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THE SILENT BLOCK: ISOLATED RIGHT PULMONARY ARTERY OCCLUSION AS A MANIFESTATION OF CTEPH IN A MIDDLE-AGED WOMAN

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ABSTRACT:

Chronic thromboembolic pulmonary hypertension (CTEPH) is a progressive form of pulmonary hypertension caused by persistent obstruction of the pulmonary vasculature following thromboembolism. It is a potentially curable condition if identified early. We report the case of a 53-year-old female who presented with exertional dyspnoea, nausea, and vomiting for 10 days, with a significant history of prior similar episodes. Imaging and diagnostic workup revealed right pulmonary artery occlusion, right heart dilation, and perfusion defects consistent with CTEPH and cor pulmonale. Management included anticoagulation and pulmonary vasodilators, with symptomatic improvement, and she was evaluated for definitive surgical intervention via pulmonary endarterectomy. This case highlights the diagnostic challenges and importance of timely recognition of CTEPH to prevent progression and improve outcomes.

KEYWORDS: Chronic thromboembolic pulmonary hypertension, CTEPH, pulmonary artery occlusion, cor pulmonale, pulmonary endarterectomy, unilateral perfusion defect

INTRODUCTION:

Chronic thromboembolic pulmonary hypertension (CTEPH)[1] is a rare yet serious cause of pulmonary hypertension characterized by persistent obstruction of pulmonary arteries by organized thrombi. Despite being classified under WHO group 4 pulmonary hypertension, CTEPH remains underdiagnosed due to its nonspecific clinical presentation and insidious onset. Unlike other forms of pulmonary hypertension, it is potentially curable with pulmonary

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endarterectomy. The presence of cor pulmonale[2], right heart strain, and unilateral perfusion defects on imaging should prompt further investigation. We present a diagnostically challenging case of CTEPH in a middle-aged woman with isolated right pulmonary artery involvement and no identifiable thrombophilic disorder.

CASE PRESENTATION:

A 53-year-old postmenopausal female presented with progressive breathlessness on exertion for 10 days, accompanied by nausea and vomiting. She had a prior history of similar complaints requiring hospitalization. There was no significant family or personal history suggestive of thrombophilia or autoimmune disease. Menarche occurred at 14 years, menopause at 43, with two normal vaginal deliveries. On examination, her pulse was 111 bpm, BP 100/70 mmHg, respiratory rate 25 cpm, and SpO_2 was 85% on room air. Bilateral pedal oedema was noted, but no pallor, cyanosis, clubbing, or lymphadenopathy. Systemic examination revealed bilateral wheeze. Cardiovascular exam revealed normal heart sounds without murmurs. Lab investigations showed normal renal function, normal coagulation profile, and mildly deranged liver enzymes (total bilirubin 3.40 mg/dL, indirect bilirubin 3.07 mg/dL, GGT 160 U/L). Haemoglobin was 14.6 g/dL, RBC count was elevated (7.34 million/µL), and D-Dimer was 0.27 ug/ml (normal). NT-proBNP was significantly elevated at 1060 pg/mL. ECG showed sinus tachycardia, right ventricular hypertrophy, and right atrial enlargement. Chest X-ray showed enlarged cardiac silhouette, oligemic right lung field[3] as shown in figure 1.

