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CASE REPORT

ADVANCED

CLINICAL CASE

Cardiac Strangulation Due to Partial Pericardial Defect Presenting as Acute Myocardial Infarction

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ABSTRACT

A 79-year-old man with chest pain and dyspnea underwent emergency percutaneous coronary intervention for acute myocardial infarction. However, he died 17 days later due to refractory heart failure. An autopsy revealed cardiac strangulation caused by herniation of the apical heart through a pericardial defect due to partial absence of the pericardium. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2021;3:1635-1638) © 2021 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 79-year-old man presented to his primary care physician with sudden onset of resting chest pain and dyspnea that progressively worsened. He was diagnosed with acute anterior ST-elevation myocardial infarction. He was referred to our hospital, and admitted to the emergency department.

MEDICAL HISTORY

The patient was an otherwise healthy adult with no history of medication use.

LEARNING OBJECTIVES

- To review the subject of partial pericardial defect.
- To consider partial pericardial defect as a rare cause of acute myocardial infarction with multivessel occlusion.

DIFFERENTIAL DIAGNOSIS

Initially, differential diagnoses included acute myocardial infarction, acute heart failure, acute aortic dissection, and acute pulmonary thromboembolism.

INVESTIGATIONS

On admission, the patient had a pulse rate of 105 beats/min, blood pressure of 109/86 mm Hg, and reduced arterial oxygen saturation of 83% on 9 L/min of oxygen. The jugular veins were distended. Coarse crackles were heard in both lung fields. The patient's skin was moist, and his peripheral extremities were cold. His cardiac enzymes were elevated. Electrocardiography showed sinus tachycardia with a heart rate of 105 beats/min, right-axis deviation; Q-waves in leads I, aVL, V_5, and V_6; ST-segment elevation in leads I, V_5, and V_6; and ST-segment depression in leads III, V_1, and V_2. Chest radiographs showed cardiomegaly and

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ABBREVIATIONS AND ACRONYMS

CK = creatine kinase

IABP = intra-aortic balloon pumping

LAD = left anterior descending artery

pulmonary congestion (**Figure 1**). Echocardiography was performed, but the area from the apex to the lateral wall was not sufficiently visible (Videos 1A and 1B). Therefore, we diagnosed the patient with acute anterior ST-segment myocardial infarction with biventricular heart failure. The patient was intubated, ventilated, and started on intra-aortic balloon pumping (IABP) counterpulsation. Coronary angiography showed severe stenosis in the mid-left anterior descending artery (LAD), and abrupt cut-offs in the distal LAD, mid-diagonal branches, and the left circumflex artery, suggesting multiple obstructions caused by coronary spasm, thrombus, or dissection (Videos 2A and 2B). The right coronary artery originated from the left coronary cusp. However, there was no significant stenosis.

