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Acute pulmonary thromboembolism – a rare case series and a short review

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Abstract:
Acute pulmonary thromboembolism is one of the common causes of vascular death after myocardial infarction and cerebrovascular accident. It is one of the leading preventable causes of death in hospitalised patients. We present three rare cases –

1. *Septic thromboembolism with factor V Leiden mutation*
2. *Idiopathic pulmonary hypertension with acute pulmonary thromboembolism*
3. *Pulmonary thromboembolism due to protein S deficiency*

Early diagnosis of such rare diseases and prompt intervention led to successful outcome in all our cases.
Keywords*: Thromboembolism, pulmonary, rare causes*

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I. Introduction:

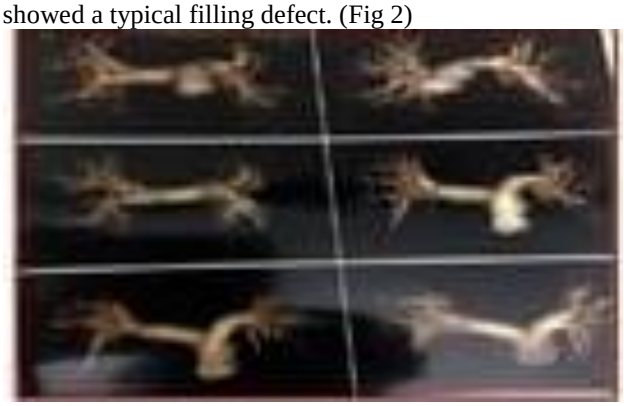
The diagnosis of pulmonary embolism (PE) remains challenging always for emergency physicians. Symptoms can beeither vague or non-existent, and the clinical presentation shares features with many other common diagnoses.

Diagnostic testing is usually complicated, as biomarkers like the D-dimer are frequently false positive with imaging like computed tomography pulmonary angiography, carries risks of radiation and contrast dye exposure. It is therefore necessary for emergency physicians to be both vigilant and thoughtful about this diagnosis[1]. In this series, we present three rare cases of acute pulmonary thromboembolism

Case 1: A seventy five year old farmer by occupation presented to us with shortness of breath, chest pain, and frothy sputum of one day duration. He had swelling of left leg with pain for two to three days. There was nohistory of fever. He was not a known case of diabetes mellitus or hypertension. On clinical examination, the patient was tachypneic, tachycardic with an oxygen saturation of 82%. There were mild crepitations in the right base. A possible diagnosis of acute pulmonary embolism was made. The complete blood count, liver function test, renal function tests electrolytes were normal. The chest XRay showed wedge shaped opacity in right mid zone (Fig1) the ECG and ECHO heart were not having significant findings. There was a reverse hallow sign in CT chest. The pulmonary angiogram showed a typical filling defect. (Fig 2)

Fig1 showing X Ray chest PA view with wedge opacity.

The pulmonary angiogram showed a typical filling defect. (Fig 2)



The D Dimer value was 800ng/l. ANA and APLA assay negative. RA factor negative.Protein C antigen 101, protein S antigen 132.8.Plasma antithrombin III 0.242g/dl.Serum homocysteine 12.36 micromols/l. Venous Doppler showed superficial thrombophlebitis with perforator incompetence in the medial aspect above the left ankle joint. Right leg was normal. Patient was started on Heparin 5000 units TDS s/c and Acitrome 2mg OD. Patient was given supplemental oxygen and noninvasive ventilation intermittently. Patient became better both in terms of symptoms and oxygenation. Factor V Leiden mutation was suspected and managed. Hence this case is reported as a rare case of septic thromboembolism with factor V Leiden mutation.

