

[CASE REPORT]

Successful Pulmonary Artery Embolization for the Management of Hemoptysis in a Patient with Eisenmenger Syndrome Caused by Patent Ductus Arteriosus

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Abstract:

The patient was a 19-year-old woman who was diagnosed with patent ductus arteriosus complicating Eisenmenger syndrome at a previous medical institution. She was referred to our hospital and arranged for lung transplantation. She developed hemoptysis after the introduction of i.v. epoprostenol, which was administered as a bridging treatment while the patient awaited lung transplantation. She continued to suffer from recurrent hemoptysis, even after switching from i.v. epoprostenol to i.v. treprostinil. Angiography of the systemic and pulmonary arteries revealed the vessel responsible for the recurrent hemoptysis and pulmonary artery embolization was successfully performed. It is essential to identify the culprit vessel and physicians must not hesitate in performing embolization when patients develop lethal hemoptysis.

Key words: hemoptysis, lung hemorrhage, pulmonary arterial hypertension, Eisenmenger syndrome, embolization, epoprostenol

(Intern Med 56: 3299-3304, 2017)

(DOI: 10.2169/internalmedicine.9071-17)

Introduction

Hemoptysis is a frequent complication of Eisenmenger syndrome (ES). It can lead to the deterioration of the patient's condition and can be life-threatening when patients show no response to conservative therapies (1). Although a few case reports have described the performance of bronchial artery embolization (BAE) for the treatment of hemoptysis in ES (2, 3), there have been no reports on pulmonary artery embolization. We herein describe the case of 19-year-old woman with ES caused by patent ductus arteriosus (PDA) in whom recurrent lethal hemoptysis was successfully treated by pulmonary artery embolization.

Case Report

The patient was a 19-year-old Japanese woman. She was diagnosed with PDA by a previous medical institution at 6 years of age; there was no indication for surgical or catheter intervention due to ES. Since then, she had been treated by home oxygen therapy and oral pulmonary vasodilators.

When she was 18 years of age, she was referred to our hospital due to a worsening of her symptoms. At her first evaluation at our hospital, electrocardiography and echocardiography showed right ventricular hypertrophy (Fig. 1), the contrast-enhanced chest computer tomography (CT) findings were consistent with PDA, lung perfusion scintigraphy showed a mottled pattern and right-to-left shunt (Fig. 2), her World Health Organization functional class (WHO-FC) was III, percutaneous oxygen saturation (SpO₂) showed differen-

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Received: February 21, 2017; Accepted: April 5, 2017; Advance Publication by J-STAGE: October 11, 2017
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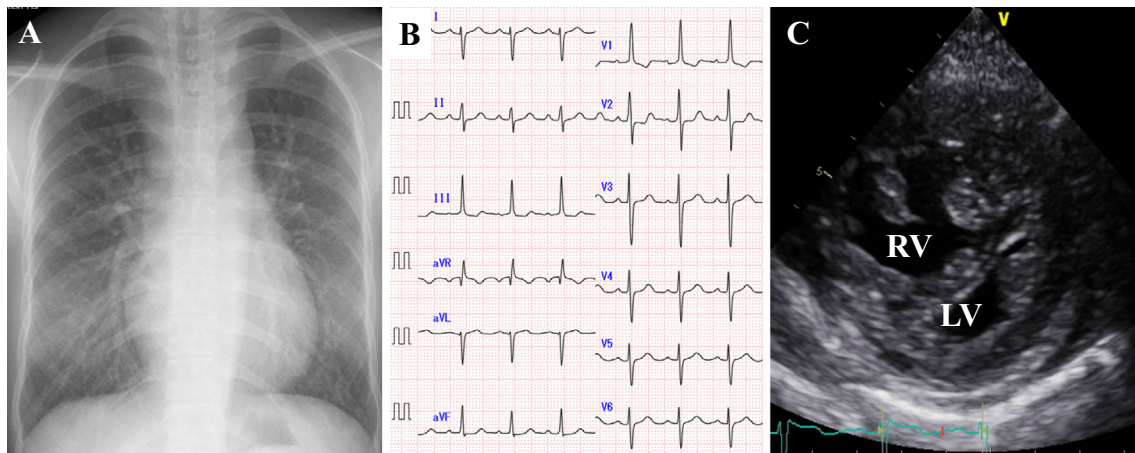