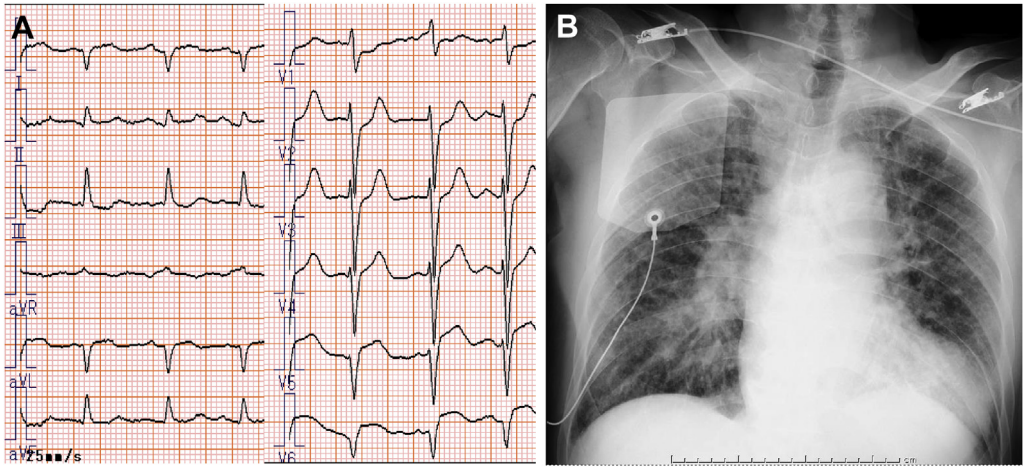
MANAGEMENT

Percutaneous coronary intervention was started approximately 8.5 hours after symptom onset. A guidewire was passed down the LAD, but could not be passed through the distal occlusion. Intravascular ultrasound showed an atherosclerotic plaque with mild calcification at the site of the mid-LAD stenosis and a drug-eluting stent was implanted. The remaining lesions were treated conservatively using anticoagulation and IABP management. Medical management was continued in the intensive care unit, with a peak creatine kinase (CK) level of 2,691 IU/L (normal values 41-153 IU/L). The next day, with trace amounts of dobutamine and noradrenaline support, the IABP was withdrawn. However, congestive heart failure was refractory to treatment. The patient died after 17 days. An autopsy revealed cardiac strangulation caused by herniation of the apical heart through a pericardial defect due to partial absence of the pericardium (**Figures 2A, 2B, and 3A**).

DISCUSSION

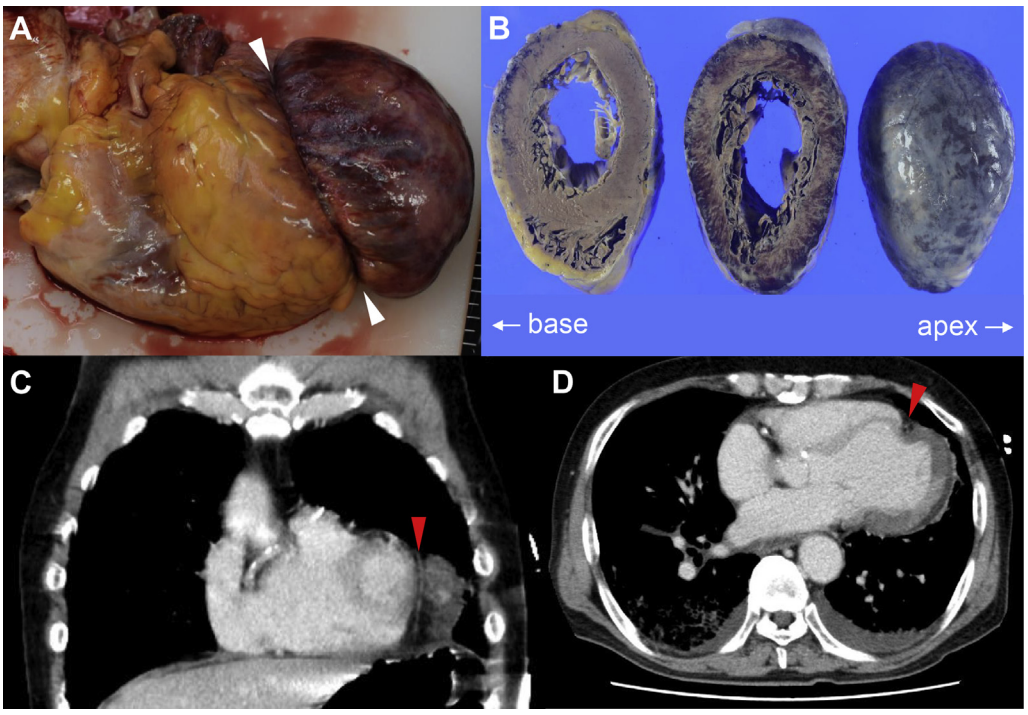
Pericardial defect is rare with a reported incidence of 0.007%-0.015% in autopsy cases (1,2). In addition, among the 34,000 patients who underwent cardiovascular surgery at the Mayo Clinic, 15 cases of a partial or complete absence of the pericardium were observed, none of which were diagnosed preoperatively (3). The Johns Hopkins Hospital reported that 24% of pericardial defect cases are partial defects (4). Pericardial defects are often asymptomatic. However, partial pericardial defects have been reported to cause chest pain, dyspnea on exertion, palpitations, and fainting; also, they have been reported to cause clinical signs such as heart murmurs, cardiomegaly, heart displacement on the chest x-ray, arrhythmias, and pneumopericardium associated with the development of spontaneous pneumothorax. Partial pericardial defect might lead to fatal complications such as sudden death or cardiac herniation through the defect. For this reason, surgical intervention is

