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CASE REPORT

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Nonsurgical treatment of a patient with decompensated right ventricular failure due to chronic thromboembolic pulmonary hypertension with proximal clot location—A case report

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Abstract

Chronic thromboembolic pulmonary hypertension (CTEPH) is a disease resulting from impaired patency of the pulmonary arteries by a clot, and the treatment method of choice is pulmonary endarterectomy (PEA). In inoperable patients, balloon pulmonary angioplasty (BPA) is recommended, but we need to implement pharmacological bridge therapy to BPA in some cases. We report a case of a 38-year-old male diagnosed with CTEPH, disqualified from PEA due to comorbidity, who developed right ventricular (RV) failure. The case shows a complex pharmacological treatment method that can be successfully used as an effective bridge therapy to BPA in patients with CTEPH and severe RV dysfunction, disqualified from surgery.

KEYWORDS

chronic thromboembolic pulmonary hypertension, right ventricular failure

INTRODUCTION

Pulmonary hypertension (PH) is defined as the elevation of mean pulmonary artery pressure (mPAP) over 20 mmHg at rest.[1] Chronic thromboembolic pulmonary hypertension (CTEPH) is a type of PH, which in almost 75% is the derivative of pulmonary embolism (PE). The pathophysiology of CTEPH is complex—mechanical obstruction of pulmonary arteries coexists with micro-angiopathy due to increased blood flow through vessels unaffected by clot.[2]

In patients with thrombotic changes accessible for surgery, the first-line treatment method is pulmonary endarterectomy (PEA).[3] In inoperable patients or subjects with persistent PH after PEA, the recommended therapy is balloon pulmonary angioplasty (BPA).[1] It has been proved that in technically operable patients who are disqualified from PEA due to reasons other than clot surgical accessibility, BPA provides significant hemodynamic and clinical improvement.[4] Irrespective of the treatment method, right ventricular (RV) failure correlates with poor clinical outcomes, and RV ejection

fraction is a strong predictor of adverse events in all CTEPH patients.[5]

CASE PRESENTATION

A 38-year-old man with a history of PE 12 months ago was admitted to the Heart Disease Center in Szczecin due to decompensated RV failure (RVF).

Four months ago, the patient was treated in the pulmonology department due to empyema of the right pleural cavity. The patient was disqualified from the pleural decortication procedure. The right lung did not fully expand, which resulted in restrictive ventilation disorders, with the diffusing capacity of the lungs for carbon monoxide reduced to 36% of the predicted value.

On admission to our Centre, the patient was in general severe condition, with hypoxemia and tachypnoe. Computed tomography angiography (CTA) visualized proximal clot location (Figure 1a). According to transthoracic heart echocardiography (TTE) we diagnosed the high probability of PH with a significant decrease in RV contractility (Figure 1b).

After diuretics therapy, we performed the right heart catheterization and pulmonary arteriography (PAr), confirming the diagnosis of CTEPH—the hemodynamic values are presented in Table 1. PAr revealed that in the right pulmonary artery (PA), only segmental arteries eight, nine, and six were permeable. Also, the permeability of the subsegmental arteries of the left PA was disabled (Figure 1c,d).

Due to severe ventilation disorders, CTEPH–TEAM disqualified the patient from PEA and qualified him for BPA and pharmacotherapy with riociguat, oral anticoagulants, and diuretics. Treatment was started with a daily dose of 3 mg of riociguat with the assumption of uptitration to achieve the maximum reduction in mPAP before BPA implementation. However, 1 month after the initiation of therapy, the patient was readmitted to the Centre due to decompensation of RVF.

