Atrial septal defect: Massive pulmonary embolism mimic in point of care ultrasound

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SUMMARY

Massive pulmonary embolism (PE), which manifests as the obstructive shock is one of the most unforgiving medical emergencies. Due to the overriding concern of adverse outcomes associated with delayed treatment, the diagnosis is commonly made based on clinical symptoms and bedside point of care ultrasound (POCUS) assessment in order to enable the timely institution of life-saving reperfusion therapy. The presence of McConnell's sign portends the possibility of PE with high specificity. Notwithstanding, this eponymous sign is also fraught with limitations as some cardiopulmonary pathologies may also manifest such a sign. Therefore, recognition of its mimickers is vital to guide clinicians to a correct diagnosis which then translate to the right treatment for patients. Herein, we report a case of undiagnosed atrial septal defect masquerading as massive PE in a 27-year-old lady, who presented with shock with concomitant McConnell's sign during the puerperal period. It is hoped that this vignette serves to remind the clinicians that it is important to be able to expound the differentials of McConnell's sign as its presence is not pathognomonic of PE. Besides, the incorporation of point-of-care cardiac doppler ultrasound (POCDUS) in an emergency setting should be considered as part of the POCUS evolving continuum to augment its diagnostic accuracy.

INTRODUCTION

Massive pulmonary embolism (PE) arises due to catastrophic pulmonary vasculature occlusion culminating in obstructive shock. Fatality is inevitable if diagnosed late or missed as its definite thrombolytic treatment is a time-sensitive therapy. Hence, majority of the massive PE cases are diagnosed based on patients' clinical conditions and point of care ultrasound (POCUS) findings.[1] Among the sonographic features of massive PE, McConnell's signs is one of the most highly cited findings, in which if present concurrently with acute hypotension will create a heightened suspicion towards massive PE.[2,3] Herein, we describe a case of undiagnosed atrial septal defect (ASD) complicated by right heart failure, mimicking a massive PE by presenting with shock and McConnell's sign during POCUS evaluation. The final diagnosis was uncovered by a comprehensive transthoracic echocardiogram and a normal CT pulmonary angiogram (CTPA) subsequently.

CASE PRESENTATION

A 27-year-old lady, who was 13 days post-spontaneous vaginal delivery, presented to the health clinic with the chief complaint of dizziness for 1 day associated with presyncope. Otherwise, she denied episodes of palpitation, pleuritic chest pain, reduced effort tolerance or haemoptysis. At the presentation in the health clinic, her blood pressure measured 76/47 mmHg with a pulse rate of 89 bpm and peripheral oxygen saturation of 92%. Physical examination was unremarkable. She was immediately transferred to the Emergency Department (ED), Hospital Tengku Ampuan Rahim, for suspected acute PE.

Assessment in ED revealed a dehydrated patient with coated tongue and fluid resuscitation was commenced promptly. Physical examination was unremarkable except for signs pertaining to dehydration. Despite 1 litre of fluid resuscitation, she remained hypotensive with blood pressure ranging from 77/57 to 90/60 mmHg and she was subsequently started on intravenous noradrenaline infusion. The presence of type 1 respiratory failure warranted her to be given supplemental oxygen of 3 L/min. To note, she was not tachycardic throughout the observation. Electrocardiogram showed left axis deviation with right bundle bunch block. (Figure 1) On the other hand, POCUS evaluation pre-fluid resuscitation demonstrated dilated right atrium and ventricle with D-shaped ventricles and kissing inferior vena cava (IVC). A repeated POCUS was performed and showed an increment of IVC diameter to 1.7 cm with dilated right atrium and ventricle with McConnell's sign observed. In view of patient's prevailing history, haemodynamic instability with POCUS findings suggestive of massive PE, she was thrombolysed with intravenous (IV) tenecteplase 6000 IU followed by intravenous heparin infusion. Her D-dimer result later was 0.9 µg/ml, which was marginally raised.

The thrombolysis therapy was uneventful, and she was admitted to high dependency ward for close monitoring. CTPA was performed the following day and there was no CT evidence of PE. In addition, bilateral lower limb doppler ultrasound was also performed and similarly did not detect any evidence of deep vein thrombosis. In order to elucidate the aetiology of the abnormal POCUS findings, a comprehensive transthoracic echocardiogram was performed, which illustrated a large secundum ASD with the dilated right ventricle (Figure 2). Anticoagulation therapy was discontinued, and the final diagnosis was revised as

