society of Vascular Pathology (Gifu, Japan, 2022) and at the 133rd Kyushu Regional Meeting of the Japanese Circulation Society (Kurume, Japan, 2022).

References

[1] Feldman DN, Roman MJ. Aneurysms of the sinuses of Valsalva. Cardiology 2006;106:73–81.
[2] Luo X, Zhang D, Li B, Qi L, Gong L, Tang Y, et al. Surgical repair of a ruptured congenital sinus of Valsalva aneurysm: 10-year experience with 286 cases. Eur J Cardiothorac Surg 2019;55:1211–8.
[3] Hiyamuta K, Ohtsuki T, Shimamatsu M, Ohkita Y, Terasawa M, Bekki H, et al. Aneurysm of the left aortic sinus causing acute myocardial infarction. Circulation 1983;67:1151–4.
[4] Kurisu S, Inoue I, Kawagoe T, Ishihara M, Shimatani Y, Nakamura S, et al. Electrocardiographic features in patients with acute myocardial infarction associated with left main coronary artery occlusion. Heart 2004;90:1059–60.
[5] Lijoi A, Parodi E, Passerone GC, Scarano F, Caruso D, Iannetti MV. Unruptured aneurysm of the left sinus of Valsalva causing coronary insufficiency: case report and review of the literature. Tex Heart Inst J 2002;29:40–4.
[6] Feldman CL, Stone PH. Intravascular hemodynamic factors responsible for progression of coronary atherosclerosis and development of vulnerable plaque. Curr Opin Cardiol 2000;15:430–40.