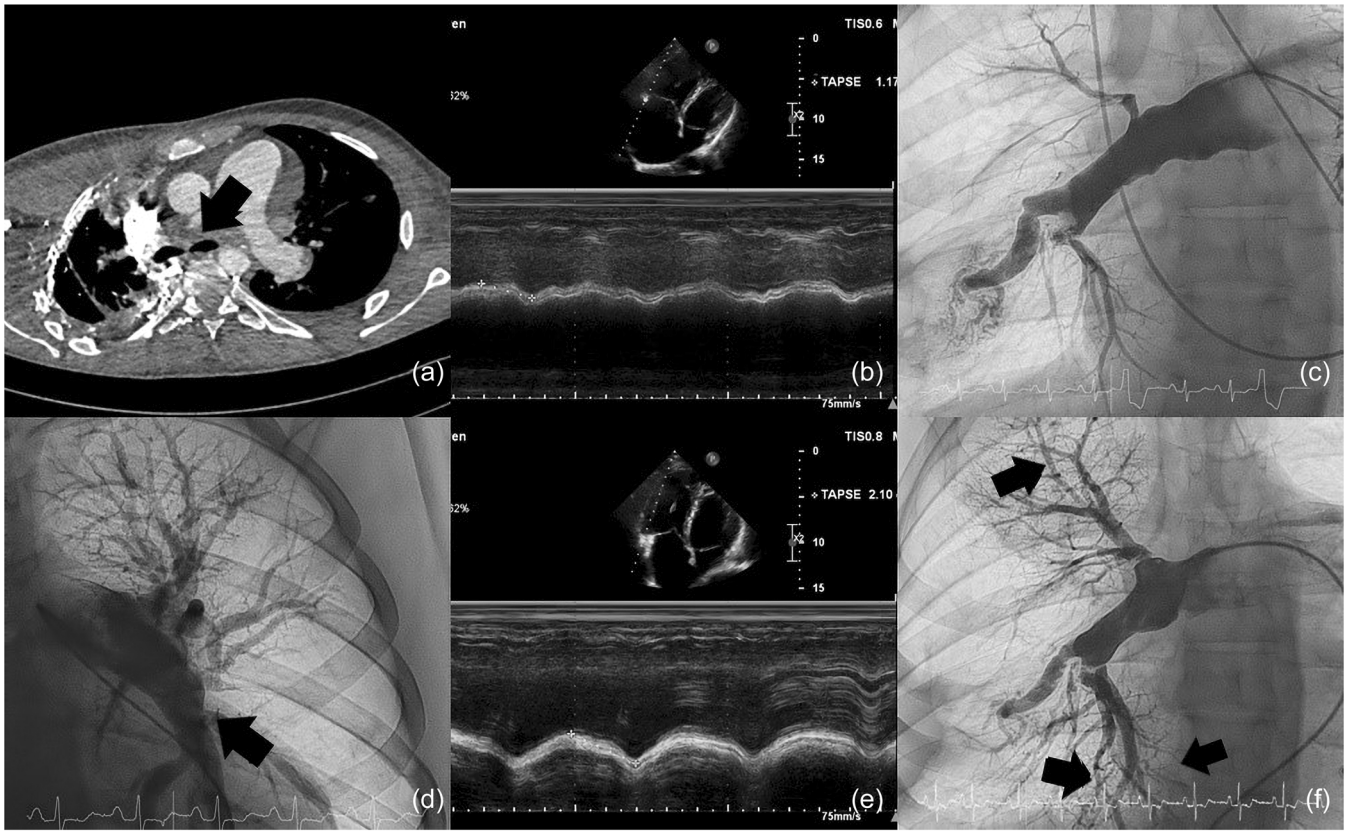

FIGURE 1 (a) Computed tomography angiography—the arrow indicates fibrotic changes in proximal part of the right pulmonary artery (RPA). (b) Transthoracic heart ultrasonography shows impaired right ventricular contractility with the decerase in tricuspid annular plane systolic excursion (TAPSE). (c) Baseline pulmonary arteriography of the RPA shows massive perfusion defects (posteroanterior view). (d) Baseline pulmonary arteriography of the left pulmonary artery (LPA) the arrow shows honeycomb-like perfusion defect in the segmental branch of LPA (posteroanterior view). (e) Transthoracic heart ultrasonography after levosimendan infusion shows significant increase in TAPSE. (f) Pulmonary arteriography of RPA after the third balloon pulmonary angioplasty session-arrows show significant perfusion improvement in superior and inferior lobes (posteroanterior view).

TABLE 1 The number, type, location of lesions, and results of BPA sessions in our patient.

BPA session	I	II	III	IV	V	VI
The number of treated PA branches	3	3	3	3	5	3
Treated PA branches and type of lesions	Left PA: subsegmental branches: A8a: web-like lesion A8b: web-like lesion A5a: web-like lesion	Left PA: subsegmental branches: A3a: web-like lesion A3b: web-like lesion Right PA: segmental branch: R8: web-like lesion	Right PA: segmental branches: A1: web-like lesion A9: web-like lesion A10: web-like lesion	Right PA: segmental branches: A1: web-like lesion A9: web-like lesion A10: web-like lesion	Right PA: segmental branches: A1: ring-like lesion A2: web-like lesion A3: web-like lesion A8: ring-like lesion A10: ring-like lesion	Right PA: segmental branch: A9: ring-like lesion LEFT PA: subsegmental branches: A6a: web-like lesion A6b: web-like lesion
Result	Complete perfusion	Complete perfusion	Complete perfusion	Complete perfusion	Complete perfusion	Complete perfusion

Abbreviations: BPA, balloon pulmonary angioplasty; Left PA, left pulmonary artery; Right PA, right pulmonary artery.