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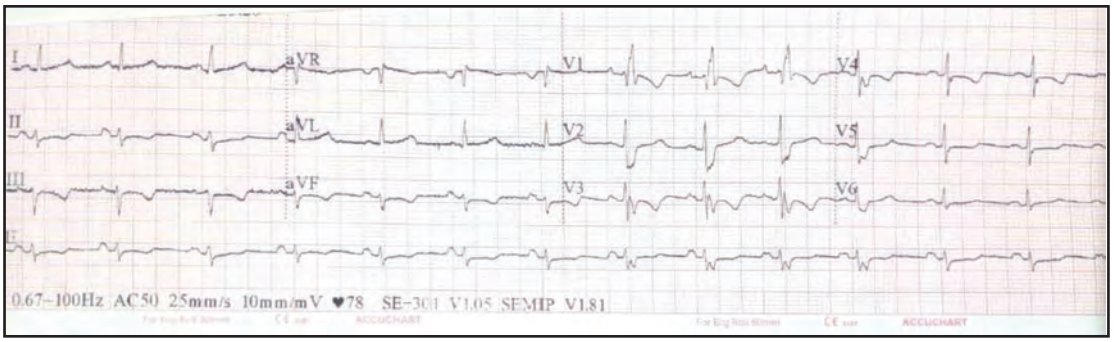

Fig. 1: ECG shows left axis deviation with right bundle brunch block. Widespread T Inversion seen from V1 to V4

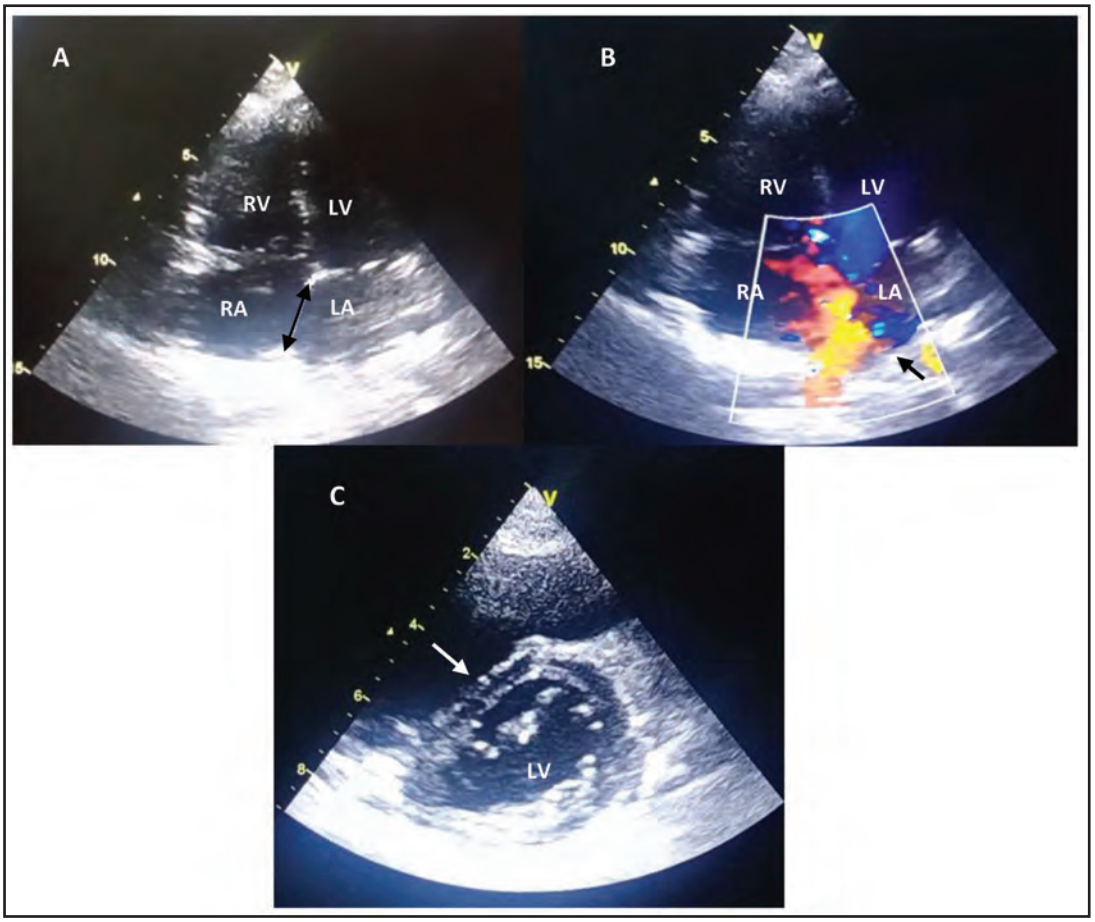

Fig. 2: Transthoracic echocardiogram. (A) ASD [double-headed arrow] with dilated RV and RA. (B) ASD with prominent left to right shunt on colour doppler USG [black arrow head] (C) Dilated LV with septal flattening. [white arrow head] ASD=atrial septal defect; LA=left atrium; LV=left ventricle; RA=right atrium; RV=right ventricle

symptomatic large secundum ASD. She was discharged home well and referred to another cardiology institution for further treatment. At the time of writing this report, she remained well and was scheduled for ASD repair.

