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Systemic Sclerosis-Associated Pulmonary Hypertension: Three Cases with Distinct Hemodynamic Profiles

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Abstract | **Case Report**

Pulmonary arterial hypertension (PAH) is a devastating complication of systemic sclerosis (SSc) and is a leading contributor to mortality and morbidity. This series demonstrates the clinical and hemodynamic heterogeneity within PAH-SSc and the importance of personalized care. Herein, we describe three connective tissue disease (CTD) women with SSc and PAH at various times of their disease. Patient 1: isolated postcapillary PAH in context of left heart disease and atrial flutter; patient 2: combined PAH with interstitial lung disease and systemic hypertension; patient 3: precapillary PAH with heart failure with preserved ejection fraction (HFpEF) profile. All were verified by right heart catheterization and treated with specific therapies for PAH. This series highlights the heterogeneity of PAH in SSc and the need for early recognition, multimodal assessment and individualized treatment to enhance functional status and quality of life in these complex patients.

Keywords: Systemic Sclerosis, Pulmonary Arterial Hypertension, Right Heart Catheterization, Endothelin Receptor Antagonists, Phosphodiesterase-5 Inhibitors.

INTRODUCTION

Pulmonary arterial hypertension (PAH) is a life-threatening complication in systemic sclerosis (SSc) and one of the leading causes of morbidity and mortality [1]. It may be precapillary only, postcapillary only or mixed, and there is considerable hemodynamic heterogeneity which makes diagnosing and managing it difficult.

Here we describe three cases with SSc and PAH that occurred at different stages of the disease and are characterized by different clinical, echocardiographic and hemodynamic features. The former case had pulmonary veinous hypertension (PVH) due to isolated postcapillary PH due to left heart disease with repeated atrial flutter, the latter with combined PVH and interstitial lung disease and systemic hypertension and the rest had precapillary PH with preserved left ventricular ejection fraction and pulmonary nodules. All patients underwent right-heart catheterization for confirmation of diagnosis and received PAH-specific therapy (macitentan, tadalafil, sildenafil or iloprost). The outcome results demonstrated a meaningful functional recovery.

This series emphasizes the necessity of an individualized assessment and therapeutic strategy in PAH secondary to SSc.

CASE PRESENTATION

Case 1

We report a 44-year-old woman with a diagnosis of systemic sclerosis (SSc) according to ACR-EULAR criteria, that was made in 2017, with cutaneous, vascular, and pulmonary manifestations. She had established cardiovascular risk factors of obesity (BMI 38.3 kg/m²) and early menopause. She had no previous history of abortion, autoimmune diseases or toxic habits. She developed progressive exertional dyspnea (WHO-FC III), peripheral edema, and palpitations five years after the onset of her disease. Work-up was consistent with postcapillary PH, interstitial lung disease (ILD) without fibrosis, and recurrent atrial flutter requiring 2 ablations.

On clinical examination, the patient was stable with scleroderma facies and malleolar/digital ulcers (figure 1). The Electrocardiogram (ECG) showed sinus rhythm (50 bpm), biatrial enlargement, complete Right Bundle Branch Block (RBBB), long QTc interval (569

ms), and low voltage. Imaging and echocardiography demonstrated severe Right Ventricle (RV) dilation and dysfunction, tricuspid regurgitation (TR), and maintained Left Ventricle (LV) function. Cardiac Magnetic Resonance Imaging (MRI) revealed a Right Ventricular Ejection Fraction (RVEF) of 14% with right atrial enlargment. Right heart catheterization (RHC) confirmed postcapillary pulmonary hypertension (mPAP 26 mmHg, Pulmonary Capillary Wedge Pressure PCWP 19 mmHg, Pulmonary Vascular Resistance PVR 1.53 WU). Following ablation, mPAP fell to 18 mmHg while PCWP remained high and cardiac index low (1.12 L/min/m²).

