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CASE REPORT

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Balloon pulmonary angioplasty under awake veno-arterial extracorporeal membrane oxygenation in a patient with class III obesity with chronic thromboembolic pulmonary hypertension complicated with multiple serious comorbidities

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Abstract

Chronic thromboembolic pulmonary hypertension (CTEPH) is a chronic disease that can rapidly deteriorate into circulatory collapse when complicated by comorbidities. We herein describe a case involving a 43-year-old woman with class III obesity (body mass index of 63 kg/m^2) and severe CTEPH associated with total occlusion of the left main pulmonary artery who subsequently developed circulatory collapse along with multiple comorbidities, including acute kidney injury, pulmonary tuberculosis, and catastrophic antiphospholipid syndrome. The patient was successfully treated with two sessions of rescue balloon pulmonary angioplasty with veno-arterial extracorporeal membrane oxygenation (V-A ECMO) support under local anesthesia without sedation, at cannulation and during the V-A ECMO run, to avoid invasive mechanical ventilation. This case suggests the potential usefulness of rescue balloon pulmonary angioplasty under awake V-A ECMO support for rapidly deteriorating, inoperable CTEPH in a patient with class III obesity complicated with multiple comorbidities.

KEYWORDS
balloon pulmonary angioplasty, chronic thromboembolic pulmonary hypertension, comorbidity, obesity, veno-arterial extracorporeal membrane oxygenation

CASE DESCRIPTION

A 43-year-old woman, with class III obesity (body weight of 168 kg and body mass index [BMI] of 63 kg/m^2) and a prior history of type-2 diabetes mellitus, asthma, and uterine cancer which was treated surgically, and without any episodes of venous thromboembolism, was admitted to our hospital due to severe dyspnea, hemoptysis, and generalized edema. She presented with World Health Organization functional class (WHO-FC) IV symptoms

and a high brain natriuretic peptide (BNP) level of 2,054 pg/mL, in addition to continuous wheezing due to asthma. Chest radiography revealed cardiomegaly (cardiothoracic ratio of 57%) with consolidation in the left lower lung field, and electrocardiography revealed sinus rhythm with a heart rate of 94 beats/min and right axis deviation, low voltage, and a negative T wave at V1–V3. Transthoracic echocardiography demonstrated an estimated systolic pulmonary arterial pressure of 92.3 mmHg and the severely dilated right ventricle with the compression of the left ventricle and slight pericardial effusion (Figure 1a). Blood tests revealed a serum creatinine level of 2.44 mg/dL (0.72 mg/dL at baseline), indicating acute kidney injury (AKI), as well as C-reactive protein and d-dimer levels of 10.62 mg/dL and 5.71 μg/mL, respectively. Although the patient tested negative for all autoantibodies, she did test positive for lupus anticoagulant twice. Noncontrast-enhanced computed tomography (CT) demonstrated reticular infiltration in the left lung with left pleural effusion (Figure 1b), when we refrained from using contrast agent due to AKI. Lung perfusion scintigraphy demonstrated complete perfusion defects in the left lung and multiple segmental defects in the right lung, indicating central-type chronic thromboembolic pulmonary hypertension (CTEPH) (Figure 1c). Right heart catheterization revealed a mean pulmonary artery wedge pressure of 10 mmHg, a pulmonary arterial pressure (PAP) of 120/28 (59) mmHg, pulmonary vascular resistance (PVR) of 5.8 Wood units, and noninvasive blood pressure of 98/59 (72) mmHg, indicating over-systemic precapillary pulmonary hypertension (PH). Despite continuous intravenous anticoagulation, increased inotropic agents, and the careful introduction of intravenous epoprostenol and tadalafil, phosphodiesterase type-5 inhibitor, she had refractory right-sided heart failure, resulting in circulatory collapse. Eleven days after admission, we initiated veno-arterial extracorporeal membrane oxygenation (V-A ECMO) with femoral–femoral 23-French–16-French cannulas and pump flow of around 2.0 L/min without sedation, to avoid worsening of pulmonary hemodynamics owing to positive end-expiratory pressure and invasive mechanical ventilation (MV)-related complications, followed by continuous renal replacement therapy. Moreover, she was diagnosed as catastrophic antiphospholipid syndrome (CAPS) as follows; (1) more than three organs involving thrombosis (CTEPH, old cerebral infarction, and left internal jugular vein), in addition to proteinuria, (2) development of manifestations simultaneously or in less than 1 week, except cerebral infarction of unknown onset, (3) confirmation of small pulmonary artery occlusion, and (4) confirmation of the presence of lupus anticoagulant twice at least 6 weeks apart, according to the preliminary criteria for CAPS.[1] By the multidisciplinary CTEPH team, operability for pulmonary thromboendarterectomy (PTE) was assessed and concluded as inoperable at our center due to class III obesity, risk of invasive MV for unstable hemodynamics even under V-A ECMO (mean PAP of 53 mmHg, elevated right atrial pressure of 23 mmHg, and a further increased BNP level of 3,470 pg/mL), and multiple serious comorbidities; AKI, active pulmonary tuberculosis, and CAPS.