Figure 1. The clinical images at our first evaluation. A: Chest X-ray showed heart enlargement (cardiothoracic ratio, 54.5%). B: Electrocardiography showed right axis deviation and right ventricular hypertrophy. C: Echocardiography showed a D-shaped ventricle with a tricuspid regurgitation pressure gradient of 124 mmHg and right ventricular systolic dysfunction (fractional area change, 30%). LV: left ventricle, RV: right ventricle

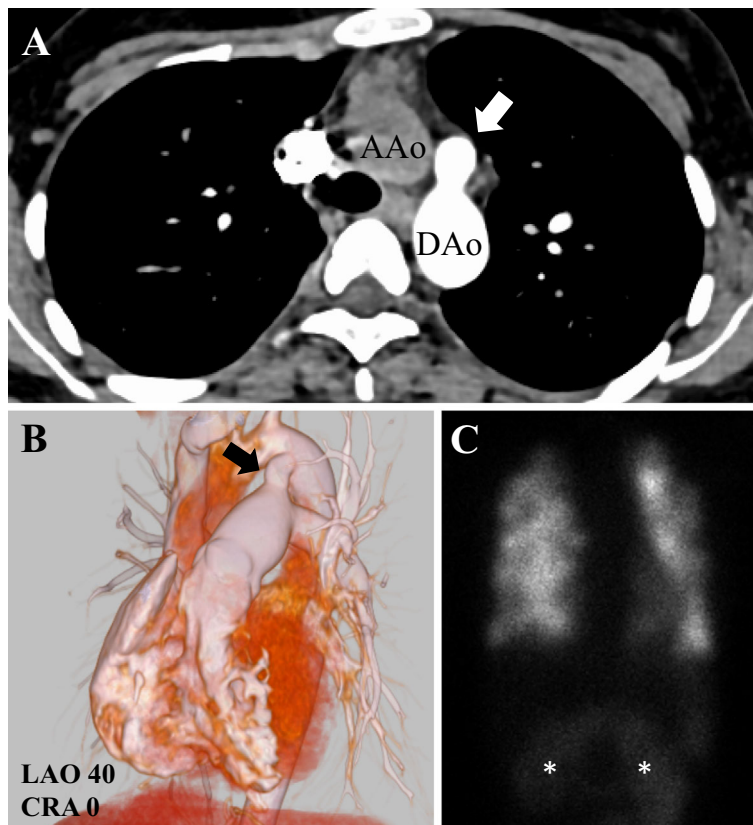


Figure 2. Representative clinical images of Eisenmenger syndrome. A: Contrast-enhanced chest CT detected right to left shunt via PDA (white arrow) to the descending aorta in the pulmonary arterial phase. B: A 3D image revealed PDA. C: Lung perfusion scintigraphy showed a mottled pattern suggesting severe remodeling of the pulmonary vessels and the uptake of tracer by the kidneys (asterisk), suggesting right to left shunt. AAo: ascending aorta, CT: computed tomography, DAo: descending aorta, PDA: patent ductus arteriosus

tial cyanosis, the oxygen saturation in the left finger (87%) was lower than that in the right finger (92%) without oxygen therapy, and right heart catheterization showed severe pulmonary arterial hypertension (mean pulmonary artery

pressure, 70 mmHg; cardiac index, 2.71 L/min/m²; pulmonary vascular resistance, 16.4 Wood units; pulmonary-systemic blood flow ratio, 0.86). We therefore arranged for lung transplantation and introduced i.v. epoprostenol as a

bridging-therapy.

