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Trivial Complaint in an Eisenmenger Syndrome Patient: A Case Report

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Abstract

Acute pulmonary embolism (PE) is a life-threatening cardiorespiratory condition. Shortness of breath and cyanosis in an active daily living patient with Eisenmenger Syndrome (ES) obscuring the early diagnosis of acute PE. We reported a case of a 41-year-old lady with underlying Eisenmenger syndrome presented to Emergency Department with sudden onset of palpitation. Otherwise, she did not complain of any worsening shortness of breath or reduced effort tolerance. Eisenmenger syndrome causes chronic hypoxemia because of the right-to-left shunting of blood, leading to a pro-thrombotic state and increasing the risk of blood clot formation. People who have Eisenmenger syndrome are more likely to develop venous thromboembolism (VTE), which includes deep vein thrombosis (DVT) and pulmonary embolism (PE). The increased risk of VTE is due to blood flow stasis, endothelial dysfunction, and abnormalities in blood composition. Compensation of the heart causing normalization of the blood pressure in ES may mask the features of massive PE and make it more challenging in managing this kind of patient. However, with the complaint of acute onset palpitation trigger the managing team to think about PE. Computed Tomography Pulmonary Angiography (CTPA) had done, and there were significant blood clots in the right main pulmonary artery. Luckily, she survived after thrombolytic therapy, warded a few days, and discharge well with a life-long oral anticoagulant.

Keywords
Eisenmenger Syndrome, Pulmonary Embolism, Thrombolytic Therapy

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Introduction

Eisenmenger Syndrome (ES) is the most severe form of pulmonary arterial hypertension in a patient with underlying congenital heart disease. It is a long-standing left-to-right cardiac shunt until it eventually leads to a cyanotic right-to-left shunt. The clinical baseline for an ES patient is always in a cyanotic and dyspnoeic state [1]. However, they may present with a trivial complaint with underlying sinister complications. As the disease progresses, the impact of polycythaemia may lead to a hypercoagulable state and an increased risk of PE [2]. Increasing age, presence of biventricular dysfunction, dilatation of the pulmonary arteries and consequently reduced pulmonary flow velocity favour for development of pulmonary embolism (PE) [3]. This condition will further lead to the formation of pulmonary artery thrombosis in ES patients. We report a case of an ES patient who complained of palpitation without any evidence of tachycardia or hypotension, with a final diagnosis of acute massive PE.

Case Presentation

A 41-year-old lady, a known case of ES with atrial septal defect (ASD) and pulmonary hypertension, presented to Emergency Department (ED) with a chief complaint of sudden onset of palpitation while she was asleep. She was on Sildenafil and compliant with the medication. Premorbid, she was cyanotic and could ambulate with minimal shortness of breath. She was able to perform daily housekeeping independently with long-term oxygen therapy (LTOT) of 3 litre/minute through a nasal prong. She described the palpitation was associated with diaphoresis and dizziness.

She was triaged to the critical area as she appeared tachypnoeic and lethargic. On examination, there was presence of clubbing with central and peripheral cyanosis. Her oxygen saturation (SpO2) on room air was 89% and improved to more than 90% after administration of high flow mask oxygen 15 L/min. She was afebrile. Her blood pressure was 110/70 mmHg and her heart rate of 65 beats per minute. Cardiac examination revealed parasternal heave, loud second heart sound and ejection systolic murmur at the left sternal edge. The lungs examinations were unremarkable. There was no calf swelling or tenderness. Arterial blood gases (ABG) under high flow oxygen supplementation showed pH: 7.28, PaO2: 81.1mmHg, PaCO2: 32 mmHg, bicarbonate: 16.5 mmol/L and base access: -10.5. She also had polycythaemia with haemoglobin (Hb): 19.6 gm/dL and haematocrit: 57.6%. The electrocardiogram (ECG) showed right bundle branch block (RBBB) with 1st-degree heart block (Figure 1), which was similar to her baseline ECG.