In view of the seriousness of the disease and the contradictory results, therapy with macitentan and intravenous iloprost was started. It was not possible to operate on tricuspid regurgitation. The patient was discharged at a 6-month follow-up with significant clinical improvement.

Figure 1: "a" and "b": ulceration on the pulp of the right second finger; "c": ulceration on the left third toe; "d": Scleroderma facies

Case 2

We present the case of a 71-year-old woman followed since 1997 for systemic sclerosis with cutaneous, cardiac, and pulmonary involvement. Her cardiovascular risk factors included long-standing hypertension treated with Angiotensin II Receptor Blocker (ARB) and hydrochlorothiazide, overweight (BMI 26.1 kg/m²), and postmenopausal status. Medical history included hypothyroidism and osteoporosis under treatment. She had no surgical history, miscarriages, autoimmune disease, or familial predisposition.

Over time, she developed exertional dyspnea World Health Organization Functional Class (WHO-FC II) and abrupt-onset palpitations. Evaluation revealed combined pre- and postcapillary pulmonary hypertension, frequent ventricular extrasystoles, and mild pulmonary fibrosis.

On examination, she was hemodynamically stable with elevated blood pressure (160/60 mmHg) and signs of right heart failure (jugular venous distension, hepatojugular reflux, minimal ascites). Chest auscultation revealed fine bilateral basal crackles. Her skin displayed typical scleroderma facies and sclerodactyly without telangiectasias or ulcers.

ECG showed sinus rhythm, complete RBBB, left axis deviation, and QTc of 390 ms. Chest X-ray revealed cardiomegaly and basal opacities. High-Resolution Computed Tomography (HRCT) confirmed basal honeycombing and ground-glass opacities (<10% fibrosis) (figure 2). Right heart catheterization revealed mPAP 25 mmHg, PCWP 16 mmHg, and PVR 3.25 WU.

She was treated with inhaled corticosteroids and long-acting bronchodilators for ILD, and macitentan with tadalafil (later switched to sildenafil). After 2 years, right heart failure signs resolved and 6-Minute Walk Distance (6 MWD) improved from 390 m to 415 m.