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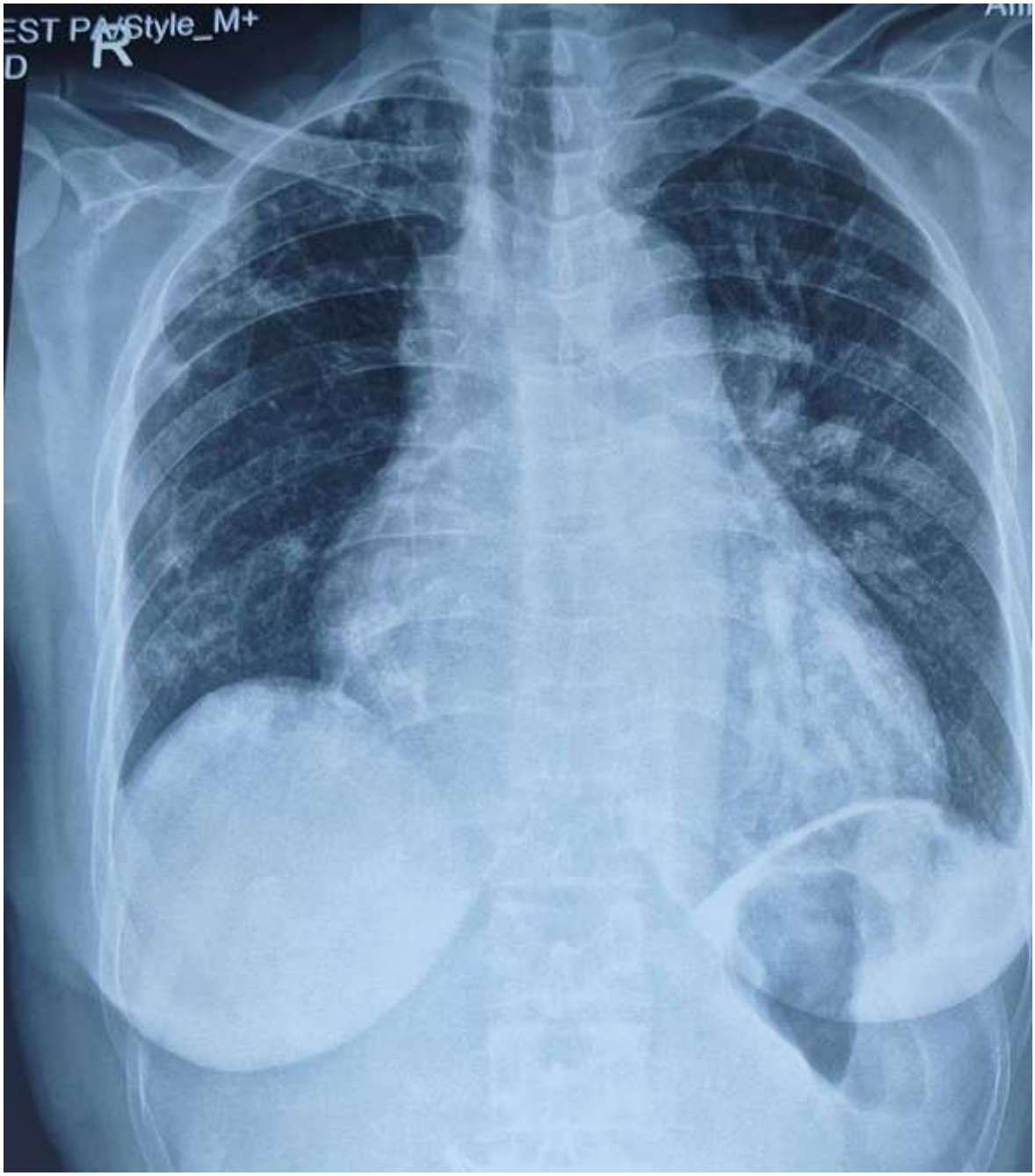

FIGURE 1 : CHEST X-RAY (PA VIEW)

2D echocardiography revealed a dilated right atrium and right ventricle with RV dysfunction (TAPSE = 12 mm), LVEF = 50 % severe tricuspid regurgitation, right pulmonary artery 8mm (no flow visualised), severe pulmonary artery hypertension[4] (RVSP = RAP + 81 mmHg), and D-shaped left ventricle. CT pulmonary angiography as shown in figure 2 and 3 showed total occlusion of the right pulmonary artery with wall enhancement and a diffusely narrowed lumen,

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consistent with organized thrombus, dilated main pulmonary artery and left pulmonary artery. Mosaic attenuation and aortopulmonary collaterals[5] were also present.

