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Case Report

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A Case of Pulmonary Artery thrombosis in situ: A Great Masquerade: Case Report

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ABSTRACT

We present a case of pulmonary artery thrombosis in situ (PATis) (a pioneer report in Sri Lanka), which involves de novo clot formation within the pulmonary arteries due to complex pathophysiology. A 69-year-old with multiple comorbidities presented with syncope, dyspnoea, fever, pleuritic chest pain with tachypnoea, hypoxia and hypotension. Initially treated as sepsis triggered Multi-Organ Dysfunction (NSTEMI with HF, AKI and Type-1 respiratory failure) that initially improved with multi-organ support (NIV, Inotropes, diuretic infusions, therapeutic LMWH, and antibiotics) despite oxygen dependency with dangerous levels of desaturation. Further, Echocardiogram and CTPA confirmed PATis, but an extensive etiology screen was negative. He improved long-term anticoagulation highlighting the importance of detecting PATis with masquerading clinical presentation for better outcomes.

Keywords: Anticoagulation, denovo pulmonary embolism, pulmonary artery thrombosis in situ (PATis),

INTRODUCTION

We discuss a diagnostic masquerade, a case of pulmonary artery thrombosis in situ (PATis) or de novo formation of clots within the pulmonary artery rather than from distant sources unlike pulmonary embolism (PE). It arises often due to factors like endothelial dysfunction and systemic inflammation. PATis has gained attention due to its association with various clinical scenarios, particularly during the COVID-19 era (1) remains under-reported in Sri Lankan literature in which a case discusses about the first Experience with Catheter-Directed Thrombolysis for Massive PE at THJ (2), then another describes the management of PE in a chest trauma patient (3) and the next case describes an unusual phenomenon of IVC Thrombosis and PE in Bechet's disease (4). None of them explains a similar phenomenon of our case and reporting this case is of paramount value.

CASE REPORT

A 69-years-old male with hypertension, IHD, COPD (GOLD- 1) and dyslipidemia presented with two episodes of exertional syncope and progressively worsening exertional dyspnea over the past two weeks with low-grade fever, pleuritic type of chest

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pain for two days with worsening functional capacity (MRC 0 to 4). Otherwise, systemic enquiry and etiology analysis was unremarkable. He had a NSTEMI in 2023 and on medical follow up.

On examination, he was tachypneic and febrile with oxygen saturation of 84-86% on RA, improving to 92-94% with 10L/min oxygen via face mask. He had low volume pulse, hypotension with postural drop. His HR 92 bpm, with a RR of 24/min with bilateral fine to coarse crepitations (left > Right).

Initial POCUS revealed bilateral B-profile in the lung bases and a full, non-collapsing IVC. Initial ABG was abnormal (pH: 7.23, pCO2: 44 mmHg, pO2: 55 mmHg, HCO3: 19.1 mEq/L, Lactate: 2.65 mmol/L, PaO2/FiO2 ratio: 169). The ECG revealed (Figure 1) sinus rhythm with a heart rate of 75 bpm, prominent P-waves, right axis deviation, T-wave inversion in leads III, aVF, V1-V3, subtle ST depressions in V2-V3, and infrequent ectopic beats.

The working diagnosis included metabolic acidosis with respiratory failure (type-1), right ventricular strain, and moderate ARDS, with a likely mixed etiology including respiratory and renal components with possible septicemia and cardiogenic shock. The patient was initially started on CPAP ventilation, IV noradrenaline, furosemide infusions, therapeutic LMWH, and broad-spectrum antibiotics. Despite symptomatic improvement, severe exertional desaturation was noted.

Blood investigations revealed a neutrophil leukocytosis, rising S.Creatinine (from baseline 70 µmol/L to 220 µmol/L), High CRP (67 mg/L), rising Troponin I (50 times ULN), hyponatremia (Na+ 132 mmol/L) and hyperkalemia (K+ 5.6 mmol/L). Liver function tests were within normal limits.