FIGURE 1 Electrocardiogram and Chest Radiograph



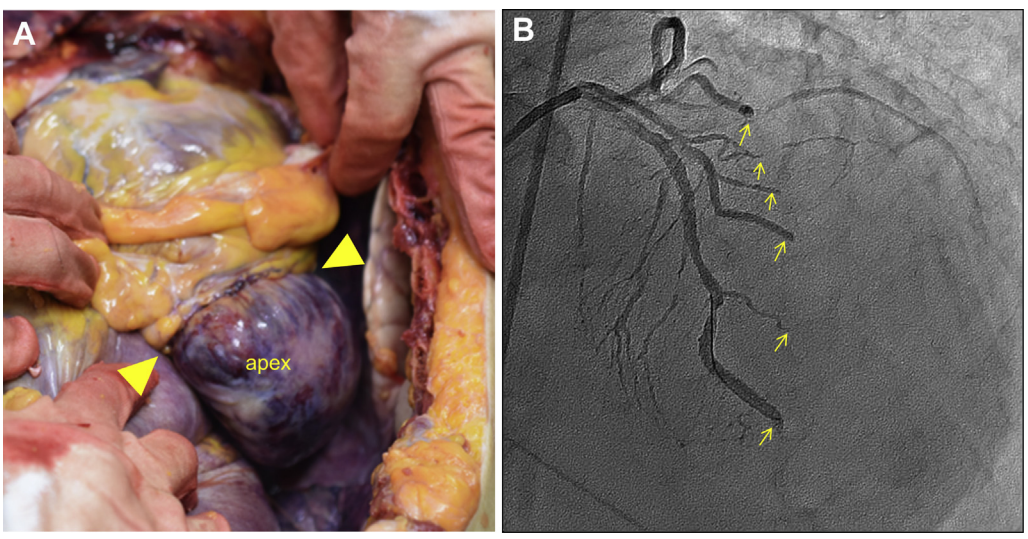
(A) Electrocardiogram showing sinus tachycardia, right-axis deviation, Q-waves in leads I, aVL, V_5, and V_6, ST-segment elevation in leads I, V_5, and V_6 induction, and ST-segment depression in leads III, V_1, and V_2. **(B)** Chest radiograph shows cardiomegaly and pulmonary congestion.

FIGURE 2 **Autopsy and Computed Tomography**



(A) Autopsy; anterior surface of the heart. The **white arrowheads** indicate the rim of the remained pericardium. **(B)** Autopsy; cross-section of the heart looking up from the apex. The section in the middle is the strangulated area. The third image shows the apex surface. **(C)** Computed tomography; coronal plane. **(D)** Computed tomography; transverse plane. The **red arrowheads** indicate a depression that may have been caused by strangulation.

FIGURE 3 **Autopsy and Coronary Angiography**



(A) Autopsy; the chest is opened through a midline sternal incision and the heart is shown. The apex side of the heart is protruding through the pericardial defect. The **yellow arrowheads** indicate the rim of the pericardial defect. **(B)** Coronary angiography; the left coronary artery (cranial view). The left anterior descending artery, diagonal branches, and circumflex artery are occluded due to external compression.

recommended for symptomatic patients with partial defects and for those with cardiac herniation (5-7).

In this case, postmortem pathology revealed that the heart had protruded through a partial pericardial defect and become strangulated. Chest radiographs showed cardiomegaly. However, there were no significant Snoopy sign or other findings suggestive of pericardial defects. On echocardiography, the area from the apex to the lateral wall was not sufficiently visible. Based on autopsy findings and computed tomography images taken during the course of disease (**Figure 2**), presence of a posterolateral shift of the cardiac apex away from the chest wall was considered. Coronary angiography showed interruption of blood flow in the midportion of the left circumflex, the first and second diagonal branches, and the distal LAD. Shower embolization, multi-vessel spasm, or epicardial adhesion was considered as a differential diagnosis, but the underlying cause was the external compression of the coronary arteries by the rim of the remaining pericardium (**Figure 3B**).

The peak CK level was 2,691 IU/L. Thereafter, CK remained at 500-1,000 IU/L. Autopsy showed myocardial necrosis in all layers of the strangulated area, with significant myocardial damage.

The diagnosis of congenital heart disease was challenging because the patient was elderly. Enlargement of the heart may have progressed gradually with age, and, when intracardiac pressure spontaneously increased, it caused the left ventricle to protrude through the pericardial defect.

CONCLUSIONS

We encountered a patient with a partial pericardial defect that presented as acute myocardial infarction from external compression of the left coronary artery. Although rare, partial pericardial defects should be included in the differential diagnosis as a cause of acute myocardial infarction. Surgical treatment should be considered if the patient is symptomatic and at risk of strangulation because this can lead to myocardial infarction, heart failure, and sudden cardiac death.

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KEY WORDS cardiac strangulation, myocardial infarction, pericardial defect

APPENDIX For supplemental videos, please see the online version of this paper.