CASE REPORT

Pharmacoinvasive strategy as rescue treatment for acute right ventricular failure from chronic thromboembolic pulmonary hypertension

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Learning points for clinicians

 Patients with CTEPH patients can develop decompensated right heart failure requiring intensive care. Timely hemodynamic optimization is key, and this entails right ventricular afterload reduction with targeted pulmonary hypertension therapy. Our case demonstrate the novel use of targeted pulmonary hypertension therapy and early Balloon Pulmonary Angioplasty in treating such patients.

Introduction

Chronic Thromboembolic Pulmonary Hypertension (CTEPH) is a disease of obstructive pulmonary artery remodelling as a consequence of thromboembolism. It is one of the etiological factors leading to pulmonary hypertension. Such patients can develop decompensated right heart failure requiring intensive care. We report an aggressive pharmacoinvasive strategy with upfront dual pulmonary hypertension therapy combined with semi-urgent balloon pulmonary angioplasty.

Case report

A 37-year Chinese man presented with 1-day history of acute breathlessness associated with fever, chills, cough with yellow sputum and increased lower-limb swelling for the past few months. The patient had a background of pulmonary hypertension secondary to CTEPH. He was pre-disposed to recurrent pulmonary embolism due to protein C deficiency. He underwent a left pulmonary endarterectomy (PEA) 4 years ago in 2014 and was given Sildenafil for residual pulmonary hypertension post-PEA but had defaulted on medications. Physical examination revealed the patient to be febrile at 39.7 degrees Celsius, with bi-basal crepitations and swollen calves bilaterally. Computed tomography of the pulmonary artery also revealed a new small sub-segmental right pulmonary artery embolus.

The patient soon desaturated and became hypotensive. He was transferred to the intensive care unit where he was mechanically ventilated and started on intravenous nor-adrenaline. He was initially commenced on nebulised Iloprost and Sildenafil for pulmonary hypertension. Intravenous antibiotics was also commenced for presumed sepsis. Subsequently, Sildenafil was switched to Riociguat and Macitentan, but there was difficulty weaning nebulised Iloprost. Given the difficulty with weaning of Iloprost, we decided to perform a semi-urgent balloon pulmonary angioplasty (BPA). He underwent a total of two sessions of BPA during his stay in the intensive care (Figure 1). The patient was successfully weaned off nebulized Iloprost after the second session of BPA. Post-BPA right heart catheterization was performed which showed improvement in pulmonary hemodynamics.

Discussion

Acute decompensated pulmonary hypertension presents a therapeutic challenge in the intensive care unit and has been associated with a high mortality. Timely hemodynamic...
optimization is key, and this entails right ventricular afterload reduction with targeted pulmonary hypertension therapy. To our knowledge, this is the first description of using a unique triple therapy combination to salvage an unstable patient from acute on chronic thromboembolic pulmonary hypertension; a combination of new generation oral agents Macitentan and Riociguat with BPA intervention.

The possibility of urgent surgical PEA must be considered carefully in CTEPH, especially when the patient is in cardiopulmonary failure. Medical treatment and/or BPA are considered if the surgical risk is prohibitive, or if the patient is deemed inopera bile because of surgical inaccessibility with distal thrombi. BPA involves using undersized balloons to break intraluminal webs and bands, and studies have found BPA to improve pulmonary hemodynamics.4

Previously, medical treatment options for CTEPH was limited. However, in recent years two newer generation drugs, Riociguat (guanylate cyclase stimulator) and Macitentan (endothelin receptor antagonist) have provided new medical treatment options for CTEPH.5,6 However, the use of Riociguat and Macitentan has not been well established in the intensive care setting to help a deteriorating, inoperable patient with CTEPH.

In the management of our patient, we administered broad-spectrum antibiotics to address pulmonary infection, as sepsis was the likely trigger for severe decompensation. Our patient was subsequently initiated on the novel use of both Macitentan and Riociguat as medical therapy, followed by BPA rescue for decompensated pulmonary hypertension from CTEPH. This combination therapy was remarkably efficacious in improving his haemodynamics, and he was able to leave the intensive care after 15 days. To our knowledge this is the first case describing the unique combination of Riociguat, Macitentan and BPA in successful treatment in a deteriorating patient with acute on chronic thromboembolic pulmonary hypertension.

Consent
Written consent was obtained from the patient prior to writing this article.

Conflict of interest: None declared.

References

Figure 1. (a) Balloon pulmonary angioplasty of the A8 branch. (b) Post intervention angiography showing recanalization of the A10 branch of pulmonary artery.