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Pulmonary Capillary Hemangiomatosis: A Rare Cause of Pulmonary Arterial Hypertension, Presenting as Supraventricular Tachycardia

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Abstract

With a prevalence of less than 1/million, Pulmonary Capillary Hemangiomatosis is a rare disorder of capillary proliferation in the alveolar septae leading to pulmonary arterial hypertension and mimics pulmonary veno-occlusive disease.

INTRODUCTION

Pulmonary capillary hemangiomatosis (PCH) is a rare cause of primary pulmonary arterial hypertension (PAH) in the world with less than 1 per million prevalence. It is characterized by the proliferation of small capillaries in the lung parenchyma and the visceral pleura [1]. Echocardiographic examination and high-resolution chest computed tomography (CT) in patients suspected to primary PAH makes the early diagnosis of this disease. However, surgical lung biopsy as the gold standard of diagnosis is a risky procedure in critically ill patients [2]. Differentiation of PCH from Pulmonary veno-occlusive disease (PVOD) is an important consideration because treatment options are different in both conditions.

CASE PRESENTATION

A 35-year female presented with shortness of breath and palpitation for 2 hours. On examination, pulse rate was 150beats/minute, BP was 120/70 mm Hg, respiratory rate 20/minute, and Oxygen saturation was 90% at room air. Her chest examination revealed a pan systolic murmur at the tricuspid region with loud P2 and bibasilar crepitations. Electrocardiography showed supraventricular tachycardia, which was converted to sinus rhythm after treatment (Fig 1). She was treated with intravenous adenosine, diuretics and supportive measures.

On routine investigations showed normal renal and thyroid functions. Autoimmune profiles were negative. 2D echocardiography examination suggest a severe tricuspid regurgitation with pulmonary artery pressure of 50mmHg, other valves were normal, chest x-ray was suggestive of the dilated pulmonary artery and cardiomegaly, and high-resolution chest CT was suggestive of ground-glass opacities forming nodules in bilateral extensive lung fields (Fig 2).

DISCUSSION

PCH results from the extensive proliferation of small capillaries within alveolar septae and this proliferation mainly in the central part as compared to peripheral part [2]. PCH Manifests as dyspnea, hemoptysis, Pulmonary Arterial Hypertension, and right heart failure [3]. Histopathologically differentiated from PVOD by the presence of abnormal proliferation of small, thin-walled capillaries expanding alveolar septae in contrast to small veins blockade by organized thrombi in PVOD. Pulmonary capillary wedge pressure is normal in both conditions.

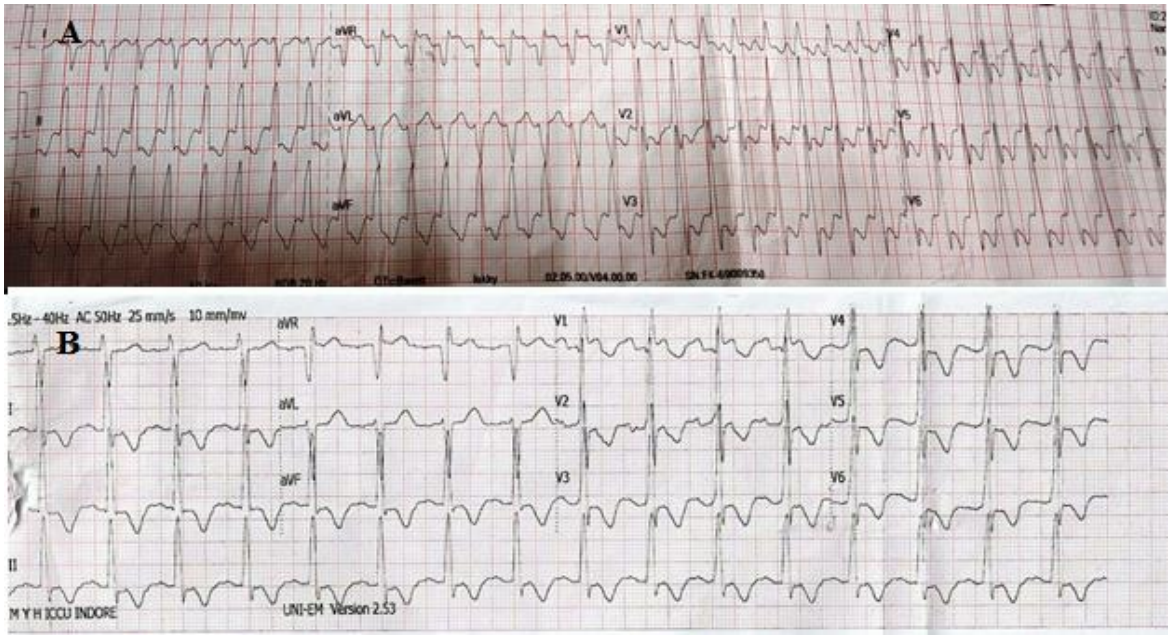

Figure 1. A) Electrocardiogram Showing SUPRA Ventricular Tachycardia. B) Electrocardiogram (after Treatment) Showing Normal Sinus Rhythm with Right Ventricular Overload.

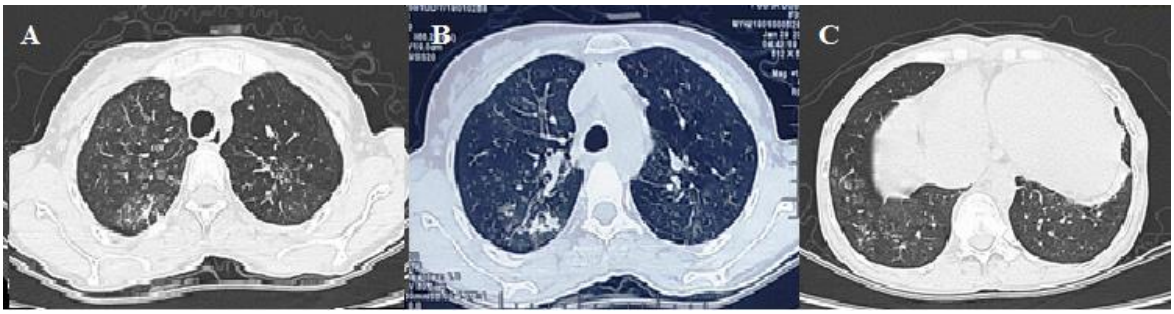

Figure 2. Axial Section High-Resolution Chest Computed Topography Showing Ground-Glass Opacities Forming Nodules in Bilateral Lung Fields. A) Apical Level, B) Middle Level, and C) Basal Level

High-resolution chest CT shows centrilobular ground-glass opacities sparing periphery is highly suggestive of PCH [4]. Treatment of this condition includes diuretics, Angiotensin II receptor blockers, warfarin, and Interferon-alpha 2a. Prostaglandin therapy, considered the treatment of choice in patients with primary PAH, may produce acute pulmonary edema and is contraindicated in patients with PCH. At present definitive treatment for PCH is lung transplantation [2].

CONCLUSION

Rare causes of PAH like PCH should be looked before labeling it as idiopathic. Usage of vasodilators can worsen the symptoms, and early lung transplantation can improve the outcome in this condition.

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Conflict of Interest

There is no conflict of interests with this article.

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