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Resolution of Cardiopulmonary Lesions After Intervention for Rapidly Progressive POEMS-Related Pulmonary Hypertension: A Case Report

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Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Patient: Male, 30-year-old
Final Diagnosis: POEMS syndrome
Symptoms: Dyspnea
Clinical Procedure: —
Specialty: Family Medicine
Objective: Rare disease

Background: Pulmonary hypertension is a severe complication of Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal protein, and Skin changes (POEMS) syndrome. It can cause severe right heart failure and even death. Vascular endothelial growth factor modulates angiogenesis and vascular hyperpermeability, which is the key mechanism behind this complication. POEMS-related pulmonary hypertension is often misdiagnosed due to its nonspecific manifestations. The optimal regimen remains undefined.

Case Report: A 30-year-old man presented with a 4-month history of dyspnea and bilateral lower extremity weakness. The patient had manifestations of volume overload and received diuretic therapy. Despite this treatment, transthoracic echocardiography showed a rapid rise in estimated pulmonary artery systolic pressure from 55 mm Hg to 109 mm Hg over a 2-month interval. Meanwhile, it also demonstrated thickening of the interventricular septum and left ventricular posterior wall, together with obvious dilation of multiple cardiac chambers. Pulmonary function tests showed severe mixed ventilatory impairment and decreased diffusing capacity of the lung for carbon monoxide. Further evaluations demonstrated more extensive multisystem involvement. Immunofixation electrophoresis and vascular endothelial growth factor testing were performed, confirming the diagnosis of POEMS syndrome. The patient initially received a lenalidomide and dexamethasone regimen and subsequently underwent autologous hematopoietic stem cell transplantation, resulting in significant symptom improvement. At the 10-month follow-up, he remained in sustained symptomatic remission, and relevant monitoring indicators remained within normal ranges.

Conclusions: Rapidly progressive pulmonary hypertension is a severe yet potentially reversible complication in POEMS syndrome. Lenalidomide and dexamethasone followed by autologous stem cell transplantation can markedly slow disease progression. Our present case illustrates rapid progression and favorable post-treatment improvement in POEMS-associated rapidly progressive pulmonary hypertension.

Keywords: POEMS Syndrome • Pulmonary Arterial Hypertension • Heart Failure
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Introduction

Polyneuropathy, Organomegaly, Endocrinopathy, Monoclonal protein, and Skin changes (POEMS) syndrome is a rare paraneoplastic disorder caused by clonal plasma cell proliferation. It is also known as Crow–Fukase syndrome (Takatsuki syndrome) in the medical literature. The acronym POEMS was first coined by Bardwick et al in 1980 to encompass the core manifestations [1]. Notably, diagnosis does not require all 5 classic POEMS criteria. Polyneuropathy and monoclonal gammopathy serve as the 2 indispensable major diagnostic criteria [2]. Progressive polyneuropathy is one of the most disabling complications. Since POEMS can mimic neuromuscular diseases such as amyotrophic lateral sclerosis, multiple sclerosis, Guillain-Barré syndrome, transverse myelitis, and vitamin D deficiency, patients presenting with predominant limb weakness are prone to misdiagnosis [3-5]. Besides peripheral polyneuropathy, common manifestations include edema, splenomegaly, and skin hyperpigmentation. Due to its rarity and variable manifestations, POEMS syndrome is often diagnosed late [4]. Without targeted treatment, patients will experience progressive systemic manifestations including osteosclerotic bone lesions, pulmonary hypertension (PH), and extravascular fluid overload [6].

The overall incidence and complete pathogenetic mechanisms of POEMS syndrome remain poorly defined. Elevated vascular endothelial growth factor (VEGF) is closely associated with disease activity and contributes to the pathogenesis of POEMS syndrome by regulating angiogenesis and vascular hyperpermeability [7]. A serum VEGF threshold of > 200 pg/mL has 95% sensitivity and 68% specificity for diagnosing POEMS syndrome [8]. As a key complication, PH occurs in 27% to 48% of POEMS patients. It is more common in those with extravascular volume overload, suggesting a common vascular mechanism for the 2 conditions [9-12]. Accumulating clinical data show cardiac involvement in POEMS syndrome. Tanaka et al reported a case of a patient with severe PH, prominent right heart dilation, and diminished right ventricular systolic function [13]. Similar findings have also been documented in another published report [14]. Wang et al documented POEMS-related left ventricular hypertrophy and myocardial dysfunction without concomitant right heart involvement [15]. These distinct findings illustrate that multiple mechanisms contribute to POEMS-induced cardiac injury. Notably, unlike the irreversible cardiac injury caused by light-chain amyloidosis, POEMS-related cardiac lesions are reversible [16]. Etiological therapy can significantly reverse myocardial injury and PH in POEMS patients [13,15,17].