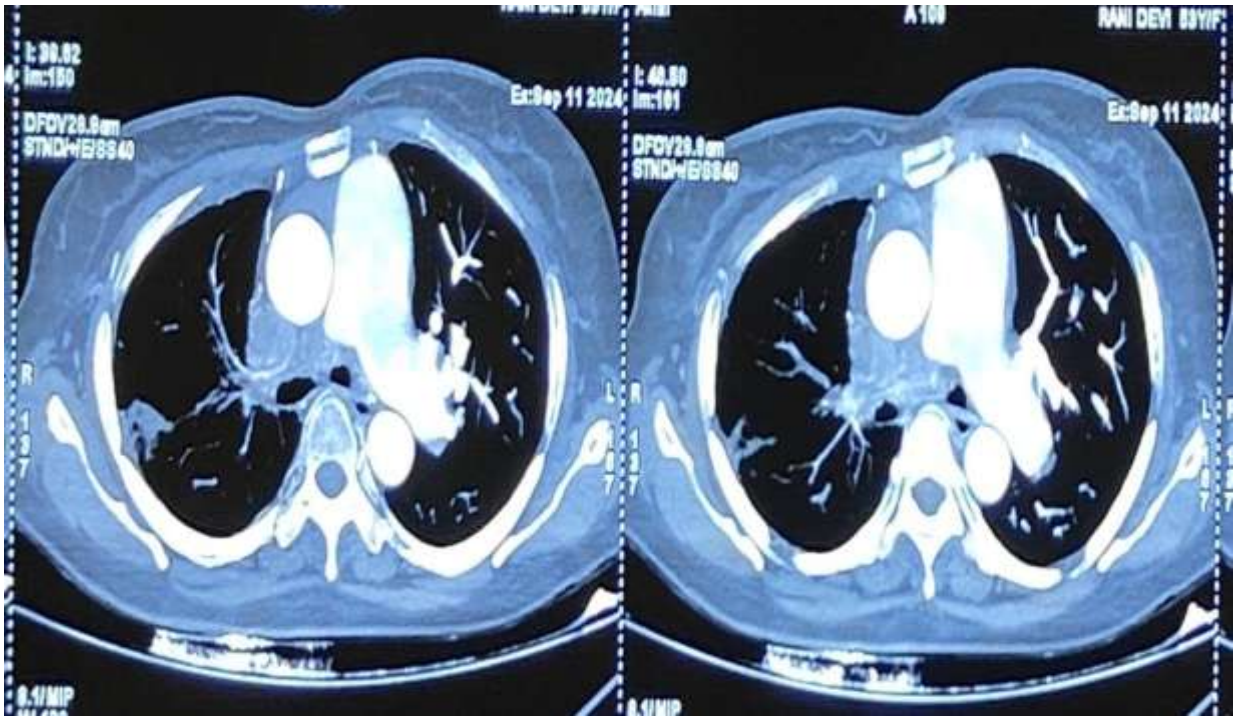

FIGURE 2 : CT PULMOMARY ANGIOGRAPHY

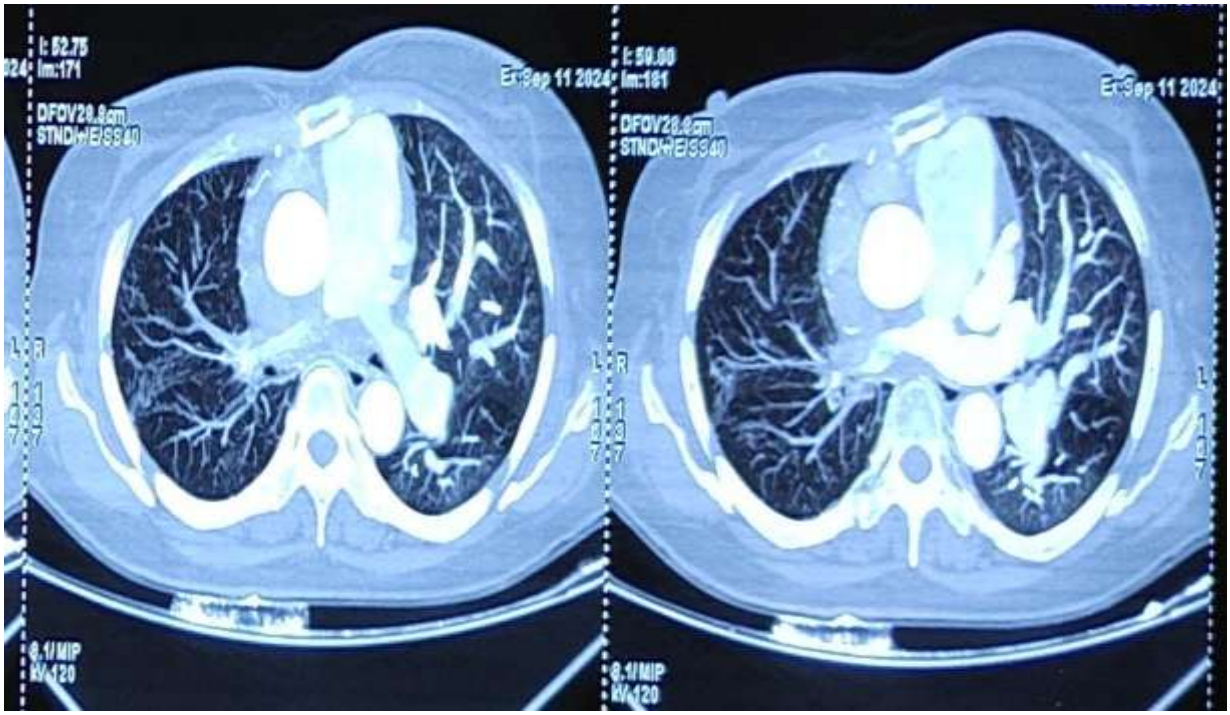

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FIGURE 3 : CT PULMONARY ANGIOGRAPHY

Lung perfusion scintigraphy with 99mTc-MAA[6] demonstrated complete lack of perfusion to the right lung, while the left lung was unaffected. Bronchoscopy and BAL were non-diagnostic, and infectious aetiologies including tuberculosis were ruled out. The patient was diagnosed with chronic thromboembolic pulmonary hypertension with cor pulmonale. Treatment was initiated with Riociguat 1 mg BID[7], Apixaban 5 mg BID, and a combination of torsemide 10 mg and spironolactone 50 mg OD. Significant symptomatic improvement was observed within two weeks. She was counselled for pulmonary endarterectomy as a potential curative option.

DISCUSSION: CTEPH is a rare sequel of unresolved pulmonary embolism, occurring in approximately 3–4% of patients after acute thromboembolism. The pathogenesis involves mechanical obstruction of pulmonary arteries by fibrotic thrombi and secondary vasculopathy of small vessels. Common symptoms such as exertional dyspnoea and fatigue overlap with other cardiopulmonary conditions, often delaying diagnosis. In our case, the patient had unilateral pulmonary artery involvement with right lung oligemia and absent perfusion—an unusual presentation. Differential diagnoses considered included idiopathic pulmonary arterial hypertension, Takayasu arteritis, fibrosing mediastinitis, and unilateral pulmonary artery agenesis[8]. However, the presence of an occluding