

Letter



Lethal Right Heart Failure due to Ruptured Aortic Arch Aneurysm into the Main Pulmonary Artery

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Ruptured aortic arch aneurysm is a fatal condition, and aortopulmonary artery fistula is an infrequent complication of ruptured aneurysm, occurring in approximately 3.7% of ruptured aneurysm cases as reported from autopsy case series.¹⁾ Sudden cardiac death due to severe pulmonary hypertension and cardiac dysfunction from high cardiac output are often the first and only signs of aortopulmonary fistula.²⁾ Thus, early and accurate diagnosis is essential in hemodynamically unstable patients. Here, we report a rare case of aortopulmonary fistula caused by abrupt rupture of an aortic arch aneurysm, which was diagnosed using echocardiography after cardiopulmonary-cerebral resuscitation (CPCR).

The patient was a 73-year-old male with a history of a cerebral aneurysm who was waiting for an aneurysm embolization. The patient received a preoperative evaluation in May 2018 at our center; chest radiography showed mild mediastinal widening (**Figure 1A**) and transthoracic echocardiography showed normal left and right ventricular contractility as well as, an incidental finding of 3.5-cm abdominal aortic aneurysm. The patient did not have any apparent symptoms from the abdominal aneurysm.

After 2 months, he was referred to our emergency unit due to sudden-onset dyspnea. At admission, the patient was conscious and alert, and had a blood pressure of 108/54 mmHg, pulse rate of 69 beats/min, oxygen saturation of 96% (room air), body temperature of 36.2°C, and respiratory rate of 22 breaths/min with dyspnea. Initial laboratory data showed white blood cell count of 14,500/ μ L, hemoglobin count of 14.9 g/dL, platelet count of 174×10^3 / μ L, mild liver and renal dysfunction, mild elevated C-reactive protein, and metabolic acidosis in arterial blood gas analysis. Brain natriuretic peptide was elevated (432 pg/mL). Troponin-I was 7.118 ng/mL, creatine kinase MB was 24.2 ng/mL and D-dimer was 6.03 μ L/mL. Chest radiography showed exacerbation of mediastinal widening and pulmonary congestion, especially in the right side of the lung (**Figure 1B**). Electrocardiogram showed sinus tachycardia, with a pulse rate of approximately 110 beats/min. We tried to perform contrast-enhanced chest computed tomography, but the patient suddenly exhibited altered mental status and decreased blood pressure after taking pre-enhanced phase computed tomography imaging (**Figure 1C and D**), which revealed an 8-cm saccular aneurysm. He subsequently suffered recurrent cardiac arrest and was resuscitated with CPCR. We therefore performed bedside transthoracic echocardiography, which revealed that left ventricular end-diastolic dimension was 49 mm, and ejection fraction was 52% without

Conflict of Interest

The authors have no financial conflicts of interest.

Author Contributions

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regional wall motion abnormality; however, the right side of the heart showed severe dysfunction (**Supplementary Video 1**). In a short time, refractory cardiogenic shock was developed and venoarterial extracorporeal membrane oxygenation (ECMO) was inserted. However, ECMO flow was very low and blood pressure was not fully recovered in spite of successful ECMO implantation. Moreover, central venous pressure was not reduced (13 cm H₂O). Transesophageal echocardiography after ECMO insertion showed an aneurysm of the aortic arch near the main pulmonary artery and a 5-mm tissue defect between them. Flow communication on color Doppler revealed that the aortic arch aneurysm rupture communicated with the main pulmonary artery, thereby causing severe systolic dysfunction of the right ventricle (**Figure 2** and **Supplementary Video 2**). When we reviewed the transthoracic echocardiography again, the aneurysm was compressing the left pulmonary artery near the pulmonary bifurcation and flow communication between the aneurysm and pulmonary artery was observed (**Figure 3** and **Supplementary Video 3**). Unfortunately, he did not survive because of ischemic brain injury.