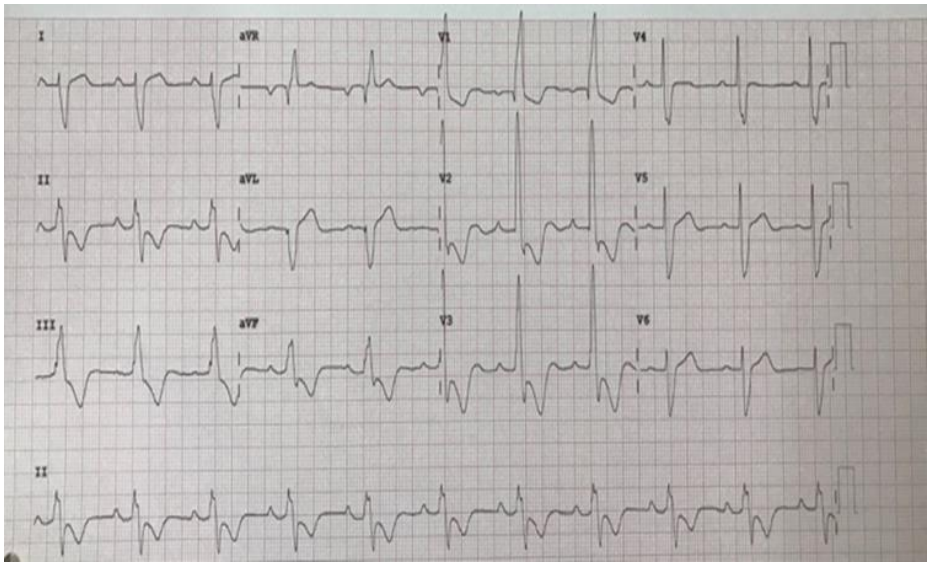

Figure 1: ECG shows RBBB with 1st degree heart block

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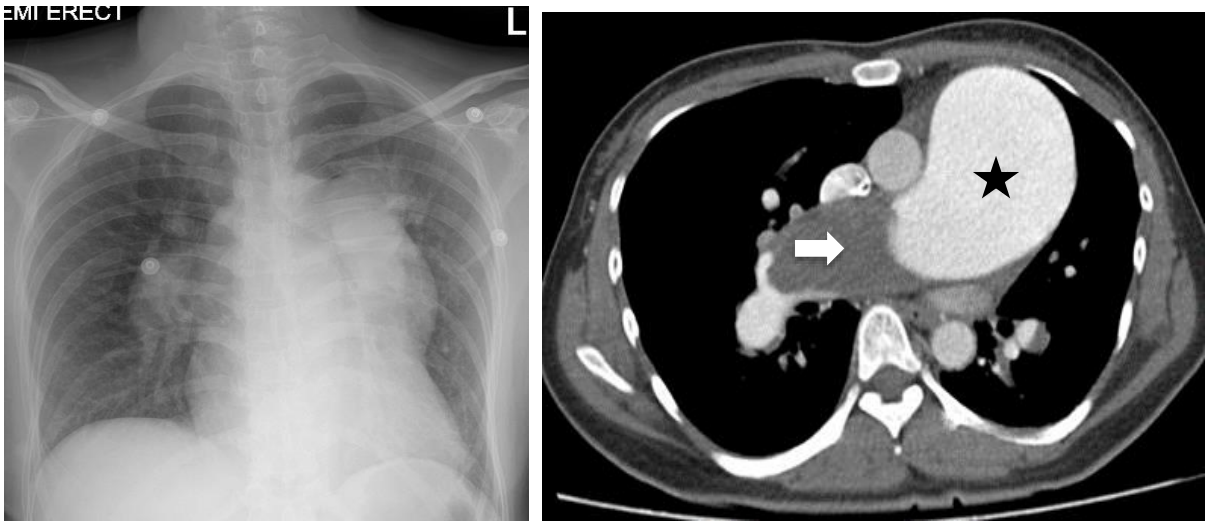
Bedside echocardiography (ECHO) demonstrated right atrium (RA) and right ventricle (RV) dilatation with a D-shaped septum, no regional wall motion abnormality and good contractility. The two-point compression test was negative.

A semi-erect chest x-ray revealed enlarged bilateral main pulmonary arteries with the pruning of peripheral vessels (Figure 2 A). She had undergone Computed Tomography Pulmonary Angiography (CTPA), which revealed massive bilateral pulmonary embolism with large ASD (Figure 2 B).



A B

Figure 2: (A)Enlarged bilateral main pulmonary arteries with pruning of peripheral vessels. (B): CT pulmonary angiography (CTPA) shows grossly dilated pulmonary trunk (★) and large thrombus in right main pulmonary artery (white arrow)

IV alteplase 10 mg bolus was administered followed by 60 mg over an hour infusion as per local protocol and subsequently admitted to cardiac care unit (CCU). She was on high flow mask oxygen supplementation and was able to wean down to her baseline oxygen therapy upon discharge after 3 days. During her stay, she developed haematuria secondary to the thrombolytic therapy. However, her symptom resolved spontaneously, and she was continued with Rivaroxaban 15 mg twice daily for 21 days and then 20 mg daily for lifelong.