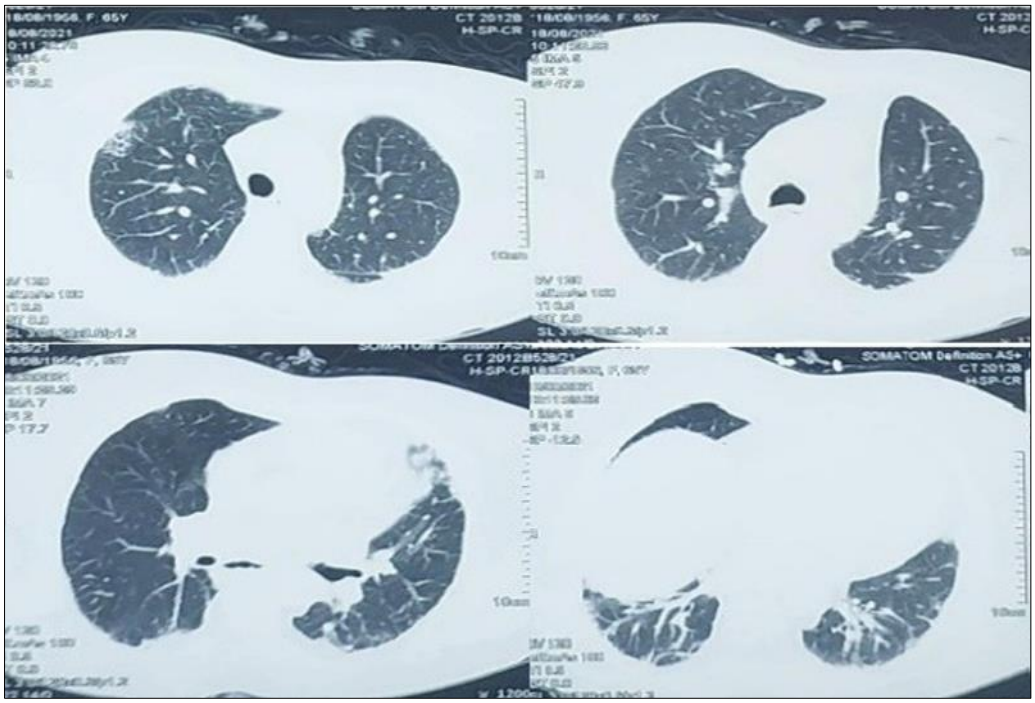

Figure 2: Chest CT scan showing pulmonary fibrosis

Case 3

We report a 59-year-old female patient, followed since 2012 for systemic sclerosis (SSc) diagnosed according to ACR-EULAR classification criteria, treated in dermatology for cutaneous, cardiovascular and gastrointestinal impairment. She had no modifiable cardiovascular risk factors and her menopausal status occurred when she was 47 years. Her past medical history was significant for a childhood history of pulmonary tuberculosis (cured), three years' history of hypothyroidism under treatment, and 2012 cholecystectomy. She had two vaginal deliveries and no toxic habits nor family history of autoimmune disease.

Tenth years from the onset of the disease, she experienced progressive exertional dyspnea (WHO-FC III) accompanied by moderate hemoptysis. The patient was stable on clinical examination, eupneic, with a body mass index of 22.8 kg/m², normal cardiac and pulmonary findings and scleroderma facies (figure 3) with telangiectasias, sclerodactyly and Raynaud phenomenon.

ECG showed sinus rhythm 80/min, right atrial enlargement, incomplete RBBB, and QTc interval 370ms. Transthoracic echocardiography (TTE) demonstrated preserved biventricular function with estimated sPAP of 41 mmHg. The HRCT revealed spiculated pulmonary nodule and interstitial changes in the right upper lobe.

Right heart catheterization revealed precapillary PH with a Heart Failure with preserved Ejection Fraction profile: mPAP 35 mmHg, PCWP 13 mmHg (increasing to 19 mmHg after volume challenge), PVR 3.47 WU, and a high cardiac index (>3.92 L/min/m²). Forced Vital Capacity (FVC) was decreased consistent with a restrictive ventilatory pattern. Immunology was positive for Antinuclear Antibodies (ANA) >1:1280, Anti-Centromere Antibodies (ACA), anti-Scl70 negative. NT-proBNP was also slightly increased (129.88 pg/mL).

The patient was started on macitentan and later on tadalafil in combination. She was in WHO-FC III at

12 months later with clinical improvement in dyspnea to WHO-FC II.

