DOI: 10.1002/pul2.12354

CASE REPORT

Pulmonary Circulation Open Access | PVRI Pulmonary Vascular Research Institute | WILEY

Pulmonary endarterectomy for chronic thromboembolic pulmonary hypertension with bronchial obstruction by a carcinoid tumor

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Funding information
None

Abstract

Pulmonary endarterectomy (PEA) is a standard treatment for chronic thromboembolic pulmonary hypertension (CTEPH). CTEPH combined with bronchial obstruction by a tumor is rare but should be assessed carefully because PEA for obstructed segments can be less therapeutic and make the subsequent surgical resection challenging. This report describes a case of CTEPH with bronchial obstruction by a typical carcinoid tumor in a 75-year-old man. On-site evaluation and removal of the obstructive tumor were performed bronchoscopically, increasing the effectiveness of subsequent PEA for all affected pulmonary segments. This report illustrates a PEA strategy to treat CTEPH with bronchial tumor obstruction.

KEYWORDS
bronchoscopic intervention, lung cancer, Thoracic surgery

INTRODUCTION

Pulmonary endarterectomy (PEA) is the most appropriate treatment for chronic thromboembolic pulmonary hypertension (CTEPH) when the disease is present in the proximal vessels.[1] Although persistent pulmonary hypertension after PEA can lead to a poor prognosis, PEA for less functional pulmonary segments, such as segments in which the bronchus is obstructed by a tumor, should be avoided to reduce the risk of intraoperative lung injury and airway hemorrhage.[2] In addition, because of the fragile state of the arterial wall after PEA, PEA can make subsequent thoracic surgery challenging.[3] Therefore, the pulmonary segment for PEA should be considered carefully when subsequent surgical resection of a tumor may be required. We herein report a case of a patient diagnosed with CTEPH and concurrent bronchial obstruction by a typical carcinoid tumor. PEA was performed for all affected pulmonary segments after bronchial patency was established by endobronchial tumor resection.

Abbreviations: BPA, balloon pulmonary angioplasty; CTEPH, chronic thromboembolic pulmonary hypertension; PA, pulmonary artery; PEA, pulmonary endarterectomy.

CASE DESCRIPTION

A 75-year-old man with a history of CTEPH refused operative treatment and was discharged. Three months later, he was readmitted because of recent worsening of his functional status and lower extremity swelling. On admission, he had symptoms of World Health Organization class IV pulmonary hypertension, and he required 5 L/min of oxygen to relieve his dyspnea. He had a previous history of pulmonary embolism and was receiving warfarin and riociguat. Angiography of the pulmonary artery (PA) confirmed central-type CTEPH without apparent recent progression (Figure 1c). Right heart catheterization revealed an elevated PA pressure of 48/16/28 mmHg with systemic pressure of 116/82/95 mmHg (systolic/diastolic/mean, respectively) and mean atrial pressure of 10 mmHg, whereas the pulmonary capillary wedge pressure was normal at 4 mmHg. The pulmonary vascular resistance and cardiac index were 547 $dyn \cdot s \cdot cm^{-5}$ and 1.82 $L \cdot min^{-1} \cdot m^{-2}$, respectively. Inotropic agents and diuretics were added preoperatively, which improved his impaired functional status only minimally. During the preoperative assessment, a growing mass was detected in the bronchus on contrast-enhanced computed tomography. The mass obstructed the B9 + 10 segments of the right lung, indicating that these segments were nonfunctional.

Bronchoscopic investigation was performed before PEA. A bronchoscope was inserted through an intubation tube under anesthesia with careful monitoring of the circulatory and respiratory systems. The bronchoscopic examination revealed a smooth-surfaced erythematous tumor at the right B9 + 10 entrance, completely obstructing the bronchus (Figure 1a). Rapid on-site evaluation of a cryobiopsy specimen revealed plasmacytoid cells without obvious malignant cells, indicating that complete surgical resection was not required. Resection of the tumor through the bronchoscope was performed concurrently, achieving bronchial patency without massive bleeding (Figure 1b). On the basis of the pathological findings, the tumor was diagnosed as a typical carcinoid tumor. PEA for all affected pulmonary segments was then scheduled. Because his functional status was severely impaired even with inotropic agents, early PEA was considered preferable to reduce the risk of the patient progressing to an inoperable physical state.