A CXR (Figure 2) revealed cardiomegaly, Lung function testing showed FEV1/FVC ratio of 88% (which questioned the pre illness diagnosis of

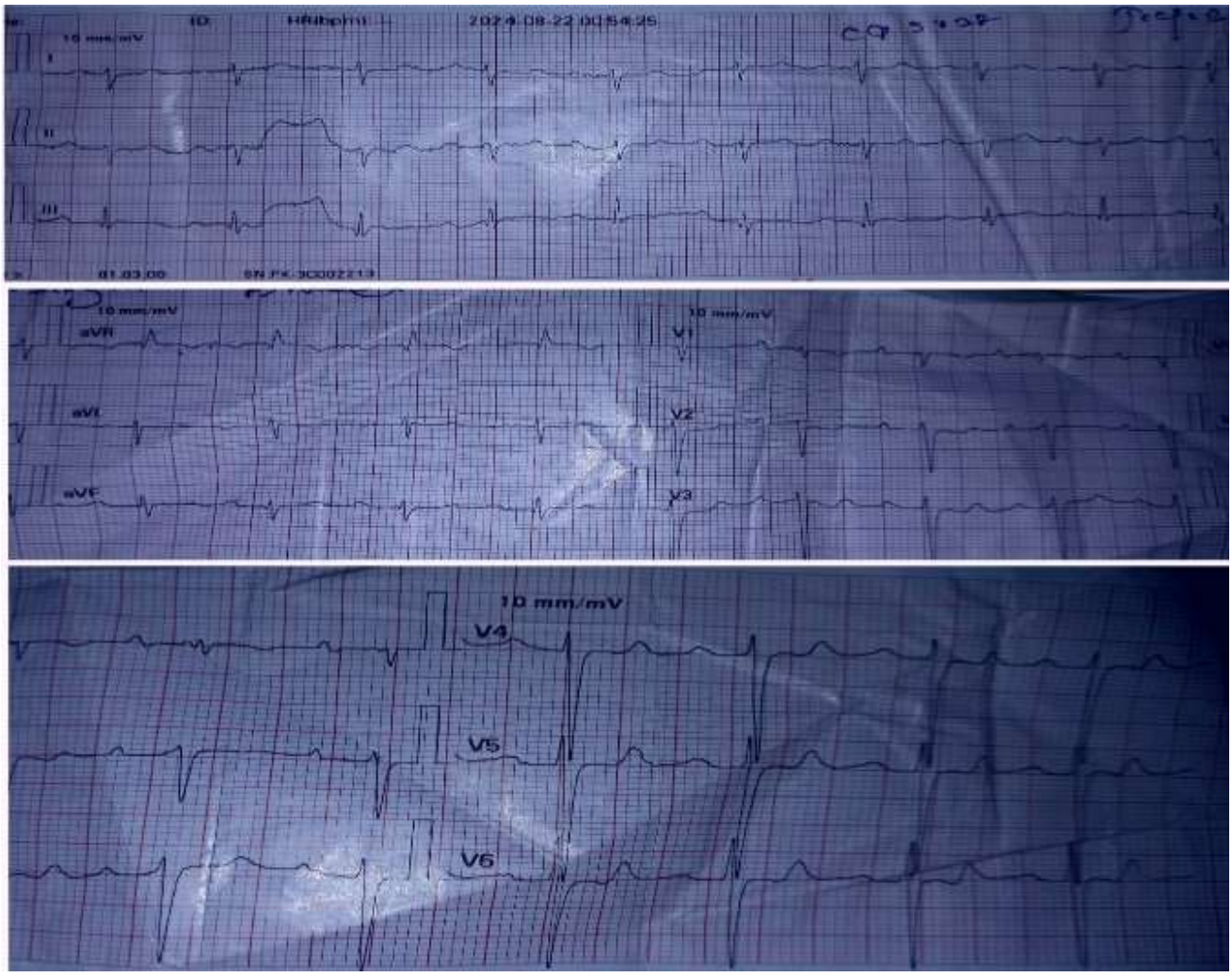

Figure 1: ECG

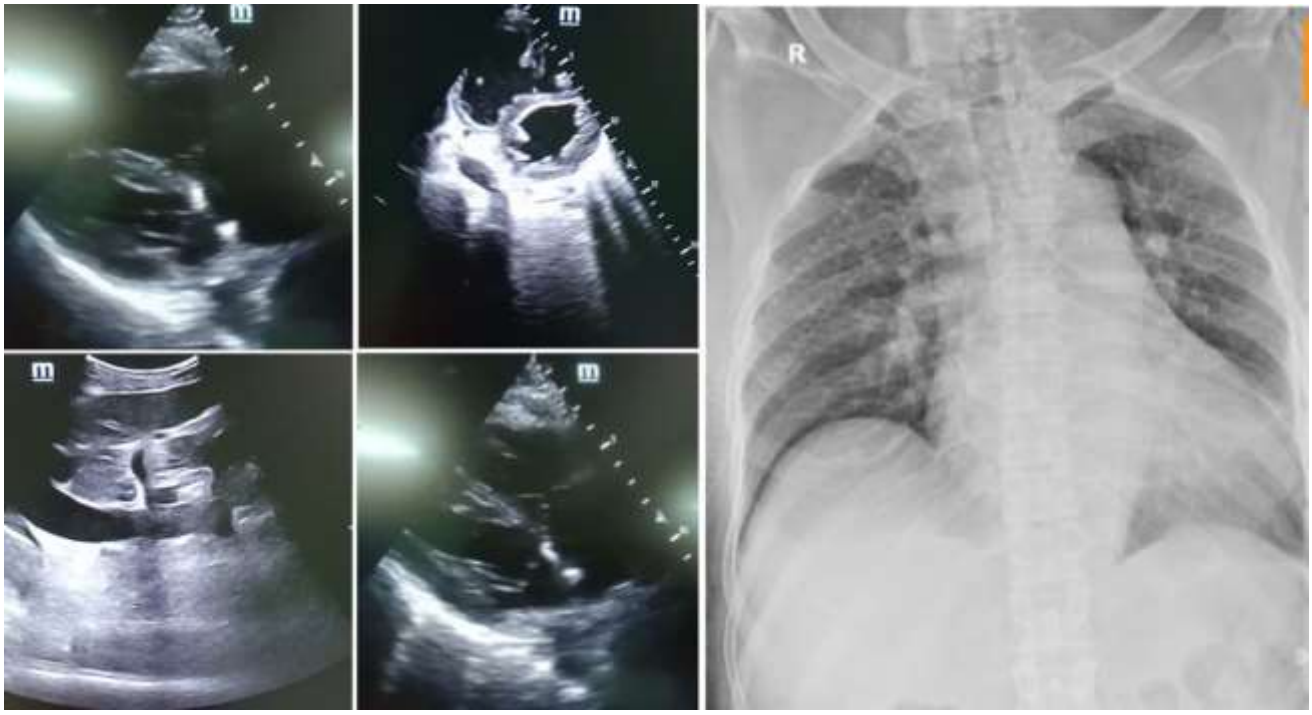

Figure 2: POCUS Echo & CXR PA

COPD). A 2D echocardiogram RA/RV dilation, severe pulmonary hypertension (TRPG of 77 mmHg), and normal EF (Figure 2). A DVT scan, USS Abdomen, ESR, thrombophilia and tumour maker screen and Covid-19 PCR were normal with elevated D-dimer levels (1.2 µg/mL). CTPA confirmed multiple filling defects in the segmental and sub-segmental branches of both the upper and lower lobes and the right