DISCUSSION

Acute presentation with haemodynamic compromise and hypoxaemia often prompts the clinician to consider the diagnosis of massive PE. Though tachycardia was absent in this case, the abrupt symptoms onset during the puerperal period and the presence of right ventricular strained ECG patterns strengthened the suspicion PE. Bedside POCUS has been recommended as the initial test in evaluating such patients with suspected massive PE, characterised by haemodynamic instability to elicit signs of acute pulmonary hypertension or right ventricular dysfunction. In highly unstable patients, reperfusion therapy should be instituted if there is supporting evidence of PE based on POCUS findings. Final confirmation with CTPA would be performed once the patient has been stabilised.[1]

Acute PE has been known to be able to contribute to RV pressure overload and dysfunction, which could be detected by POCUS. Numerous echocardiographic features of PE have been enumerated in the European Society of Cardiology guidelines.[1] They are represented by RV dilation as well as measurements that could suggest right heart strain or dysfunction, such as disturbed RV ejection pattern (60-60 sign), abnormal Tricuspid Annular Plane Systolic Excursion (TAPSE) and McConnell's signs. The caveat is that any significant cardiac or pulmonary pathology could mimic such findings. In our case, the subject demonstrated RV dilation, septal flattening as well as McConnell's signs during POCUS.

McConnell's sign, depicted as right ventricular mid-free wall akinesia with apical preservation, was first described by McConnell et al in a seminal paper published in 1996. In the original report, this finding had a 77% sensitivity, 94% specificity, positive predictive value (PPV) of 71%, negative predictive value of 96% for the presence of PE in patients.[4] In recent years, Daley et al. who examined a cohort of 136 subjects with suspected PE, demonstrated that the McConnell's sign only had a sensitivity of 33% towards acute PE, whilst the specificity remains as high as 99%.[5] On the other hand, Vaid et al who retrospectively analysed 73 patients with McConnell's sign detected by echocardiogram, demonstrated that the PPV of this sign on acute PE was only 57%.[6] Considering all these, McConnell's sign should not be used in isolation when making a diagnosis of PE in patients, as well as directing the use of reperfusion treatment in unstable patient.

A myriad of published cases illustrating McConnell's sign as PE mimickers have been reported. Rafie et al described two cases with this eponymous sign attributed to right ventricular ischaemia caused by acute occlusion of right proximal coronary artery.[7] Walsh et al highlighted a case of pulmonary hypertension due to chronic obstructive pulmonary disease and systemic lupus erythematosus with similar finding.[8] To the best of our knowledge, our case represents the first case of undiagnosed ASD mimicking as massive PE which demonstrated McConnell's signs during ED presentation. Overall, these case vignettes serve to remind the clinicians to be cognizant of the McConnell's sign differential diagnoses, as a plethora of cardiopulmonary pathology with RV strain could demonstrate such eponymous sign.

In retrospect, the pre-syncopal attack in our case represents the first manifestation of severe ASD. Though it is unclear why she was devoid of heart failure symptoms during her recent pregnancy. Yet, it is crucial to consider the possibility of undiagnosed congenital septal defects when assessing individuals with such presentation. We postulate that the McConnell's sign observed in this case was probably due to the volume gradient from the shunt. The hypotensive episode was unlikely due to an obstructive shock caused by massive PE as there was no evidence of PE based on the CTPA findings and serial normal lactate level. Furthermore, the D-dimer value was only marginally raised and bilateral lower limb doppler was negative. Hence, it is reasonable to speculate that the hypotensive episode could be caused by a combination of hypovolemia with right ventricular failure as a result of untreated ASD.

Decision on the selection of ultrasound modes depends of the clinical disease suspected by the clinicians. In general, colour Doppler ultrasound would be considered only if there is a need to measure and visualising blood flow during POCUS evaluation. For example, it would be utilised when evaluating a subject with suspected deep vein thrombosis or even in a subject with suspected vascular injury.[9,10] Our case highlights that incorporation of cardiac Colour Doppler during POCUS (POCDUS) should be considered during critical setting, especially among subjects with evidence of structural cardiac abnormality during initial assessment, as it could assist in revealing the underlying abnormal shunt or valvular pathology. Nevertheless, adoption of POCDUS in critical settings remains to be elucidated in future studies to determine its diagnostic value when being applied within a limited time under emergency situations.

CONCLUSION

Acute PE is a life-threatening condition and yet treatable with prompt diagnosis and intervention. POCUS together with typical physical signs and symptoms have been typically employed to diagnose PE, especially when CTPA is not readily available or clinical circumstances hinder such an approach. Among the echocardiography signs observed in acute PE, McConnell's sign has long been established as a reliable sign with high specificity. However, there is always an exception as reported in this case and clinician should be aware that McConnell's sign is not equivalent to acute right heart strain from PE. It is crucial to contemplate other conditions and utilise other diagnostic tools to enhance diagnosis accuracy, which in this case is by using the colour doppler. In future, the use of POCDUS can be expounded upon and studied to evaluate its value in a high-strung emergency setting.

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CONFLICT OF INTEREST

The authors have no conflicts of interest to declare.

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