At four months after the first visit to our hospital, the patient developed hemoptysis and was admitted to our hospital. The hemoptysis was controlled by 5 days of non-

Table. Laboratory Data on Admission.

Blood cell count		Biochemistry	
White blood cell (μL)	6,700	Total protein (g/dL)	6.7
Hemoglobin (g/dL)	16.7	Albumin (g/dL)	4.7
Platelet ($10^4/\mu\text{L}$)	6.5	AST (U/L)	14
Coagulation test		ALT (U/L)	9
APTT (sec)	38.1	LDH (U/L)	139
Prothrombin time % (%)	72.9	BUN (mg/dL)	15.7
FDP ($\mu\text{g/mL}$)	<2.5	Creatinine (mg/dL)	0.56
Arterial blood gas ($\text{FiO}_2 1.0$)		Uremic acid (mg/dL)	5.3
pH	7.266	Na^+ (mEq/L)	141
PaCO_2 (Torr)	56.4	K^+ (mEq/L)	3.3
PaO_2 (Torr)	55.7	Cl^- (mEq/L)	105
HCO_3^- (mEq/L)	24.9	CRP (mg/dL)	<0.03
SaO_2 (%)	82.6	BNP (pg/mL)	37

APTT: activated partial thromboplastin time, FDP: fibrin degradation products, AST: aspartate aminotransferase, ALT: alanine aminotransferase, LDH: lactate dehydrogenase, BUN: blood urea nitrogen, CRP: C-reactive protein, BNP: brain natriuretic peptide

invasive positive pressure ventilation.

At two months after the previous admission, however, she was admitted again due to massive hemoptysis. On admission, her blood pressure was 93/59 mmHg, her heart rate was 89 beats per min, the SpO_2 of the right finger was 85% with the administration of 8 L of oxygen per min and her respiratory rate was 30 breaths per min. Her complete blood counts showed a gradual decrease in her platelet count (from 171,000/ μL to 65,000/ μL) (Table) after the introduction of epoprostenol but did not show anemia. Chest radiography and CT showed an infiltrative shadow at the right middle-lower lung field (Fig. 3). The patient was intubated for positive-pressure mechanical ventilation and platelet transfusion was performed. We suspected epoprostenol-induced thrombocytopenia and switched from i.v. epoprostenol (37 ng/kg/min) to i.v. treprostinil (45 ng/kg/min), which resulted in a gradual increase her platelet count. We were able to extubate the patient on the 14th hospital day and moved her from the intensive care unit (ICU) to the ward, Rehabilitation was started on the 17th hospital day. Although her platelet count gradually increased to 98,000/ μL at one month after the switch, she had 3 more episodes of hemoptysis, after which she was managed in the ICU.

Each time, chest CT showed severe hemorrhage (Fig. 3), which was always located in the right middle lobe, suggest-