middle lobe, suggestive of pulmonary thrombosis. Additionally, RA-RV dilatation, RV strain, and septal bowing towards the left ventricle were noted, although no thrombi were observed in the main pulmonary arteries. (Figure 3). The diagnosis of pulmonary embolism,

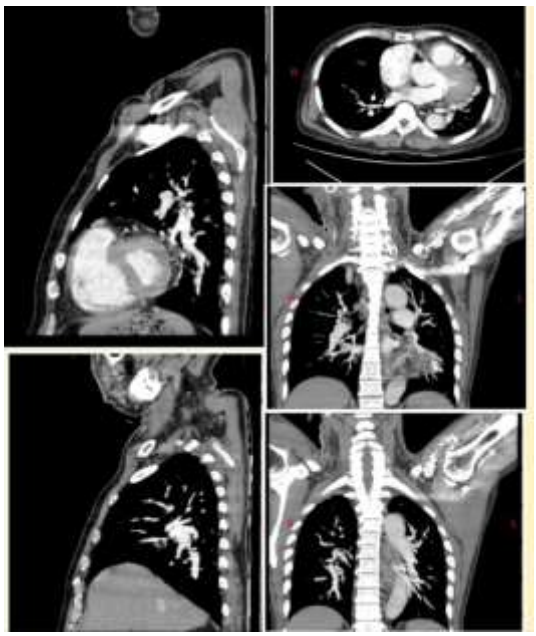

Figure 1: CTPA

likely in situ with multi-organ dysfunction (MOD) possibly due to infective etiology without any other provoked cause, was confirmed.

The patient was later started on warfarin with bridging therapy and gradually weaned off oxygen therapy after 2 weeks. He was discharged with oral anticoagulation therapy and follow-up arrangements.