Herein, we report a case of POEMS syndrome presenting with a 4-month history of progressive bilateral limb weakness and exertional dyspnea. This case serves as an illustrative example of rapidly progressive pulmonary hypertension secondary to POEMS syndrome.

Case Report

A 30-year-old man presented with a 4-month history of bilateral lower extremity weakness and exertional dyspnea. At admission, he exhibited NYHA II–III exertional dyspnea, with an untreated blood pressure of 147/90 mm Hg. Physical examination revealed symmetric and uniform hyperpigmentation involving the face, both hands, and bilateral lower extremities. Cardiopulmonary and abdominal examination showed decreased breath sounds over both lower lung fields, cardiomegaly, positive shifting dullness, and moderate bilateral lowerextremity edema. Neurological assessment demonstrated grade 4 muscle strength bilaterally in the lowerlimb proximal and distal muscle groups, hypoesthesia to pain and temperature sensation in both lower limbs, and diminished ankle reflexes (**Figure 1**). The patient was a smoker with no history of tuberculosis, chronic lung or heart disease, or other systemic comorbidities. Chest computed tomography (CT) detected small-to-moderate pleural and pericardial effusions. Transthoracic echocardiography (TTE) showed a pulmonary artery systolic pressure (PASP) of 55 mm Hg and a left ventricular ejection fraction (LVEF) of 62%. Electromyography demonstrated decreased motor and sensory nerve conduction velocities across all extremities.

Imaging confirmed multiple serous effusions. Etiologic workup first excluded typical secondary etiologies: negative antinuclear antibodies and normal C3/C4 complement levels ruled out connective tissue disorders. Tumor biomarkers and contrast-enhanced thoracoabdominal CT yielded no definitive evidence of malignancy (**Figure 2A-2D**). TTE showed absent pericardial thickening with preserved left ventricular ejection fraction, precluding constrictive pericarditis and heart failure with reduced ejection fraction. Finally, medical record review, liver biochemistry, and urinalysis eliminated cirrhosis and nephrotic syndrome from the differential diagnosis.

The patient had clinical manifestations of fluid overload and received diuretic therapy. However, TTE showed a rapid rise in PASP from 55 mm Hg to 109 mm Hg over a 2-month interval (**Figure 3**). The examination also revealed thickening of the interventricular septum (IVS) and left ventricular posterior wall (LVPW), as well as dilation of the left atrium, right atrium, and left ventricle, with LVEF remaining normal (**Figure 4**). Such rapidly progressive cardiac impairment indicated a poor prognosis. Before admission, computed tomographic pulmonary angiography (CTPA) revealed no evidence of chronic thromboembolic disease. Ventilation-perfusion scintigraphy was not performed; therefore, chronic thromboembolic pulmonary hypertension

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Figure 1. Changes in patients' physical signs and appearance. A1-C1: Facial pad atrophy (A1), sclerodermic changes (B1) and diffuse hyperpigmentation (C1). A2-C2: Facial pad atrophy, sclerodermic change, and diffuse hyperpigmentation improved after treatment.



Figure 2. Imaging findings suggest polyserositis effusion and hepatosplenomegaly. (A) A small amount of bilateral pleural effusion. (B) moderate pericardial effusion. (C) pelvic and abdominal effusion. (D) hepatosplenomegaly.

could not be completely excluded based on imaging. Arterial blood gas analysis showed no hypoxemia (PaO2: 93 mm Hg on room air), and serological tests excluded HIV infection. Detailed medical history inquiry ruled out obstructive sleep apnea, portal hypertension, schistosomiasis, connective tissue disease (CTD), and exposure to drugs or toxins known to induce pulmonary hypertension. Coronary angiography further excluded significant coronary artery stenosis. Pulmonary function testing identified severe restrictive-predominant mixed ventilatory dysfunction and decreased diffusing capacity of the lung for carbon monoxide (DLCO). These functional abnormalities occurred in a patient without pre-existing respiratory disease, which points to possible pulmonary injury mediated by systemic disease. Echocardiography revealed no significant valvular heart disease or congenital heart disease, with normal systolic and diastolic cardiac function. Nevertheless, elevated pro-BNP levels (2872 pg/ml) suggested that left-heart-associated PH may have partly contributed to disease progression. Collectively, these findings strongly suggested that other etiologies or multifactorial mechanisms were involved in the development of PH, and the patient's systemic manifestations further indicated an underlying plasma cell disorder.