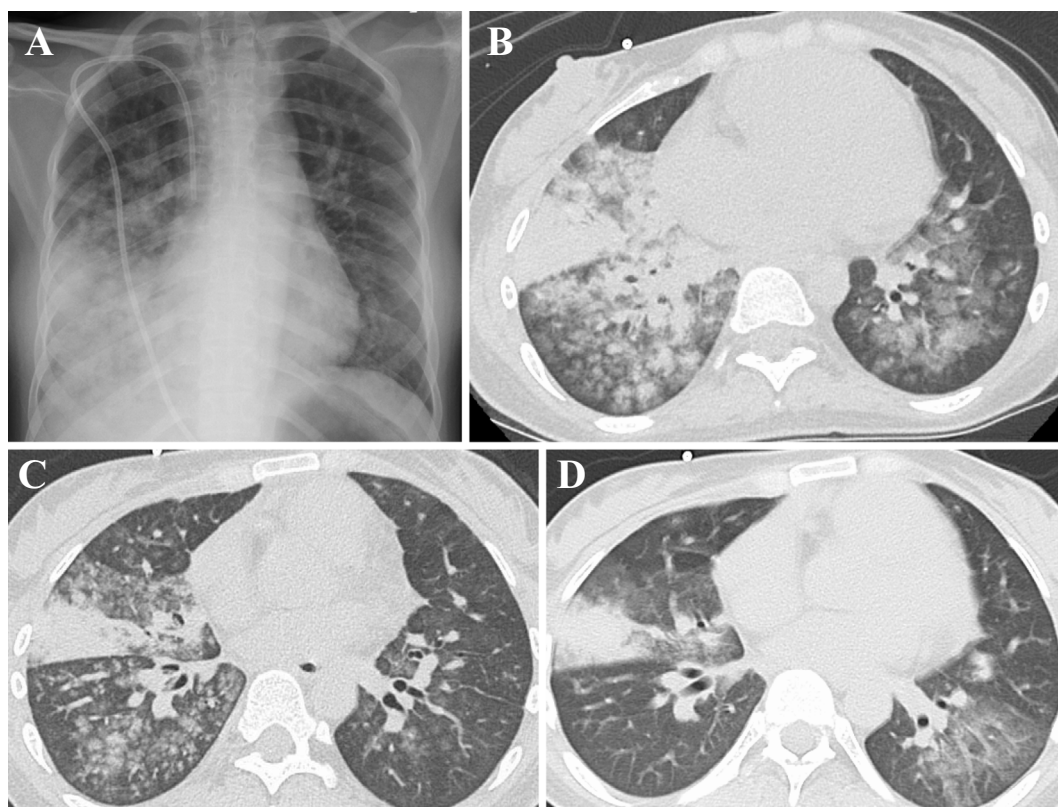


Figure 3. Chest radiography and chest CT images obtained after the development of hemoptysis. A chest radiography (A) showed infiltrative shadows at the right middle-lower lung field in the second episode of hemoptysis. Chest CT (B, C, and D were taken at second, third, and fourth hemoptysis, respectively.) showed severe lung hemorrhage originating from the vessels in right middle lobe (A4 region) and surrounding consolidation indicated trans-bronchially spreading hemorrhage. CT: computed tomography