DISCUSSION

PATis is increasingly recognized as a distinct clinical entity, separate from traditional PE. PATis occurs due to Virchow's Triad, particularly in conditions like pulmonary arterial hypertension (PAH) and COVID-19 (5, 6). Neutrophils recruitment is essential for thrombosis formation in the absence of DVT (6). Distinguishing PATis from chronic thromboembolic pulmonary hypertension (CTEPH) is important, as management strategies differ significantly. In situ, thrombus may not benefit from surgical interventions like pulmonary thromboendarterectomy (5). Literature reveals that PATis is often misdiagnosed as an embolic phenomenon, leading to inappropriate treatment (7).

In situ thrombosis may arise in the pulmonary arteries, linked to conditions such as hypoxia or

inflammatory changes, rather than dislodged thrombi from peripheral veins as in our case which had multiple segmental and subsegmental thrombi (8). In situ PAT arises often linked to conditions like COVID-19, trauma, or radiation therapy, while provoked PE typically originates from thrombi in peripheral veins, such as DVT often due to an underlying etiology (6, 9).

Diagnosis of PATis is challenging and often lacks the classic symptoms associated with PE and presents atypically and mandates a CTPA mostly. RV strain is commonly observed in PAT, with ECG changes noted in over 50% of cases. Echocardiography aids in diagnosis. Risk assessment Scoring systems like Caprini and IMPROVE VTE have shown strong correlations with PAT incidence, unlike Wells and Geneva scores, which are less effective (6). The diagnosis of isolated subsegmental PE as in our case, which can be complicated by false positives due to factors like suboptimal imaging techniques and interobserver variability among radiologists as well (10). Diagnosis often involves imaging techniques that reveal unique characteristics of in situ thrombi, such as their eccentric nature and location within the pulmonary artery (9).

Initial management includes oxygen therapy, analgesics, and anticoagulation to prevent clot progression (11, 12). Systemic thrombolysis is recommended for high-risk and unstable patients to rapidly dissolve clots, with major bleeding risk. Catheter-directed therapies are increasingly utilized, especially for patients with contraindications to systemic thrombolysis. These methods allow for targeted treatment with reduced bleeding risks [13]. Long-term anticoagulation is essential, with direct oral anticoagulants being preferred over traditional vitamin K antagonists. A multicenter study found that among patients with isolated subsegmental PE managed without anticoagulation, the cumulative incidence of recurrent venous thromboembolism (VTE) was 3.1% over 90 days. Specifically, 2.1% of patients with single isolated subsegmental PE and 5.7% with multiple isolated subsegmental PE experienced recurrent VTE (10). For patients developing CTEPH, pulmonary endarterectomy is the treatment of choice, while balloon pulmonary angioplasty may be considered for inoperable cases (13,14). But in PATis, strategies differ from those for CTEPH, necessitating tailored approaches such as anticoagulation and pulmonary vasodilators (6).

MOD in PATis is an important concern, especially in conditions like sepsis and COVID-19. In sepsis, pulmonary thrombosis can exacerbate lung injury and lead to renal and cardiac dysfunction. Mild thrombosis may prevent bacterial spread, while severe cases cause hypoxia and endothelial damage (15). COVID-19 has been linked to extensive thrombotic events, resulting in MOD. The cytokine storm and endothelial dysfunction contribute to this progression, leading to conditions like MI and PE (16,17).

Multi-Organ Ultrasonography technique has shown high sensitivity and specificity for diagnosing PE and timely management (18).

CONCLUSION

The diagnosis and management of PATis associated MOD remains challenging, highlighting the need for targeted therapies and further research into underlying mechanisms. This case contributes to the limited literature on PATis, emphasizing the importance of tailored management strategies.

Author declaration

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Authors' contributions:
Conceptualization and Design: NMMR; Data Collection and Patient Care: NMMR, IKJ, NA, PF, MTFH; Literature Review: NMMR, JFS; Writing - Original Draft: NMMR; Writing - Review & Editing: NMMR, IKJ, NA, JFS; Supervision: NMMR; Approval of Final Manuscript: All authors.

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Written informed consent was obtained from the patient before they participated in the case, ensuring their understanding of the purpose, procedures, and potential implications involved.

Statement on data availability:
All data generated during this study are included in this published article.