Ruptured aneurysm of the aortic arch into the pulmonary artery and fistula formation is a rare but life-threatening condition. Generally ruptured aneurysm presents as chest pain with cardiac tamponade or hypovolemic shock due to hemorrhage into other spaces (e.g. aorto-

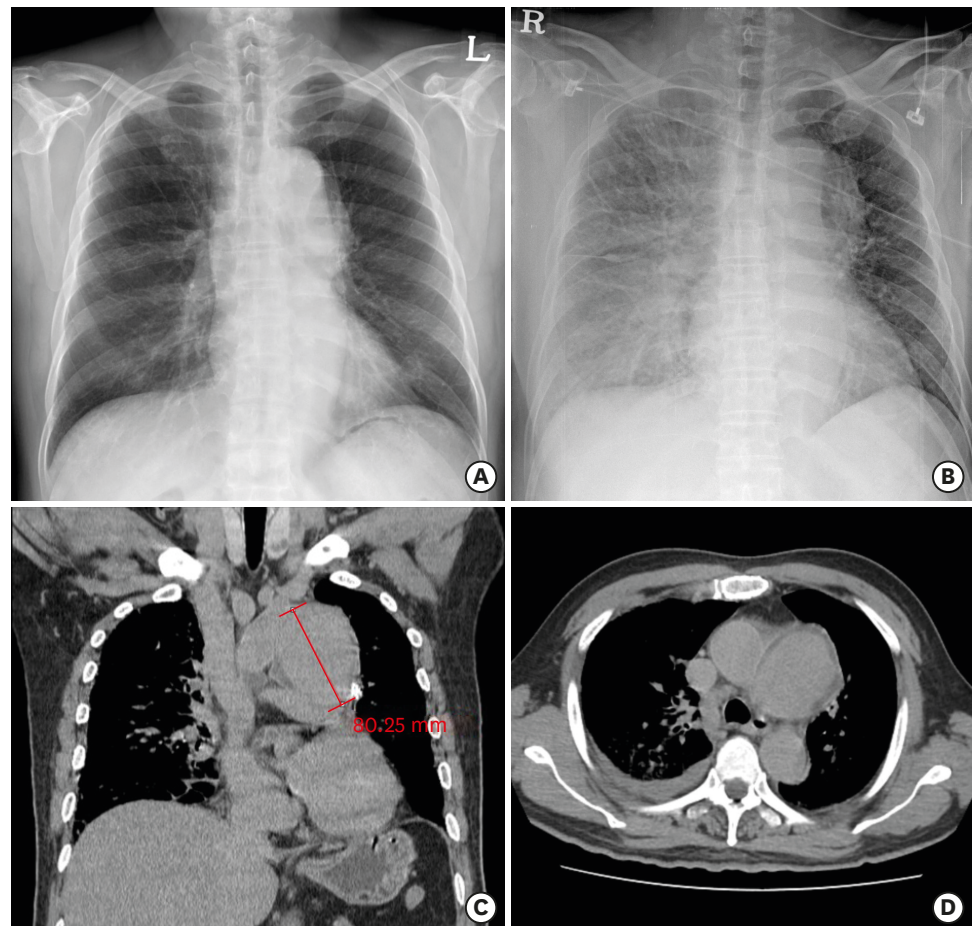


Figure 1. (A) Initial preoperative chest X-ray showing mild mediastinal widening. (B) Chest X-ray in the emergency unit showing exacerbation of mediastinal widening and haziness of the right lung. (C) Coronal section of pre-enhanced phase computed tomography showing the aortic arch aneurysm with a maximum diameter of 8.0 cm (D) Axial section of pre-enhanced phase computed tomography. White arrow indicates the saccular aneurysm.

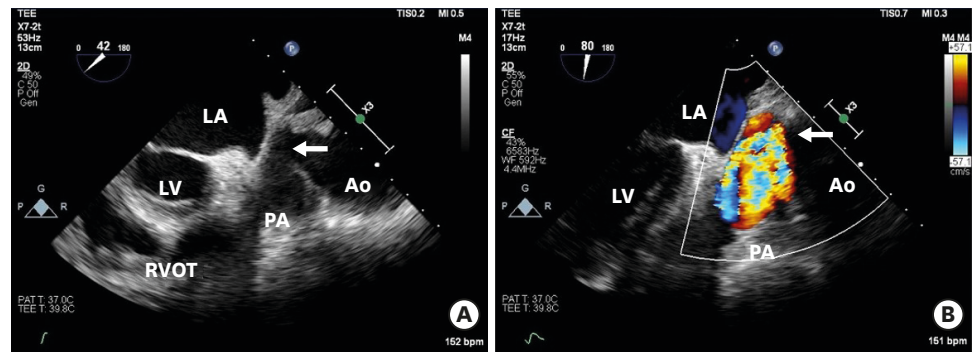


Figure 2. Transesophageal echocardiography in the ECMO insertion state. (A) White arrow indicates the tissue defect between the Ao and the main PA; (B) Doppler color-flow mode showing the flow communication between the Ao and the main PA. Ao = aortic arch aneurysm; ECMO = extracorporeal membrane oxygenation; LA = left atrium; LV = left ventricle; PA = pulmonary artery; RVOT = right ventricle outflow tract.