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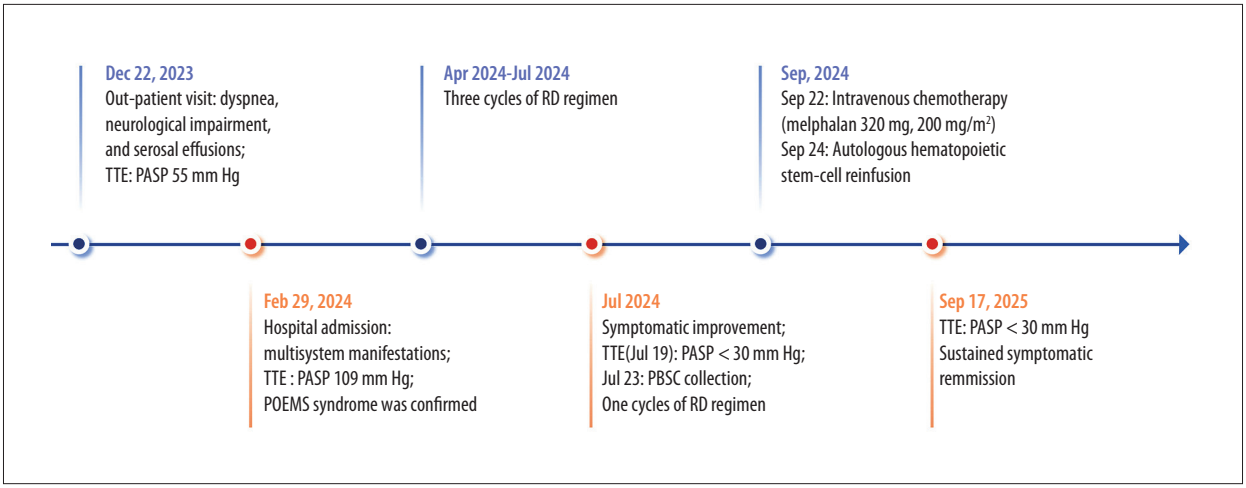


Figure 3. Timeline of clinical course and management. PASP, pulmonary artery systolic pressure; TTE, Transthoracic echocardiography; RD, lenalidomide-dexamethasone; PBSC, peripheral blood stem cell.