One week after endobronchial resection, when no occurrence of airway hemorrhage on resumed anticoagulation therapy with a heparin drip was confirmed, PEA was performed for all affected PA segments under deep hypothermic circulatory arrest (Figure 1e). The patient was successfully weaned off cardiopulmonary bypass. He was extubated when the absence of massive airway hemorrhage and reperfusion lung injury was confirmed (1 day postoperatively). Postoperatively, his functional status and dyspnea were improved. A continuous heparin drip was administered until the international normalized ratio reached the therapeutic range with warfarin therapy. Follow-up contrast-enhanced computed tomography and PA angiography showed improved perfusion of the PA (Figure 1d). Right heart catheterization before discharge revealed a normalized PA pressure of 20/4/10 mmHg, with systemic pressure of 109/70/74 mmHg (systolic/diastolic/mean,

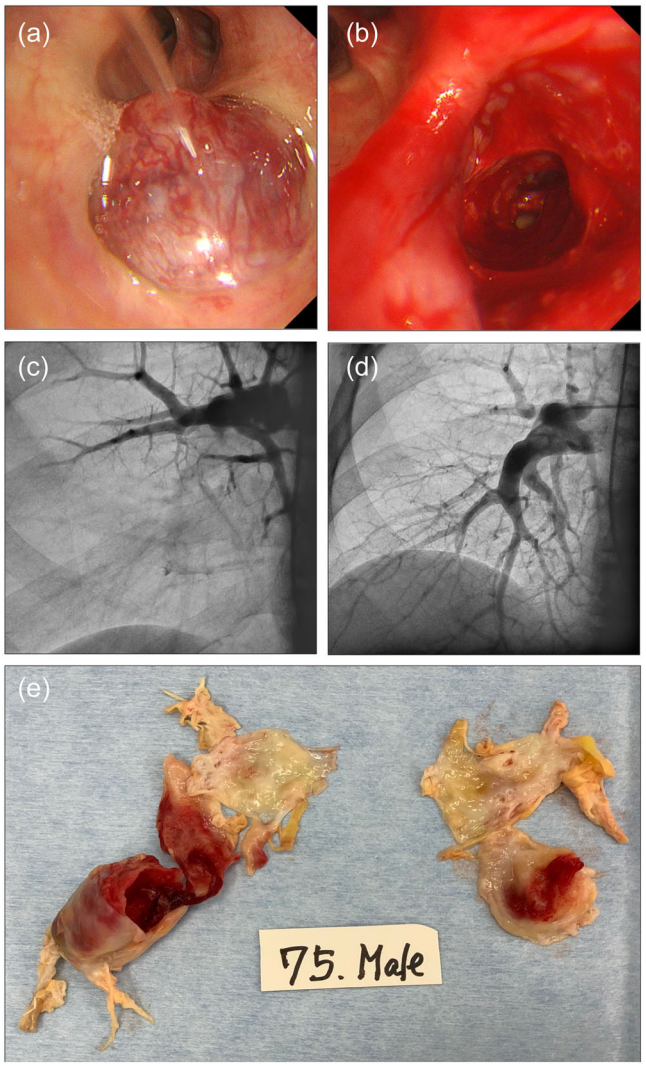

FIGURE 1 (a) A smooth-surfaced erythematous tumor was found on bronchoscopy, and (b) endobronchial resection of the bronchial carcinoid tumor was performed through bronchoscopy. (c) Preoperative angiography of the pulmonary artery confirmed central-type chronic thromboembolic pulmonary hypertension with a poorly perfused right lower segment. (d) Angiography before discharge showing improved perfusion of the pulmonary arteries. (e) Specimens removed from the pulmonary arteries during pulmonary endarterectomy.

respectively), mean atrial pressure of 1 mmHg, and pulmonary capillary wedge pressure of 4 mmHg. The pulmonary vascular resistance and cardiac output index were 130 dyn·s·cm^{-5} and 2.14 L·min^{-1}·m^{-2}, respectively. No additional procedures, such as balloon pulmonary angioplasty (BPA) or postoperative medication, such as riociguat, was required. At the latest follow-up, 12 months after the carcinoid resection and PEA, the patient was alive and well with no signs of recurrent disease. However, long-term follow-up is necessary to monitor for recurrence.

DISCUSSION

Bronchial carcinoid tumors are rare neuroendocrine tumors with an incidence of approximately 1 per 100,000 persons. Complete surgical resection is the treatment of choice, as for all other pulmonary tumors. However, for typical bronchial carcinoids, endobronchial resection has recently been considered a less or minimally invasive alternative with an excellent long-term outcome.[4] The appropriate interval between endobronchial resection and subsequent cardiac surgery requiring cardiopulmonary bypass is unclear. However, we assessed that the hemorrhagic risk during cardiopulmonary bypass was acceptable in this case because perioperative massive bleeding during endobronchial procedures is extremely rare,[5] and the patient tolerated the resumption of anticoagulation for 1 week after the endobronchial resection. In addition, because his severely impaired functional status was minimally responsive to medical therapy, even with inotropic agents, we assessed that a longer interval would worsen the patient's condition and possibly lead to inoperability and an inability to tolerate PEA, outweighing the hemorrhagic risk associated with CPB with a shorter interval.

