An Atypical Presentation of Massive Pulmonary Embolism

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Abstract

Background: Pulmonary embolism (PE) is an obstructive disease of the pulmonary arterial system caused by the embolization of thrombus originating from the deep veins of the lower extremities [1]. PE is the third leading cause of cardiovascular related deaths after coronary arterial diseases and stroke [2]. Its incidence rises in older age group [3]. The frequency of developing pulmonary embolism in young patients is low but once developed it has a high potential for mortality if not diagnosed and managed early. Hyper-coagulable states like protein C and anti-thrombin III deficiency have been reported to cause cerebrovascular thrombosis but rarely been reported to cause PE. Case Report: We present a case of a 27 year old male who presented to the Emergency Department with complaints of low backache and giddiness. Patient was found to be tachycardic, tachypneic and in shock. Patient had a low probability of PE with a Well’s score of 1.5 but was diagnosed as having massive bilateral acute pulmonary embolism with deep vein thrombosis secondary to protein C deficiency. Conclusions: It is imperative for emergency physicians to have a high index of suspicion in young patients presenting with atypical symptoms and low clinical probability for PE in order to thrombolysed the patient on time.

Keywords: Adult, Pulmonary Embolism, Protein C Deficiency, Pulmonary Artery, Venous Thrombosis.

Introduction

Pulmonary embolism (PE) is an obstructive disease of the pulmonary arterial system caused by the embolization of thrombus originating from the deep veins of the lower extremities [1]. PE is the third leading cause of cardiovascular related deaths after coronary arterial diseases and stroke [2]. Its incidence rises in older age group [3]. The frequency of developing pulmonary embolism in young patients is low but once developed it has a high potential for mortality if not diagnosed and managed early. Hyper-coagulable states like protein C and anti-thrombin III deficiency have been reported to cause cerebrovascular thrombosis but rarely been reported to cause PE. It is often a dilemma for emergency physicians whether or not to pursue the diagnosis of PE in patients who present with atypical signs and symptoms of pulmonary embolism. Despite diagnostic advances, delay in diagnosing pulmonary embolism is common and represent an important clinical issue. We report a case of a young male who presented with atypical symptoms with no co-morbidities and was diagnosed with PE and treated for the same.

Case Report

A 27 years old male patient was brought to the Emergency Department (ED) with history of left low backache since 2 days. He also complained of giddiness followed by a fall on the presenting day. There was no history of fever, chest pain or shortness of breath. The patient did not have any significant past medical or surgical history. He was an occasional smoker and there were no significant family history.

On examination the patient was tachycardic, tachypneic and in shock with a heart rate of 140/min, respiratory rate of 34/min and unrecordable initial blood pressure. Patient was normo-thermic and saturating at 80% on room air. On secondary
survey patient was pale and there was a lacerated
wound over left occipital region and bilateral
rhonchi were present on chest auscultation with
bilateral equal air entry. Rest of the examination
was unremarkable. Patient was started on oxygen
inhalation and intravenous fluids to which the
patient responded well.

ECG was done which showed sinus
tachycardia and a chest x-ray showed haziness
in the left lung base. To rule out a cardiac cause
2D echocardiography was done which showed
dilatation of the right atrium and right ventricle
and trace tricuspid regurgitation with normal
ejection fraction [Fig.1]. Patients Well’s score was
1.5 which comes under low probability group but
keeping in view his echocardiography findings, he
was further evaluated for PE. He had a D-dimer
of >5000 ng/mL (0.0-750.0) and subsequently
Computed Tomography pulmonary angiography
(CT PA) was planned. The CTPA showed soft tissue
density filling defect noted in the distal segment of
the left main pulmonary artery extending upto the
sub-segmental divisions of almost all lobes of left
lung while on right side there was mild eccentric
filling defect in the right middle lobar division not
extending upto the sub-segmental level [Fig.2-5].
The patient was diagnosed with bilateral acute
massive thromboembolism and was thrombolysed
with tenecteplase. Doppler study of the lower limbs
revealed a large echogenic thrombus in the right
common femoral vein [Fig.6]. Patient was shifted
to the Cardiac Care Unit for further management.
Subsequent blood work revealed the patient to be
deficient in protein C as 33% (70-140) and anti-
thrombin III as 35% (83-128).

Patient was further managed conservatively
with anti-coagulants and other supportive
management. He responded well to given medical
therapy and was discharged in a stable condition
with the final diagnosis of massive acute bilateral
pulmonary thrombo-embolism and deep vein
thrombosis (DVT) secondary to protein C
deficiency.

Discussion

The risk of blood clots is increased by cancer,
prolonged bed rest, smoking, stroke, certain genetic
conditions, estrogen-based medication, pregnancy,
obesity, and after some types of surgery [4]. About
90% of emboli are from proximal leg DVTs or
pelvic vein thromboses. Clinically apparent DVT
is present in only 11% of confirmed cases of
pulmonary embolism. The classic presentation
of PE is the abrupt onset of chest pain, breathing difficulty and hypoxia but some patients may have no obvious symptoms at presentation. The diagnosis of pulmonary embolism should be suspected in patients with respiratory symptoms unexplained by an alternative diagnosis. Evidence-based literature supports the practice of determining the clinical pre-test probability of pulmonary embolism before proceeding with diagnostic testing [5]. The three validated systems include the Modified Wells Scoring System, the Revised Geneva Scoring System, and the Pulmonary Embolism Rule Out Criteria (PERC) [6-8]. Low probability PE can be ruled out with D-dimer testing [9]. CTPA is the gold standard for diagnosing pulmonary embolism [10].

A hyper-coagulation workup should be performed if no obvious cause for embolic disease is apparent and the patient does not have any risk factors for the same. Protein C is a 62-kD, vitamin K-dependent glycoprotein synthesized in the liver. The activation of the protein into activated protein C (aPC) is catalyzed by thrombin when it is bound to the endothelial glycoprotein thrombomodulin [11,12]. The catalytic activity of aPC is greatly enhanced by the vitamin K-dependent cofactor protein S [13]. A deficiency of protein C disturbs the delicate balance between pro-coagulant and anti-coagulant proteins and engenders a pro-thrombotic state. Protein-C and anti-thrombin III deficiency leads to a three-fold to seven-fold increase in risk of thrombosis [14]. Protein C deficiency however leading to PE has rarely been reported. For almost one-quarter of PE patients, the initial clinical presentation is sudden death [15]. Early thrombolytic therapy has been shown to have beneficial outcomes in patients having massive PE.

Conclusion

In our case report, the patient had a Well’s score of 1.5 and was found to have massive PE on CTPA along with DVT secondary to protein C deficiency. It is imperative for astute emergency physicians to have a high index of suspicion in young patients presenting with atypical symptoms and low clinical probability for PE in order to thrombolyse the patient on time.

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