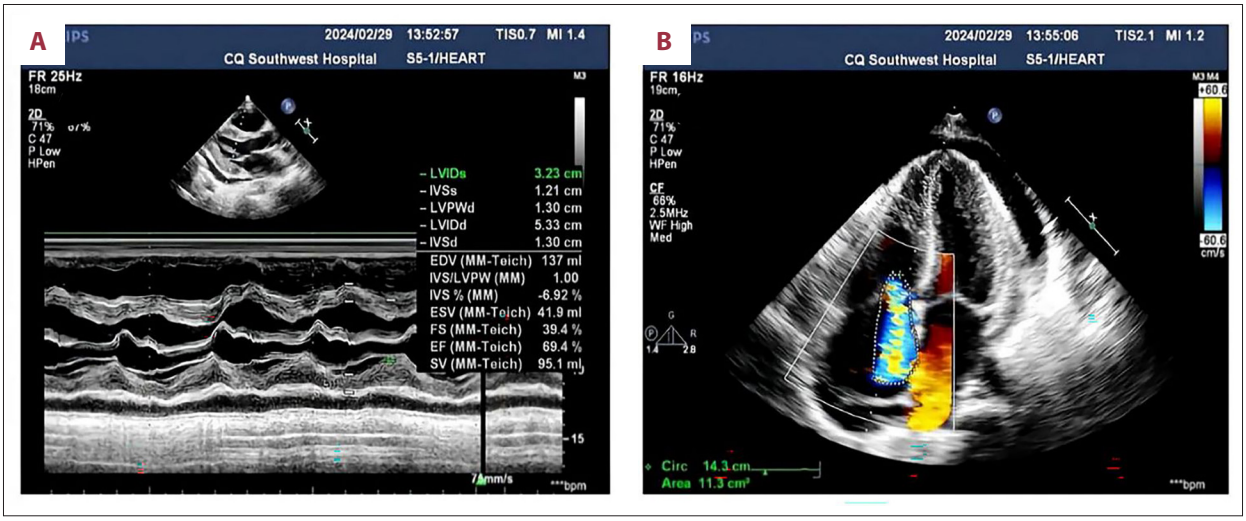


Figure 4. Echocardiographic assessment of cardiac structure, function, and pulmonary vascular changes. (**A**) Echocardiographic images (M-mode) show EF = 69%; LA (40 mm) and RA (45 mm) are enlarged; RV (24 mm) is moderately enlarged. IVS (12.1 mm) and LVPW (12.9 mm) are slightly thickened. (**B**) PASP is significantly elevated (109 mm Hg). EF, ejection fraction; LA, left atrium; RA, right atrium; RV, right ventricle; IVS, interventricular septum; LVPW, left ventricular posterior wall; PASP, pulmonary artery systolic pressure.

We next performed targeted examinations to confirm POEMS syndrome. The patient fulfilled the diagnostic criteria, including 2 mandatory major criteria, 2 additional major criteria, and 4 minor criteria (**Table 1**). He initially received the lenalidomide and dexamethasone (RD) regimen in our hospital's Hematology Department. The RD regimen was administered in 28-day cycles: lenalidomide 25 mg once daily on days 1 to 21, and dexamethasone 40 mg once weekly (days 1, 8, 15, and 22), followed by a 7-day rest interval before the next cycle. After 4 cycles of the RD regimen, hematopoietic stem cell collection was completed on July 23, followed by autologous stem cell transplantation (ASCT) on September 24 at Peking Union Medical College Hospital (**Figure 3**). All therapies were initiated after assessment for infection, cardiopulmonary function, hepatic and renal function, and coagulation status.

Following targeted therapy, TTE-estimated PASP improved markedly, with complete reversal of cardiac structural abnormalities. At the 10-month follow-up, these improvements persisted (**Table 2**). Therapeutic effect assessment revealed obvious declines in the proportion of peripheral blood plasma cells and serum VEGF concentrations. Neurological function was mostly restored, with only mild foot drop left as a residual manifestation, and organomegaly improved substantially (**Table 3**). The patient achieved substantial symptomatic relief, with restored independent ambulation, improved daily

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Table 1. Diagnostic criteria and patient's conditions.

Diagnostic criteria	Actual clinical features
Mandatory major criteria (2 required)	1. **Polyneuropathy** (bilateral neurogenic damage) 2. **Monoclonal plasma cell proliferative disorder** (SPE: M protein in the γ region; IPE: IgAλ monoclonal protein; BMB: 7% plasma cells)
Major criteria (at least 1 required)	1. Castleman disease 2. **Sclerotic bone lesions** (increased skeletal metabolism in red marrow regions) 3. **Elevated VEGF* level** (VEGF: 460.7 pg/ml, normal < 160 pg/ml)
Minor criteria (at least 1 required)	1. **Organomegaly** (Hepatosplenomegaly, multiple lymphadenopathy) 2. **Extravascular fluid accumulation** (pleural effusion, pericardial effusion, ascites, bilateral lower extremity edema) 3. **Endocrine abnormalities** (testosterone, prolactin, FT3, TSH) 4. **Skin changes** (cutaneous hyperpigmentation) 5. Papilledema 6. Thrombocytosis, polycythemia
Other signs and symptoms	Additional symptoms: pulmonary hypertension, restrictive lung disease and weight loss

Bold text denotes the diagnostic criteria met by the patient. Abbreviations: IFE, immunofixation electrophoresis; SPE, serum protein electrophoresis; BMB, bone marrow biopsy; VEGF, vascular endothelial growth factor.

Table 2. Changes in pre- and post-treatment echocardiographic parameters.