Discussion

Diagnosing acute PE is always a significant challenge to physicians, especially in patients with abnormal clinical baselines. In conjunction with this problem, it is challenging to diagnose when the patient's blood investigations, ECG and chest radiograph showed no significant changes compared to her baseline findings of ES. Furthermore, the echocardiography finding was compatible with right ventricular dysfunction which commonly seen in ES patient suffering from pulmonary hypertension. In addition to her primary complaint of sudden onset palpitations without evidence of tachycardia, it is a dilemma for the attending physicians to obtain a working diagnosis. Palpitation is the perception of one's own heartbeat, which is not always due to an increased heart rate. It can indicate a harmless condition, for instance, if triggered by anxiety, or it may signal a significant issue, such as cardiac arrhythmia. The specific symptoms, associated factors, and potential risk factors will dictate the specific examination and assessment required to arrive at a diagnosis [1]. Furthermore, she encountered only minor symptoms such as diaphoresis and dizziness. Typical clinical

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signs and symptoms of adults with PE are dyspnoea, fatigue and syncope [2,5-6]. However, this patient also complaint of dizziness which can be usual symptoms in ES due to hyperviscocity syndrome as her haematocrit reveal polycythemia [7]. At the beginning, physicians also in difficulty whether this patient might have acute coronary syndrome which may present as sudden onset palpitation and diaphoresis. Additionally, clinical presentations like palpitation mimic many other clinical conditions, resulting in mistaken diagnosis [5]. In spite of that, dizziness and diaphoresis can be symptoms of PE even though it carries small percentage [2].

Palpitation, tachycardia and RBBB are the most frequent clinical characteristics for patients with acute PE [8-9]. Besides, in this case despite of complaint sudden onset of palpitation, her heart rate was normal and her ECG was similar to her baseline ECG. Despite that, presence of RBBB may give diagnostic value or marker for obstruction of main pulmonary trunk [9].

Her normal blood pressure throughout the hospital stay was also not a typical finding of acute massive PE that supposed to have hypotension [10]. Possible explanation is ES with pulmonary hypertension has the capability to compensate the right ventricular strain. Normal findings for the two-point compression test are expected in ES patient as they are prone for *in situ* thrombosis [11]. D-dimer may not be helpful because of they may have higher circulating level of this biomarker [12]. Relying too much on the low baseline oxygen saturation (SpO2 of 72±9%) based on the previous study [11] may also lead to misdiagnosis.

Her chest radiograph was compatible with ES, which described as the pruning of pulmonary arteries and cardiomegaly [7], but no finding suggested of PE. The ECHO findings for this patient were also consistent with right ventricular dysfunction which commonly seen in ES suffering from pulmonary hypertension. These presentations exert influence on treating physician from considering of PE.

Patients with heart disease are at greater risk of developing venous thromboembolism and most of them seemed to have isolated PE without deep venous thrombosis [12] as similar with this case whereby two-point compression test was negative.

Even though the features may resemble acute PE in normal patients, it may be tricky in patients with underlying ES, where those clinical features were their baselines. As the ES progresses, the impact of polycythaemia and hypercoagulable state endangers the patient with the risk of thromboembolic events. Extreme pulmonary hypertension may conceal the pulmonary thrombosis, hence impeding the diagnosis. Even massive PE may present with only minimal exacerbation of symptoms and signs compared to pre-existing conditions.

Conclusion

Minor complaints such as sudden onset palpitation, even in the absence of tachycardia, should be considered seriously. Diaphoresis and dizziness might represent associated symptoms with acute PE in an ES patient. Therefore, any trivial complaint should be taken undoubtedly as thrombolysis is a life-saving treatment if timely diagnose. In fact, many fatal deaths of acute PE were due to underestimated and underdiagnosed.

Competing interests

The authors declare no conflict of interest regarding this case report.

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Patients' Consent for the Use of Images and Content for Publication
Verbal consent was obtained from the patient for publication of this case report with the accompanying images.

What is new in this case report compared to the previous literature?

- Palpitation is common symptoms patient complain to health facilities. However, do not forget to exclude life-threatening conditions such as pulmonary embolism as a first impression in patients with a higher risk of thromboembolism.
- We should trigger our thinking regarding more serious conditions even though physical findings do not tally with the complaints or may be normal findings.
- Details investigation and high index of suspicious should apply in certain cases like this patient.
- A multi-disciplinary approach is necessary to improve patient evaluation and provide the best care to the patient.

Implication to Patient
In those with underlying structural heart disease, when they complain of sudden onset of new symptoms, the treating physician should first think of any life-threatening condition. Delay in working diagnosis leads to severe complications and carries high mortality to the patient.

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