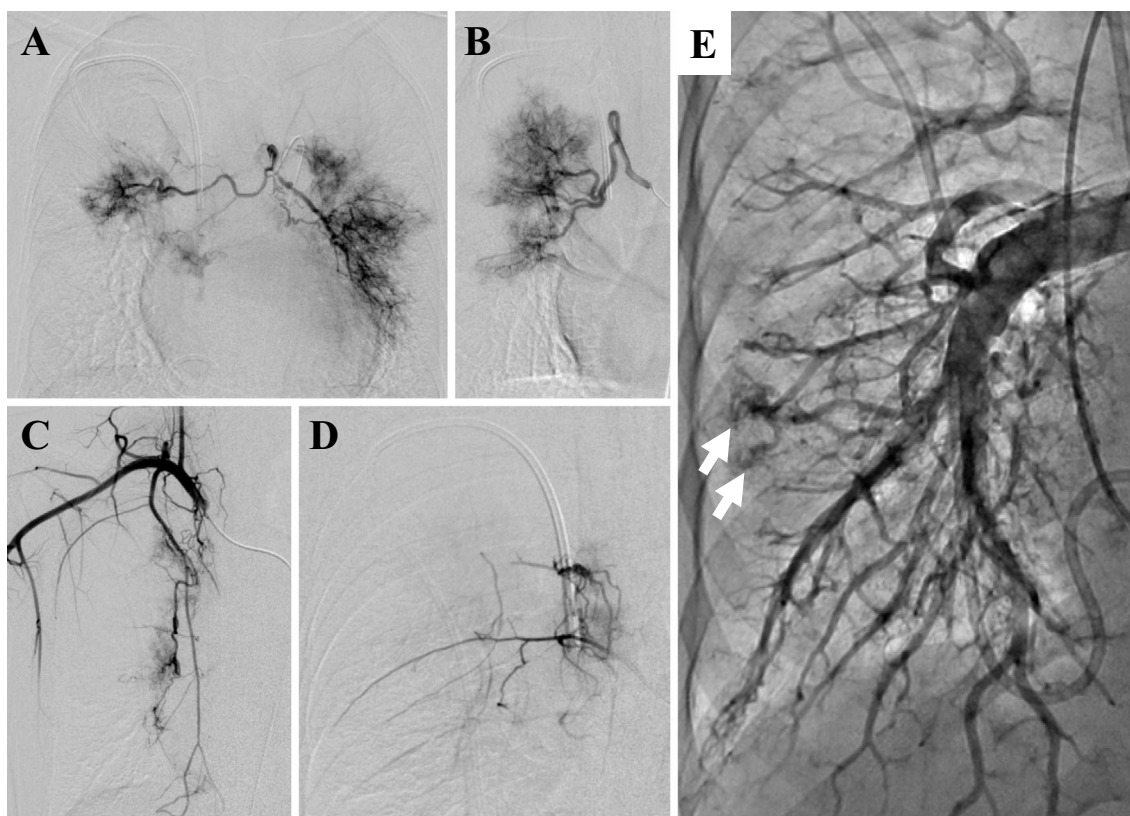


Figure 4. Angiography of the systemic and pulmonary arteries. Although angiography of the left bronchial artery (A), right bronchial artery (B), right intrathoracic artery (C) and right intercostal artery (D) revealed no culprit lesion that could have caused hemoptysis. Angiography of the right pulmonary artery showed a fragile vascular network in the peripheral region of the right middle lobar artery (white arrows in E).

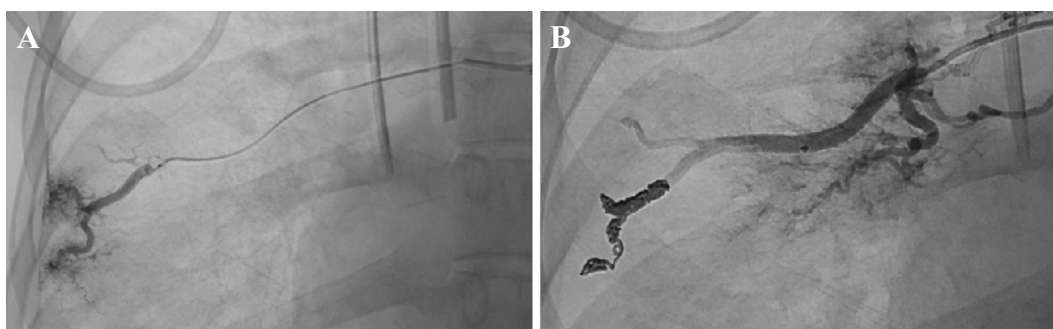


Figure 5. Selective angiography before and after embolization. The culprit lesion of hemoptysis was embolized with a ϕ 2.0mm C-STOPPER Coil (PIOLAX, Yokohama, Japan).

ing that the vessels in the lesion were fragile. To identify the culprit vessel, we performed angiography of the bronchial arteries, intercostal arteries, a right intrathoracic artery and the pulmonary arteries (Fig. 4). Although no culprit lesion was identified in the systemic arteries, we could find that the A4 branch in the peripheral region of the right middle lobar artery was fragile (white arrows in Fig. 4E). Embolization was necessary for the patient to survive.

The information on pulmonary artery embolization for patients with ES was extremely limited. Thus, we held a multidisciplinary conference that included a cardiologist, a radi-

ologist, a respiratory surgeon and an anesthesiologist, to discuss the safety and efficacy of this intervention. As a result, we decided to perform pulmonary artery embolization. The peripheral segment of right middle lobar artery was catheterized with a 2-French Estream microcatheter (Toray Medical, Tokyo, Japan). Through the microcatheter, the lesion was embolized with ϕ 2.0 mm C-STOPPER Coil (PIOLAX, Yokohama, Japan) (Fig. 5). The intervention was successfully performed without any complications other than a mild deterioration of the patient's hemodynamics (the mean pulmonary artery pressure increased from 64 to 66 mmHg, the

cardiac index decreased from 1.92 to 1.62 L/min/m², and the pulmonary vascular resistance increased from 20.5 to 25.3 Wood units.). She has had no other episodes of hemoptysis for 13 months and has been on the waiting list for lung transplantation.