Parameters	Dec 22, 2023	Feb 29, 2024	Jul 19, 2024	Sep 17, 2025
PASP (mm Hg)	55	109 (severe)	Normal (< 30)	Normal (< 30)
RAD (mm)	41	45	Not measured	36
RVEDD (mm)	23	31	25	20
TR Severity	Mild	Severe	Mild	None
Pericardial effusion	Small-to-moderate	Small-to-moderate	Small	None
IVS (mm)	10.8	12.1	9	10.3
LAD (mm)	43	40	33	35
LVD (mm)	56	53	47	50
EF (%)	62	69	64	68
E/E' ratio	6	Not performed	> 10	10

The patient received autologous stem-cell transplantation in September 2024. Abbreviations: PASP, pulmonary artery systolic pressure; RAD, right atrial diameter; RVEDD, right ventricular end-diastolic diameter; TR, tricuspid regurgitation; IVS, interventricular septum; LAD, left atrial diameter; LVD, left ventricular diameter; EF, ejection fraction; E/e' ratio, ratio of early transmitral inflow velocity E to early mitral annular tissue-Doppler velocity e'.

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living ability, and complete resolution of exertional dyspnea and limb weakness (**Figure 1B**). Blood pressure stabilized and pulmonary function parameters improved markedly (**Table 4**).

Discussion

In routine etiological workups for PH, clinicians primarily screen for cardiopulmonary and connective tissue diseases, while POEMS syndrome is rarely included in standard screening panels. Accumulating clinical evidence has confirmed that POEMS syndrome can induce severe cardiopulmonary damage, potentially leading to acute respiratory failure and hemodynamic instability [3,7,11]. Several cohort studies focusing on pulmonary manifestations in POEMS syndrome have reported dyspnea in 28% to 34% of affected patients [8], among whom 56.4% present with pleural effusion, 62.8% with mediastinal or hilar lymphadenopathy, and 35.5% with PH [11]. The reported prevalence of PH in POEMS syndrome varies across studies, and the routine use of TTE for PH screening may overestimate its incidence [18]. Notably, PH serves as an independent negative prognostic indicator in POEMS syndrome and significantly impairs

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Table 3. Therapeutic effect assessment: Multi-dimensional follow-up outcomes.

Assessment Items	Before treatment	After treatment	10-month Follow-Up
IFE	IgA λ type positive	IgA λ type positive	Negative
Plasma cell ratio	7%	Not tested	2%
Serum VEGF	460.7 pg/ml	61.97 pg/ml	69 pg/ml
ONLS Score	0 + 6 = 6	1 + 3 = 4	0 + 2 = 2
Organomegaly	Hepatomegaly, splenomegaly, lymphadenopathy	Not tested	Reduced volume
Testosterone	0.83 ng/ml	Not tested	2.68 ng/ml
Thyroid function	FT3 2.24 mmol/L; TSH 7.14 uIU/ml	Not tested	FT3 5.06 mmol/L; TSH 1.96 uIU/ml
Hyperpigmentation	Mild	Attenuated	Resolved
NT-proBNP	2872 pg/ml	Not performed	Not performed

Normal reference ranges: Testosterone: 1.75-7.81 ng/ml; FT3: 3.1-6.8 mmol/L; TSH: 0.27-4.2 uIU/ml; VEGF: < 160 pg/ml.
Abbreviations: FT3, free triiodothyronine; IFE, immunofixation electrophoresis; ONLS, Overall Neuropathy Limitations Scale; TSH, thyroid-stimulating hormone; VEGF, vascular endothelial growth factor; NT-proBNP, N-terminal pro-B-type natriuretic peptide.

Table 4. Therapeutic effect assessment: Follow-up outcomes of pulmonary function and blood pressure.

Assessment items	Baseline (March 2024)	10-month follow-Up
Pulmonary function conclusion	Severe restrictive-dominant mixed ventilatory dysfunction	Normal
FVC% pred	74.2%	86.18%
FEV_1/FVC	89.79%	91.57%
FEV_1% pred	45.1%	93.76%
MVV (L/min)	142.82	156.35
DLCO (mmol/min/kPa)	4.32	8.65
Blood Pressure (mm Hg)	147/90	136/85

Abbreviations: FEV_1, forced expiratory volume in one second; FVC, forced vital capacity; MVV, maximum voluntary ventilation; % pred, percentage of predicted value; DLCO, diffusing capacity of the lung for carbon monoxide

overall survival (OS), with a median OS of only 54 months in PH-complicated patients [19]. A single-center prognostic study from Peking Union Medical College Hospital identified advanced age (over 50 years), PH, pleural effusion, and reduced estimated glomerular filtration rate as independent risk factors for decreased OS [17]. Taken together, these results suggest that PH is a common and clinically important complication of POEMS syndrome, even though it is not included in the official diagnostic criteria. Enhanced clinical awareness of the association between POEMS syndrome and PH is essential.