Aside from high-dose steroids for CAPS, we performed two rescue balloon pulmonary angioplasty (BPA) procedures through the right internal jugular vein at an interval of 3 days. To guide BPA, we adopted selective angiography and thin-slice noncontrast-enhanced CT using the lung window setting to visualize the bronchial tubes that normally run in along the pulmonary arteries (PA) (Figures 1d, 1e, 1f).[2] The first BPA targeted right A3, A5, and A6, whereas the second one targeted right A7, A1, A2, A3, and A5, including optimization of the vessels treated in the first session, without any complications. After BPA, a significant decrease in BNP levels from 3,470 to 288 pg/mL and PAP from 120/28 (59) to 46/26 (33) mmHg had been noted. She was sequentially weaned from awake V-A ECMO and renal replacement therapy, underwent four additional BPA procedures for the right PA, including optimization of the vessels treated previously, and was switched from intravenous epoprostenol to oral selexipag, selective prostacyclin receptor agonist. After improvements in AKI, contrast-enhanced CT confirmed total occlusion of the left main PA (Figures 1g, 1h), without any changes in the vessel wall characteristic of vasculitis such as Takayasu arteritis including the systemic arteries. Follow-up catheterization indicated a PAP of 40/14 (28) mmHg and PVR of 1.6 Wood units with oral double PH-specific therapies and maintenance-dose prednisolone. After confirming that the patient's pulmonary tuberculosis was inactive following two antituberculosis agents and rehabilitation, she was discharged at a body weight of 118 kg (BMI of 44 kg/m^2) on warfarin, with WHO-FC II symptoms.

DISCUSSION

Reports have shown that WHO-FC IV, age >70 years, the severity of preoperative PVR, the presence of right ventricular failure, obesity, and PH duration affected postoperative survival in patients with CTEPH undergoing PTE.[3] Gilbert et al. reported that only BMI was significantly higher in nonsurvivors after PTE.[4] However, Fernandes et al. reported in 476 patients undergoing PTE that there were no differences in overall in-hospital mortality and postoperative complications among four groups categorized