PA hypertension limits a patient's quality of life and life expectancy, with an estimated 1-year mortality rate of 30%–40% without treatment in symptomatic patients.[1] Although PEA for CTEPH provides excellent outcomes, with a 5-year overall survival rate of 90% for patients who survive to hospital discharge, PEA is a highly complex surgical intervention with a multitude of potential morbidities and risks.[1] Persistent pulmonary hypertension after surgery is the most important cause of early postoperative mortality. The overall mortality rate is 10 times higher in patients with than without persistent pulmonary hypertension.[6] Persistent pulmonary hypertension can result from incomplete removal of thrombi,[6] indicating that complete removal of thrombi should be performed during PEA.

Notably, thrombus removal procedures can cause pulmonary hemorrhage, which is also associated with postoperative mortality. Significant pulmonary hemorrhage can occur in 1% of PEA procedures and is associated with a mortality rate of nearly 70%.[7] Although several risk factors have been identified,[2] PA hemorrhage in the absence of PEA is less likely. Additionally, considering the fragile state of the PA even 2 years after a previous PEA, there is a high risk of disastrous events during pulmonary resection in areas where PEA has been performed previously.[3] These risk factors for postoperative complications of PEA and subsequent pulmonary resection make PEA for CTEPH with bronchial obstruction by a tumor challenging.

BPA is another well-established interventional treatment for patients with CTEPH that provides effective improvements in pulmonary hemodynamics and long-term prognosis.[8] However, whereas the long-term prognosis after PEA in operable CTEPH patients is established,[6] recent evaluation of the long-term prognosis after BPA has been performed only for patients with inoperable CTEPH, chiefly involving peripheral distribution of lesions in the pulmonary arteries.[8] In addition, BPA carries the risk of interventional complications. Pulmonary injury and hemoptysis are the most commonly reported complications, accounting for up to 10% of cases after BPA, and PA perforation, dissection, and rupture have been reported in <3% of cases. These complications are serious, and the risk is significantly higher when BPA is performed for proximal lesions.[8] Therefore, in our patient, subsequent BPA after endobronchial resection of the tumor was not considered to provide a better long-term prognosis or lower the operative risks compared with subsequent PEA.

On the basis of recent reports of the prognosis in patients with a concurrent carcinoid tumor and CTEPH and knowing the risks of the procedures, we considered that endobronchial resection of the bronchial carcinoid tumor through a bronchoscope with subsequent PEA for all affected pulmonary segments was the most effective therapeutic option for our patient. This case report highlights a potential surgical strategy that cardiothoracic surgeons should consider for patients with CTEPH and a bronchial tumor.

A STATEMENT OF GUARANTOR

N/A

AUTHOR CONTRIBUTIONS

Yuki Monden assisted during surgery and wrote the first draft of the manuscript. Dai Une and Motomi Ando

participated in the multidisciplinary meeting discussing the patient's case, performed the surgery, contributed to the second draft of the manuscript, and critically reviewed and revised the manuscript. Sho Mitsumune participated in the multidisciplinary meeting discussing the patient's case, performed the bronchoscopy and carcinoid removal, and reviewed and revised the manuscript. Hiroto Shimokawahara and Hirofumi Okada diagnosed the patient with CTEPH, participated in the multidisciplinary meeting discussing the patient's case, were involved in the postoperative care, and reviewed and revised the manuscript. Kenji Yoshida, Shutaro Kato, Suzuka Kamaguchi, and Mikizo Nakai participated in the multidisciplinary meeting discussing the patient's case, were involved in the patient's postoperative care, and reviewed and revised the manuscript.

ACKNOWLEDGMENTS

We thank Angela Morben, DVM, ELS, and Jane Charbonneau, DVM, from Edanz (https://jp.edanz.com/ac) for editing a draft of this manuscript.

CONFLICT OF INTEREST STATEMENT

The authors declare that there is no conflict of interest.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

ETHICS STATEMENT

The patient provided written consent for the publication of his information and images. Institutional review board approval was not required because of the retrospective nature of this clinical case report.

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How to cite this article: Monden Y, Une D, Mitsumune S, Shimokawahara H, Okada H, Yoshida K, Kato S, Kamaguchi S, Nakai M, Ando M. Pulmonary endarterectomy for chronic thromboembolic pulmonary hypertension with bronchial obstruction by a carcinoid tumor. Pulm Circ. 2024;14:e12354. https://doi.org/10.1002/pul2.12354