Our patient presented with acutely progressive dyspnea and rapidly worsening PASP estimated by TTE, indicating critical disease progression. Right ventricular dilation and severe tricuspid regurgitation pointed to a high risk of cor pulmonale. Meanwhile, concomitant pleural effusion, restrictive ventilatory impairment, and decreased DLCO indicated a poor prognosis. POEMS syndrome can induce systemic microvascular endothelial injury, interstitial edema, and subtle pulmonary interstitial lesions, which can reduce pulmonary diffusion capacity. In addition, systemic fluid overload and serous cavity effusions can restrict thoracic mobility and further lead to restrictive ventilatory impairment. Previous studies have documented fatal outcomes when intervention was delayed in a similar case [3].

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Notably, IVS thickening was detected in our patient. With no history of hypertension, valvular lesions, or preexisting cardiac illnesses, the cardiac structural abnormalities were presumed to be associated with POEMS-syndrome-induced myocardial injury. Previous reports have also verified POEMS-related myocardial damage, which mainly manifested as LV hypertrophy and regional myocardial motion abnormalities [15]. Takahashi et al further confirmed myocardial interstitial edema in POEMS patients via cardiac magnetic resonance imaging [20]. Distinct from irreversible myocardial hypertrophy induced by light-chain amyloidosis, POEMS-related cardiac injury is mainly caused by interstitial edema of cardiomyocytes, which renders the lesion reversible. Mechanistically, VEGF is a key pathogenic factor triggering PH and myocardial edema in POEMS syndrome, which makes it suitable for monitoring treatment response [21,22].

Stratification of disease involvement is essential for determining optimal regimens. Radiotherapy serves as first-line therapy for patients with limited bone lesions and no marrow infiltration, whereas those with diffuse skeletal lesions or marrow involvement require systemic treatment, with ASCT preferred for eligible candidates [19,23]. To date, no randomized controlled trials have established a standardized treatment for POEMS syndrome, and all strategies are based on retrospective case series. Systemic treatment includes alkylating agents combined with glucocorticoids, immunomodulatory drugs, and ASCT. Anti-VEGF agents like bevacizumab only confer limited clinical benefits and are inferior to therapies targeting the primary clonal plasma cell lesion [24]. A recent cohort study demonstrated that standardized systemic treatment achieves a 72% 6-year progression-free survival rate and an 89% 10-year OS rate [25].

This case reminds us to focus on the correlation between cardiopulmonary injury and POEMS syndrome. Targeted etiological screening should be performed in patients with unexplained PH accompanied by multisystem manifestations highly suggestive of POEMS syndrome. Follow-up results further confirm that severe multisystem damage caused by POEMS syndrome can be reversed. A major limitation of this report is the absence of confirmatory right heart catheterization (RHC) data. RHC was not performed because of the patient's rapid, unstable disease progression in the early phase and economic constraints. Accordingly, this case only offers therapeutic insights into PH diagnosed by TTE. Given the lack of RHC data, difficulties existed in the diagnosis and etiological differential diagnosis of PH. In particular, our patient presented with systemic volume overload, which can induce secondary cardiac insufficiency and may be important contributor to PH. However, after standardized diuretic treatment, the TTE-estimated PASP did not improve correspondingly, indicating that isolated volume overload could not fully explain the persistent PH. RHC is therefore recommended for clinically stable patients to confirm the PH diagnosis and establish accurate baseline hemodynamic parameters. Further large-scale, multi-center prospective studies are warranted to verify the generalizability of this treatment strategy and optimize standardized management protocols for POEMS-related cardiopulmonary complications.

Conclusions

POEMS syndrome-associated PH poses substantial diagnostic difficulties, as its nonspecific cardiopulmonary manifestations often delay diagnosis. High clinical suspicion and targeted etiological workup should be prioritized in patients with unexplained rapidly progressive PH accompanied by multisystem manifestations suggestive of POEMS syndrome.

Department and Institution Where Work Was Done

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Patient Permission

Written informed consent has been acquired from the patient for the publication of this case report, including clinical records and imaging materials. Any personal identifiers have been anonymized in the manuscript to maintain confidentiality.

Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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