Case report

Saddle pulmonary embolus in a young man with homocysteinemia

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Abstract

Acute pulmonary embolism (PE) is a life-threatening condition, requiring immediate management. This case report summarises the evaluation and management of a patient who presented with a saddle pulmonary embolus. Since the patient had features suggestive of significant right heart dysfunction, he was thrombolysed with streptokinase but developed cardiogenic shock while on the streptokinase infusion. Therefore, catheter fragmentation and thrombectomy was performed with a view to break and remove the clot. Despite this, his condition deteriorated and unfortunately he died in the ICU. The thrombophilia screen sent before death showed significantly elevated serum levels of homocysteine, which could potentially have caused a hypercoagulable state. This case re-emphasises the mortality associated with major PE and the need for a detailed thrombophilia screen in patients with deep vein thrombosis or PE without any major clinical risk factors.

Keywords: Homocyteinemia, pulmonary embolus.

Introduction

Saddle pulmonary embolus (SPE) is a large thromboembolus that lodges at the bifurcation of main pulmonary artery. It is usually less than massive but has the potential to cause sudden haemodynamic collapse. Immediate recognition and prompt treatment can significantly reduce mortality from SPE. In this case report, we discuss the management of SPE leading to cardiogenic shock in a young man. Although the outcome in this case was death, the finding of significant homocysteinaemia during the thrombophilia screen prompted us to publish this report.

Case Report

A 25 year old, obese (BMI 35 kg/m^2) professional presented to the emergency department with two days history of breathlessness and giddiness. He did not complain of any chest pain, cough or fever. Three days before the presentation, he had an episode of syncope in the toilet. There was no other significant history, except for a 12-pack year smoking history. In the emergency department he was fully conscious, tachypneic at 32 breaths per minute, needed supplemental oxygen to maintain oxygen saturation above 94%, tachycardic at 132/min but normotensive.

His electrocardiogram showed sinus tachycardia with the classical $S_1Q_3T_3$ changes suggesting

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right ventricular strain. A bedside screening transthoracic echocardiogram showed severely dilated right atrium and right ventricle (RV) with flattening of the interventricular septum. Therefore, PE was suspected and a CT pulmonary angiogram (CTPA) was ordered. He was started on therapeutic dose of low molecular weight heparin and was transferred to the medical intensive care unit for close haemodynamic monitoring and further management.

His initial blood investigations revealed a positive troponin and an elevated d-dimer at 7.06 μg/ml with normal haemogram, biochemistry and coagulation parameters. CTPA revealed an SPE *(Figure 1)* with RV strain. Thrombolysis was considered in view of the presence of SPE with hypoxia, RV strain and a positive troponin. A bolus of 250000 units Streptokinase (STK) was administered and an infusion commenced at a dose of 100000 units/h. Soon after admission to the ICU, he became hypotensive and developed cardiogenic shock. Since he developed cardiogenic shock while on streptokinase, it was decided to attempt catheter directed thrombus fragmentation and removal. He was taken to the interventional cardiology lab where the clots were fragmented and suctioned using a 6 Fr catheter. Despite this, the cardiogenic shock worsened and he died in the ICU soon after the procedure. The thromboembolism screen sent on admission revealed a homocysteine level of > 65 μmol/L.