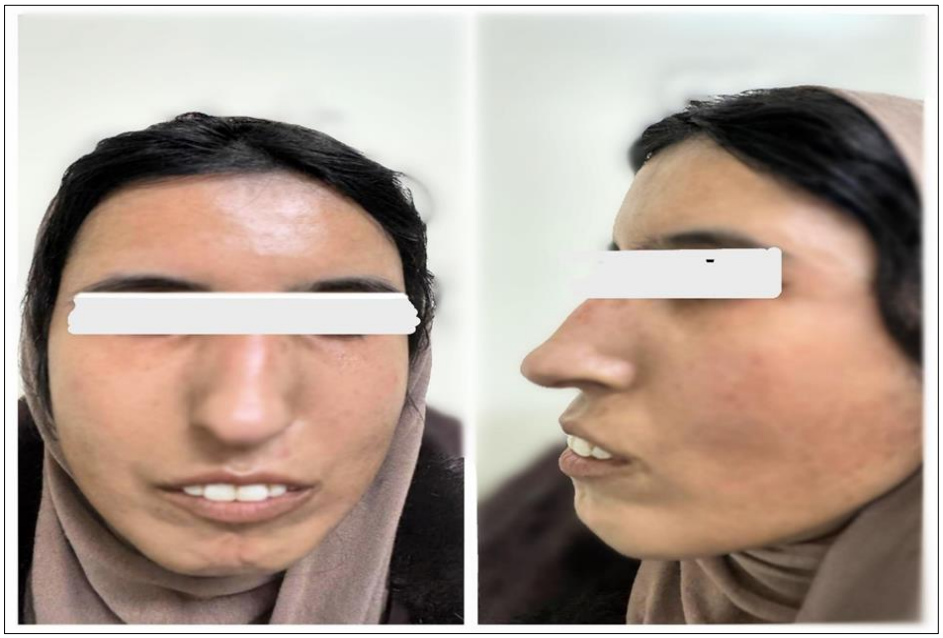

Figure 3: Scleroderma facies

DISCUSSION

Pulmonary artery hypertension (PAH) represents a significant cause of morbidity and mortality in systemic sclerosis (SSc), and occurs in about 8 to 12% of the patients with an insidious onset and a variable clinical course. The three cases reported in the article demonstrate the variable combinations of pathogenic factors and clinical presentation of PAH in the setting of SSc, highlighting the importance of individual diagnostic and treatment strategy [1].

PAH was mainly post-capillary in the first case, being secondary to rhythm-related left-left heart failure, with severe right ventricular (RV) deterioration and preserved left ventricular (LV) function. Although the hemodynamic results implied isolated postcapillary PH, the greater degree of RV failure and the discrepancy between invasive and non-invasive results supported the option to start dual PAH-targeted therapy (macitentan and iloprost). This case underscores the significance of combining multimodal assessment (ECG, cardiac MRI, echocardiography, and RHC) to guide therapy, in addition to purely categorization schemes [2, 3].

The second case demonstrated a more classical combined pre- and post-capillary PH, together with long-standing systemic hypertension and mild ILD. After 25 days of intolerance to macitentan-tadalafil the patient responded to sildenafil. By 2 years, her pulmonary artery pressure had normalized and symptoms had improved. This case underscores that among SSc patients with cardiac comorbidities and preserved LV function, even mild ILD can result in a mixed PH profile that demands judicious vasodilator titration, taking into account both efficacy and tolerability [4].

The third patient evolved with mixed PH 10 years after the SSc diagnosis, in which there were no cardiovascular risk factors. Her hemoptysis and severe shortness of breath were found to be secondary to combined PH in the context of classic features of scleroderma such as Raynaud's phenomenon, sclerodactyly, and cutaneous telangiectasias. Per initial non-emergent pulmonary exam and the hemodynamics, RHC determined the need for targeted treatment. This case underscores the need of systematic screening and early hemodynamic evaluation in SSc patients, even in the absence of manifest clinical or imaging signs of PH [5].

Collectively, these cases demonstrate for the multifaceted picture of PH in SSc; a single classification may be insufficient to encompass the interactions of the vascular, myocardial and pulmonary features. Right heart catheterization is the reference standard for diagnosis and is used for stratification but should be interpreted in the context of overall clinical and imaging findings [6]. From a therapeutic point of view, an individualized strategy according to the most prevalent pathophysiological mechanism, concomitant diseases and drug tolerance of each patient is mandatory [7].

The early recognition, multi-modality investigation and personalized treatment of PH in systemic sclerosis can significantly enhance the

functional status and quality of life, a scenario observed in all the three cases within this series [8].

CONCLUSION

This case series demonstrates the spectrum of clinical and hemodynamic pulmonary hypertension in systemic sclerosis. Accurate definition by right heart catheterization and adjunctive imaging is crucial for management. Although all patients had an autoimmune background, patient response was individualized to assessment and treatment. The use of targeted therapies, such as endothelin receptor antagonists and phosphodiesterase-5 inhibitors, resulted in amelioration of functional status in all cases. These results also underscore importance of early detection, individualized management, and the collaboration by a multidisciplinary team for improvement of prognosis and quality of life in patients with SSc-associated PAH.

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