Discussion

ES is characterized by congenital heart defects with elevated pulmonary arterial pressure and pulmonary vascular resistance, which results in reversed or bidirectional shunts (4). The clinical presentation of ES includes central cyanosis, dyspnea, fatigue, syncope, and right heart failure (5). The survival of patients with ES is reduced in comparison to patients with simple congenital heart disease without ES. According to a large cohort of patients with adult congenital heart disease in the UK, the median life expectancy of patients with ES is 42 years (6). Hemoptysis is a frequent complication and a major cause of mortality in ES (1). The management of hemoptysis is important for patients with ES because the frequency increases with age, with almost all patients experiencing hemoptysis by their forties (4).

In this case, although various pathophysiological mechanisms have been suggested as causes of hemoptysis in ES, we think that there were two causes of the recurrent hemoptysis (7). The first possible cause was epoprostenol, which contributes to impaired platelet aggregation and thrombocytopenia (8). Although switching from i.v. epoprostenol to i.v. treprostinil has the potential to increase the platelet level (9), it is not a radical treatment for recurrent hemoptysis. We realized that care should be taken in the introduction of epoprostenol to patients with ES, not only because the hemodynamic improvement is insufficient, but also because of the increased risk of hemoptysis. The second possible cause was the fragility of the severely remodeled pulmonary artery. We were able to identify the fragile A4 branch of the right pulmonary artery by performing angiography of all of the related systemic and pulmonary arteries. This lesion might be derived from pressure and volume overload due to PDA complicating ES. Further hemoptysis was avoided for more than 1 year after the embolization of the lesion. Thus, it is essential to perform angiography for identification of the culprit vessel and physicians should not hesitate to perform embolization of the culprit vessel in case of lethal hemoptysis.

Although there have been a few reports on BAE in patients with ES complicating hemoptysis (2, 3), we were unaware of any case reports regarding pulmonary artery embolization in such patients. We thought that pulmonary artery embolization was very distinct from BAE, which does not directly affect the pulmonary hemodynamics. Our most serious concern was that the procedure might lead to the deterioration of the patient's pulmonary hemodynamics and then to an elevation of the pulmonary arterial pressure, resulting in the deterioration of lethal hemoptysis. We held a

multidisciplinary conference in our hospital to discuss the risk of uncontrollable hemorrhage during the intervention. Considering the extremely limited information and our lack of experience, we prepared a backup system for rescue lung lobectomy. However, we successfully embolized the vessel with no sign of hemorrhage or severe pulmonary infarction.

To the best of our knowledge, this is the first report of successful pulmonary artery embolization in a patient with ES suffering from recurrent hemoptysis. We were able to save the patient by a careful examination to identify the culprit vessel and a close troubleshooting discussion. Although embolization is not a definitive treatment, it should be considered for patients suffering from recurrent hemoptysis. It can be expected to be an effective bridging treatment for patients waiting for lung transplantation.

Author's disclosure of potential Conflicts of Interest (COI).

Kazuhiko Nakayama: Research funding, Actelion Pharmaceuticals Japan Ltd. and Bayer Holding.

Acknowledgement

We thank Hiromi Matsubara in Okayama Medical center for the technical guidance on pulmonary artery embolization, Hiroyuki Fukushima in Keio University School of Medicine and Osamu Yamada in National Cerebral and Cardiovascular Center Hospital for discussing the indications for pulmonary artery embolization, and Hiroshi Date in the Department of Thoracic Surgery in Kyoto University and Masato Minami in Osaka University Hospital for determining the indications for lung transplantation.

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Intern Med 56: 3299-3304, 2017