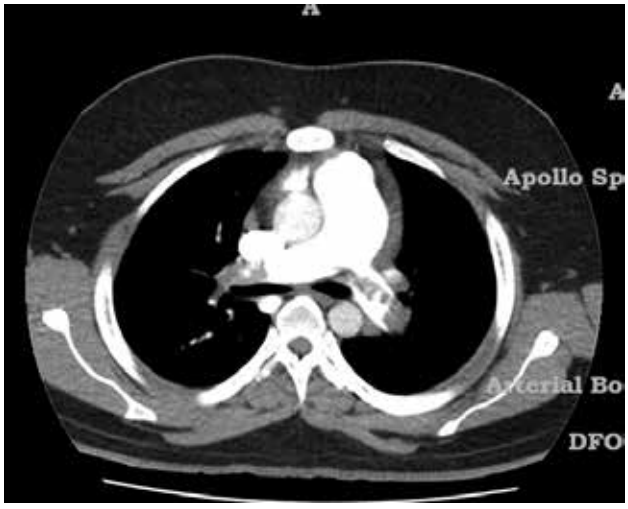

Fig. 1: CTPA showing saddle embolus

Discussion

SPE is seen on CTPA in 2-5% of all patients with radiologically confirmed pulmonary emboli (PE)[1]. However, the true frequency is difficult to determine, as many patients are too unstable for CTPA and several of them are identified only at autopsy. Although it indicates a large clot burden, one large retrospective study showed that only 8% of patients with SPE had persistent shock and 14% had transient hypotension.[1] Therefore, SPE is generally considered submassive or PE in transit. The clinical presentation of SPE varies widely from acute dyspnoea to frank cardiogenic shock. If it is associated with arterial hypotension and cardiogenic shock, the mortality can be as high as 34%.[1]

Several investigations are ordered to confirm PE, rule out other possible diagnoses and plan treatment. In most patients, a 12 lead ECG shows sinus tachycardia with non-specific T wave changes. The typical $S_1Q_3T_3$ pattern is seen only in 25-50% of the cases, but ECG is helpful in ruling out other life-threatening cardiac conditions.[2] A plain chest film might show nonspecific pulmonary oligaemia but more importantly it helps to rule out other causes of dyspnoea. Transthoracic echocardiogram shows enlargement of right heart chambers, flattening of the interventricular septum and dilatation of inferior vena cava (IVC) secondary to RV pressure overload. Acute pressure overload and right heart dilatation causes myocardial injury, which is reflected by an elevation of the troponin level. D-dimer is another frequently ordered investigation in patients suspected to have deep vein thrombosis (DVT) or PE. It has a strong negative predictive value and helps to rule out DVT or PE.[3] Our patient had S1Q3T3 on ECG, pulmonary oligaemia on chest x-ray and an enlarged right heart on transthoracic echocardiogram with an elevated troponin.

Massive PE, which is defined as PE with hypotension and cardiogenic shock, is a definitive indication for thrombolysis.[4] In submassive PE, if there is significant RV dysfunction and elevated troponin, the patient would certainly benefit from thrombolysis.[4] Since the available evidence suggests that severe RV dysfunction in PE is associated with worse prognosis, thrombolysis must be considered in such patients.[5]

Our patient was not hypotensive at presentation, but because of the severe RV dysfunction and positive troponin, it was decided to administer thrombolysis. Although tPA is the commonly used drug, STK and urokinase are also approved for thrombolysis in PE. Superiority of one agent over the other has not been established in clinical studies, but there is a suggestion that shorter drug infusions achieve rapid clot lysis compared to longer infusions.[3] Since tPA is given over 2 hours as compared to 24 hours for STK, it is commonly chosen[3]. Streptokinase was chosen for our patient because it is less expensive and easily available.

After the patient is clinically stable, investigations to identify the source of the clot must be carried out. They include bilateral lower limb venous doppler for deep vein thrombosis (DVT) and imaging of the abdomen and pelvis, if clinical examination suggests the possibility of an intra-abdominal pathology. A thrombophilia screen is ordered if there are no specific major known clinical risk factors for DVT, especially if the patient is young. An initial screen in such a situation would include Factor C and S assay, antithrombin III levels, lupus anticoagulant and homocysteine levels. In our patient serum homocysteine level was significantly elevated. Moderate hyperhomocysteinemia has been shown to be an independent risk factor for atherosclerotic and thromboembolic disease.[6,7] Various mechanisms including endothelial dysfunction and excessive platelet aggregation have been proposed as mechanisms for the procoagulant effect of homocysteine.[7] Deficiency of folate, B_{12} and B_6 are associated with elevated levels of homocysteine[8] and diets rich in fruits and vegetables, folic acid, B_6 and B_{12} supplementation have all been shown to decrease the levels of homocysteine.[9,10] Studies have shown that reducing the levels of homocysteine might offer clinical benefit.[10]

To conclude, SPE is associated with significant mortality in patients with cardiogenic shock and in patients without major clinical risk factors for DVT or PE, a detailed thrombophilia screen might help in identifying the cause.

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