esophageal fistula).²⁾ In our present case, the aneurysm ruptured into the main pulmonary artery, which primarily presented as right heart failure due to left-to-right shunt, thereby causing volume overload in the pulmonary circulation. Hydrostatic pressure due to increased blood flow increased the venous pressure and resulted in pulmonary capillary damage and immediate subsequent pulmonary edema.³⁾ Because the aneurysm was compressing the left pulmonary artery, the shunt flow was confined to the right pulmonary artery and resulted in unilateral pulmonary edema.

Complications of aortic arch aneurysm show rapid progression and severity, such as acute heart failure, pulmonary congestion and cardiac arrest. Therefore, it is necessary to diagnose quickly and accurately for management. Currently chest computed tomography is the mainstay diagnostic method for thoracic artery diseases, with a 92% accuracy in detecting abnormalities of the thoracic aorta,⁴⁾ however, conducting computed tomography may not be feasible in hemodynamically unstable patients. Conversely, echocardiography, especially through the transesophageal approach is useful for finding the exact location of the lesion even in unstable patients.⁴⁾ Echocardiography allows for clear visualization of the aorta and its major branches, and transesophageal echocardiography has been shown to be superior to transthoracic echocardiography for assessing the aortic arch and the descending thoracic

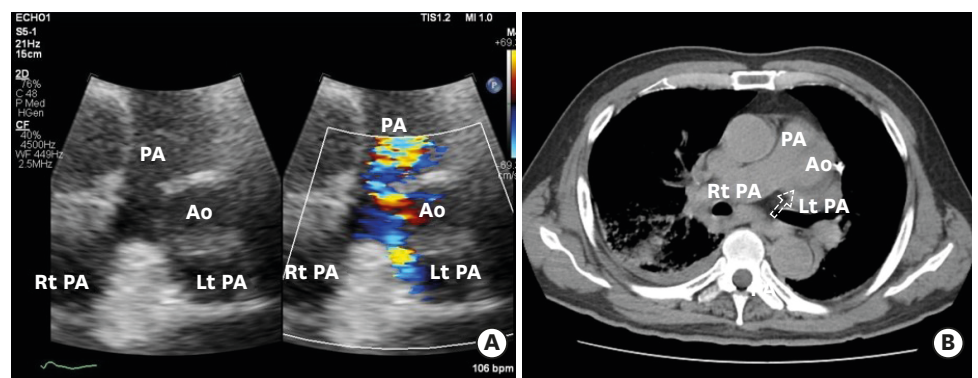


Figure 3. (A) Transthoracic echocardiography. Short-axis view (left panel) shows the aneurysm compressing the Lt PA near the PA bifurcation, and the color Doppler mode (right panel) shows the flow communication between the aneurysm and the PA. (B) Pre-enhanced phase computed tomography showing the aneurysm compressing Lt PA (dotted arrow). It is consistent with the findings of the transthoracic echocardiography. Ao = aortic arch aneurysm; Lt PA = left pulmonary artery; PA = pulmonary artery; Rt PA = right pulmonary artery.

aorta.⁴⁾ In our present case, transesophageal echocardiography was essential in confirming the presence and precise location of the aneurysm rupture.

Our case report adds information to the rare and life-threatening condition of ruptured aneurysm of the aortic arch into the pulmonary artery. If sudden right heart failure develops in patients with aortic arch aneurysm, consider complications such as aortopulmonary fistula and manage them immediately. Also, bedside echocardiography is useful for making an early and accurate diagnosis of the aneurysm rupture in hemodynamically unstable patients.

SUPPLEMENTARY MATERIALS

Supplementary Video 1

The initial bedside transthoracic echocardiography, which revealed that normal left ventricular ejection fraction and severe dysfunction of right ventricle.

[Click here to view](#)

Supplementary Video 2

Transesophageal echocardiography in the ECMO insertion state. The flow communication on color Doppler showed that the aortic arch aneurysm rupture communicated with the main pulmonary artery.

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Supplementary Video 3

In transthoracic echocardiography, flow communication between the aneurysm and pulmonary artery was observed.

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