REFERENCES

1. Su M, Chang K, Goel K. Pulmonary artery thrombosis in situ: not all chronic clots are CTEPH. Chest. 2023;164(4):A6023-A6023. https://doi.org/10.1016/j.chest.2023.07.3883
2. First experience with catheter-directed thrombolysis for massive pulmonary embolism at teaching hospital, Jaffna, Sri Lanka: a case report. (https://www.researchgate.net/publication/383581782_First_Experience_With_Catheter-Directed_Thrombolysis_for_Massive_Pulmonary_Embolism_at_Teaching_Hospital_Jaffna_Sri_Lanka_A_Case_Report)
3. Wanigasekara V, Nanayakkara SD. Management of acute pulmonary embolism following chest trauma in a resource-limited setting – A case report. Sri Lankan J Orthop Surg. 2022;8(1):96-102. https://doi.org/10.4038/tsljos.v8i1.15
4. Karunanayaka SK, Jayawardhane VV, Premawansa G. An unusual presentation of Behçet's disease with inferior vena cava thrombosis and pulmonary embolism. Galle Med J. 2021;26(2):67-70. http://doi.org/10.4038/gmj.v26i2.8089
5. Baranga L, Khanuja S, Provancha JP, Gosselin MV, Walsh J, Arancibia R, et al. In situ pulmonary arterial thrombosis: Literature review and clinical significance of a distinct entity. AJR Am J Roentgenol. 2023. https://doi.org/10.2214/AJR.23.28996
6. Porembskaya O, Zinserling V, Tomson VV, Toropova YG, Starikova EA, Maslei VV, et al. Neutrophils mediate pulmonary artery thrombosis in situ. Int J Mol Sci. 2022;23(10):5829. https://doi.org/10.3390/ijms23105829
7. Ahsino F, Zagaouch D, Soualhi M, Zahraoui R, Bourkadi JE. Fungal origin of in situ pulmonary artery thrombosis: A case report. Int J Adv Res. 2022;10(9):197-201. https://doi.org/10.21474/IJAR01/15342
8. Hirmerova J, Bilková S, Woznica V. Isolated pulmonary embolism: A specific clinical entity? Vnitřní lékařství. 2023;69(1):8-13. https://doi.org/10.36290/vnl.2023.001
9. Ahuja J, Shroff GS, Benveniste MF, Marom EM, Truong MT, Wu CC. In situ pulmonary artery thrombosis: Unrecognized complication of radiation therapy. AJR Am J Roentgenol. 2020;215(6):1329-1334. https://doi.org/10.2214/AJR.19.22741
10. Risk for recurrent venous thromboembolism in patients with subsegmental pulmonary embolism managed without anticoagulation. Ann Intern Med. 2022;175(4):W43-W44. https://doi.org/10.7326/L22-0038
11. Silva ACdF, Azevedo A. Management of pulmonary thromboembolism. Res Soc Dev. 2023;12(6):e22612642098. https://doi.org/10.33448/rsd-v12i6.42098
12. The management of pulmonary embolism. Anaesth Intensive Care Med. 2023;24(2):115-122. https://doi.org/10.1016/j.mpaic.2022.12.001
13. Gupta A, Thulasidasan N, Mak SM, Rawal B, Padley SPG, Karunanithy N. Contemporary interventional management of pulmonary embolism. Clin Radiol. 2023;78(5):315-322. https://doi.org/10.1016/j.crad.2023.01.005
14. Ribas JS, Sánchez-Corral MÁ, Riera-Mestre A. Update in the management of chronic thromboembolic pulmonary hypertension. Med Clin. 2024. https://doi.org/10.1016/j.medcle.2023.08.007
15. Evans CE, Zhao YY. Impact of thrombosis on pulmonary endothelial injury and repair following sepsis. Am J Physiol Lung Cell Mol Physiol. 2017;312(4):L441-L448. https://doi.org/10.1152/AJPLUNG.00441.2016
16. Rodríguez C, Luque N, Blanco I, Sebastián L, Barberà JA, Peinado VI, et al. Pulmonary endothelial dysfunction and thrombotic complications in patients with COVID-19. Am J Respir Cell Mol Biol. 2021;64(4):407-415. https://doi.org/10.1165/RCMB.2020-0359PS
17. Habibi H, Afsharkia P, Jahangiri M, Mostafavi S. COVID-19 with multi-organ arterial and venous thrombosis: A case report. Sonography. 2024. https://doi.org/10.1002/sono.12458
18. Aktürk ÜA, Koak ND, Ernam D. The role of multi-organ ultrasonography for diagnosing non-massive pulmonary thromboembolism. Biomed Res. 2017;28(18):8044-8049 https://www.semanticscholar.org/paper/The-role-of-multi-organ-ultrasonography-for-Akt%C3%BCrk-Koak/0aa417803cd54f675686fbeae8fa122d072a5428?utm_source=direct_link