thrombus with vascular wall enhancement on imaging and mosaic attenuation on CT strongly supported the diagnosis of CTEPH. Echocardiography and elevated NT-proBNP indicated right heart strain. Importantly, V/Q scanning remains the most sensitive tool for screening CTEPH, and our case showed complete absence of right lung perfusion. Management strategies for CTEPH include anticoagulation, pulmonary vasodilators (e.g., Riociguat), and surgical pulmonary endarterectomy—the only potentially curative approach. For inoperable cases, balloon pulmonary angioplasty or medical therapy remains the alternative. Our patient responded well to initial medical therapy and is being evaluated for surgery.

CONCLUSION:

This case emphasizes the importance of maintaining a high index of suspicion for CTEPH in patients with unexplained dyspnoea and evidence of right heart dysfunction. Unilateral perfusion defects should prompt thorough imaging to rule out thromboembolic disease. Early identification and referral for surgical assessment are crucial, as pulmonary endarterectomy offers curative potential in appropriately selected patients. Multimodal imaging and a structured diagnostic approach are key to managing this rare yet treatable cause of pulmonary hypertension.

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DECLARATION OF PATIENT CONSENT: The author certifies that he has obtained the patient's consent for her clinical information to be published in the journal. The patient understands that her name and initials will be kept confidential.

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