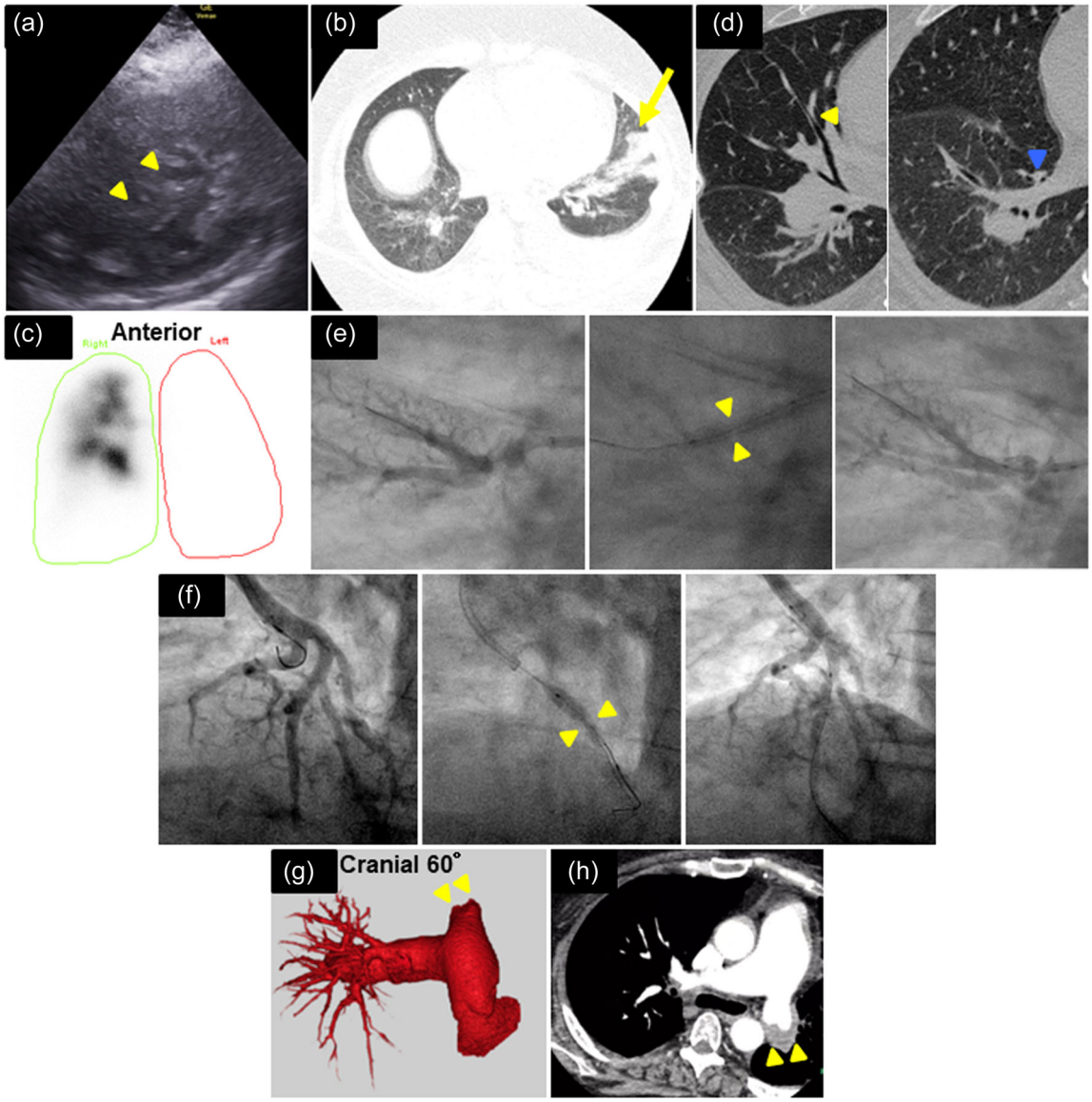

FIGURE 1 Transthoracic echocardiography, noncontrast-enhanced computed tomography (CT), lung perfusion scintigraphy, selective pulmonary angiography before and after balloon pulmonary angioplasty, and contrast-enhanced CT. (a) Transthoracic echocardiography in the parasternal short axis view showing a poor image due to class III obesity and the severely dilated right ventricle with the compression of the left ventricle (yellow arrowheads). (b, d) Noncontrast-enhanced CT showing reticular infiltration in the left inferior lobe (arrow) with left pleural effusion, implying the focus of the pulmonary tuberculosis. Thin-slice noncontrast-enhanced CT with the window setting adjusted to the lung window showing the bronchial tubes and concomitant arteries such as right A5 (yellow arrowhead) and A7 (blue arrowhead). (c) Lung perfusion scintigraphy showing complete perfusion defects in the left lung and multiple segmental defects in the right lung. (e, f) Selective pulmonary angiography for right A5 (e) and A7 (f) before and after balloon pulmonary angioplasty. Note: An indentation in the balloons (yellow arrowheads) was observed just before the balloons were fully dilated. (g, h) Contrast-enhanced CT with a three-dimensional image (anteroposterior–cranial view) indicating total occlusion at the origin of the left main pulmonary artery (yellow arrowheads).

by baseline BMI, although improvements in PVR were greatest in the lower BMI groups.[5] Interestingly, they included only one patient with a BMI of >55.[5] Apart from class III obesity (BMI of 63 kg/m^2), our patient suffered from multiple comorbidities, including AKI, uncontrolled asthma, active pulmonary tuberculosis, and CAPS almost simultaneously, where CAPS seemed to most affect hemodynamic worsening through multiple small vessel thrombosis. Although there are some CTEPH centers that may perform PTE on this patient, those comorbidities were considered inoperable for PTE.

Although CTEPH is generally a slowly progressive disease,[6,7] right-sided heart failure and pulmonary hemodynamics can rapidly deteriorate into circulatory collapse

when complicated by comorbidities. Only two reports have demonstrated survival in patients with CTEPH who underwent rescue BPA under V-A ECMO, which was managed with invasive MV under general anesthesia.[8,9] Recently, Montero et al. reported that "awake V-A ECMO" management in patients with refractory cardiogenic shock was associated with reductions in V-A ECMO-related complications, and 60-day and 1-year mortality rates.[10] They mainly included dilated cardiomyopathy, acute myocardial infarction, and fulminant myocarditis, after excluding pulmonary embolism, where awake V-A ECMO was defined as an invasive MV duration of ≤50% of the V-A ECMO run.[10] In the present report, we kept her awake without intubation or sedation during V-A ECMO, enabling reductions in invasive MV-related complications and treatment decision-making. To our knowledge, this is the first case to survive circulatory collapse of severe CTEPH following rescue BPA under awake V-A ECMO. Our findings may expand the application of awake V-A ECMO to right-sided circulatory collapse, including severe PH.

In conclusion, rescue BPA under awake V-A ECMO may be an important option in patients with CTEPH with class III obesity, multiple serious comorbidities, and subsequent circulatory collapse, if they are not candidates for PTE.

AUTHOR CONTRIBUTION

Tsukasa Sato and Shigefumi Fukui collected and interpreted data, and wrote and revised the manuscript; Takao Nakano, Kaoru Hasegawa, Hisashi Kikuta, Takeyoshi Kameyama, Yuko Shirota, Tomoyuki Endo, and Shunsuke Kawamoto revised the manuscript; Koji Kumagai, Hideo Izawa, and Tatsuya Komaru conceived the study, revised the manuscript, and supervised the study project.

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CONFLICT OF INTEREST STATEMENT

The authors declare no conflict of interest.

ETHICS STATEMENT

The subject of this report provided written informed consent for treatment and for her report to be submitted for publication.

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