On admission, the patient was in a severe general condition with shortness of breath at rest. According to CTA, we excluded the recurrence of PE. The TTE picture was comparable to the baseline one. It was not possible to perform rescue BPA without general anesthesia, but the procedure was at high risk of death.

Initially, we implemented an infusion of dobutamine and furosemide and maintained riociguat at the achieved daily dose of 4.5 mg, but due to the lack of clinical improvement, the dobutamine infusion was discontinued after 48 h.

We decided to implement levosimendan as a 24-h infusion.

TTE performed 3 days after the end of levosimendan infusion showed a significant improvement in RV systolic function (Figure 1e). After achieving stabilization of the patient clinical condition, the first BPA session was performed. After the procedure, the patient was discharged in good general condition.

During the next 12 months, we performed six BPA sessions. The number, localization, and type of treated lesions during each BPA session are presented in Table 1. We did not observe any complications of the procedure.

The riociguat therapy was maintained. The hemodynamic parameters improved markedly (see Table 2). Since using levosimendan and initiating BPA, no further exacerbations of heart failure have been observed, and the patient is now in WHO functional class II and achieves 570 meters in 6 min walk test.

DISCUSSION

Our case report demonstrates both the difficulties in decision-making, as well as the successful complex treatment in inoperable patient with decompensated RVF due to CTEPH and proximal clot location in PA.

PEA is the first-line treatment method in CTEPH patients, but about one-third of this population is disqualified from surgery.[2,6] Inoperability may result from the surgical accessibility of clot and comorbid conditions, which increase the risk of anesthesia and surgical procedures. Inoperable patients with CTEPH and proximal changes in PA are at increased risk of RVF development. It has been proved that subjects with proximal clot localization have higher mPAP and pulmonary vascular resistance.[7] On the other hand, a retrospective study published in 2020 showed that thromboembolism (TE) is the second most common cause of RVF, and the 1- and 5-year survival rate in patients with TE and RVF is 71% and 49%, respectively.[8]

In our patient, the BPA procedure was not possible due to dyspnea at rest, and the risk of anesthesia was too high.

TABLE 2 Hemodynamic changes at the baseline and during treatment.

Hemodynamics	Baseline	Seven days after levosimendan infusion, before the I session of BPA	After the I session of BPA	After the VI session of BPA
mPAP (mmHg)	53	50	48	38
sPAP (mmHg)	95	82	80	70
PVR (Wood Units)	10.47	9.78	10.25	6.59
CO (L/min)	3.82	4.09	3.9	4.25
CI (L/min/m^2)	1.99	2.13	2.03	2.21
RAP (mmHg)	14	8	8	4
PAWP (mmHg)	13	10	8	10

Abbreviations: BPA, balloon pulmonary angioplasty; CI, cardiac index; CO, cardiac output; mPAP, mean pulmonary artery pressure; PAWP, pulmonary artery wedge pressure; PVR, pulmonary vascular resistance; RAP, right atrial pressure; sPAP, systolic pulmonary artery pressure.

Therefore, we decided to implement levosimendan—the inotropic and vasodilating agent, which has proven effectiveness in reducing the mean and systolic PA pressure in patients with heart failure.[9] We observed that levosimendan is an effective medicine that improves RV contractility and can be used safely with CTEPH-targeted drugs and diuretics.

A study published in 2019 showed that treprostinil administered subcutaneously significantly improves physical performance in patients with inoperable CTEPH and persistent PH after PEA.[10] In Poland, however, this therapy is available only when standard pharmacological and interventional treatments do not result in clinical improvement. Thus, we could not qualify our patient for treprostinil therapy.

In conclusion, comorbidity is a common reason for disqualification from PEA in patients with CTEPH, and the subpopulation with RVF presents a particular therapeutic challenge. The selection of appropriate pharmacotherapy, including adequate inotropic agents, such as levosimendan, can significantly improve the clinical condition of patients and be the bridge therapy to the BPA procedure.

AUTHOR CONTRIBUTIONS

Marta Braksator, Magdalena Jachymek, and Amena Rahmani collected the data, wrote the preliminary manuscript, and created the table. Marta Braksator, Katarzyna Widecka, and Maciej Lewandowski created the picture. Maciej Lewandowski and Łukasz Jodko revised the manuscript. Małgorzata Peregud-Pogorzelska supervised and critically revised the manuscript. All co-authors have made a substantial contribution to the design, data collection and analysis of the research and the drafting of the manuscript and have reviewed and accepted the contents of the manuscript before its submission.

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CONFLICT OF INTEREST STATEMENT

The authors declare no conflict of interest.

DATA AVAILABILITY STATEMENT

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

ETHICS STATEMENT

Informed consent was obtained from patient for publication.

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