CASE 2

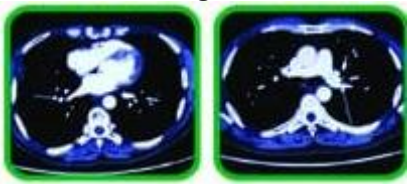
A 29 year old female presented with shortness of breath for 2 months, progressed to grade IV. There was history of hemoptysis around 100 ml for 1 day. She was not a known case of hypertension, bronchial asthma, coronary heart disease or pulmonary tuberculosis. She was married and having 2 children. On examination she was anemic, tachypneic with a heart rate of 110/min. On cardiovascular system examination, there was a loud P2 with no murmur. Routine blood investigations like CBC, RFT, LFT, electrolytes, HIV were negative. Sputum AFB negative. ECG showed right axis deviation, right atrial and ventricular enlargement with S1 Q3T3 pattern

ECHO showed RA/RV dilatation with severe pulmonary hypertension. Plasma D dimer 562 ng/ml. CT chest showed ground glass haziness over left base which coincided with findings in CT pulmonary angiography. Bilateral venous doppler in the lower limbs were normal. ANA and APLA were negative. Protein C antigen 104.4 and Protein S antigen 118. Factor V Leiden mutation not detected. Patient was started on Oxygen, Heparin bolus followed by infusion 18 units/kg/hr. Tablet Bosentan 62.5 mg PO BD, Tab Tadalafil 20 mg OD, Acitrome 2 mg OD. Patient became symptomatically better and repeat ECHO showed a decrease in pulmonary gradient pressure from 100 to 65.

CASE 3

A 50 year old male presented with complaints of right sided chest pain and shortness of breath for 10 days. There was a history of blood streaked sputum for 10 days. He was a known case of diabetes and hypertension 3 years, smoker and alcoholic for 24 years. There was tachypnea, mild tacyhcardia with hypoxia with saturation 90-92%. Respiratory system was normal except minimal right infrascapular creptitations. Other systems were normal. Routine blood investigations like CBC, RFT, LFT normal. RBS 250, Sputum AFB negative. ECG S1q3T3 pattern. ECHO, USG abdomen, venous doppler of bilateral lower limbs normal. D dimer 4542.5 FEU mcg/l. Protein S 39% (normal 77-143%). Other protein assays normal. Partial filling defects in the right descending pulmonary artery extending to lower lobe segmental branches, partial thrombus in left pulmonary artery bifurcation.

Fig 3



CT angiography findings shows typical filling defect

This patient was diagnosed as pulmonary thromboembolism with protein S deficiency with diabetes and hypertension. Patient was managed with oxygen, bronchodilators, oral anti diabetic and antihypertensives.

Tablet acenocoumarol was started. Patient improved symptomatically and was discharged with advice for regular follow up.

II. Discussion

Pulmonary embolism (PE) is the third most common cause of cardiovascular death after myocardial infarction and stroke.

PE occurs when clots formed in the deep venous system dislodge or break loose, travel through the heart, and become lodged in the pulmonary vasculature. While small PE can frequently lyse spontaneously, larger PE can cause a sudden and persistent rise in pulmonary artery pressure, which can lead to circulatory collapse[2].

There are multiple factors that can increase one's chance of developing DVT and PE. These factors can beeither inherited or acquired. One of the common causes of inherited conditions is factor V Leiden mutation with a prevalence of 4%. The commonest acquired risk factors for Venous ThromboEmbolism include: increasing age, venous insufficiency, obesity, smoking, rheumatologic or cardiovascular disease, previous VTE, and antiphospholipid antibody syndrome[3]. In all our cases, there was mild hypoxia which was correctable with oxygen therapy and occasional noninvasive ventilation. There was no circulatory collapse in all our cases.

In our first case of septic thromboembolism with factor V Leiden mutation, the prognosis was good because of good LV function without RV dilatation[4].

In our second case, the prevalence of pulmonary arterial hypertension and idiopathic acute pulmonary thromboembolism is 5.9 cases per million adult population. Usually, the presentation of age is 35-40 years, which in our case was a 29 year old. The literature shows a female predominance in such conditions.

Table 1 showing differences between primary pulmonary hypertension and pulmonary hypertension with thromboembolism[5]

VARIABLES	PPHT	CPTE-PHT
Age	20-40	>50
Female Male Ratio	4:1	1:1
Clinical Course	Continued deterioration	Intermittent stabilization
Perfusion Lung Scans	No segmental perfusion defects	Segmental/large perfusion defects
Pulmonary artery systolic pressure	>60mmHg	<60mmHg
Pulmonary Angiogram	Pruning	Intraluminal filling Defects
Confounding problems with angiogram.	Thrombi may occur on or distal to PPh lesion	Pruning can also suggest PE
Therapy	Anticoagulation	Anticoagulation

Our case is a rare case of idiopathic pulmonary hypertension with pulmonary thromboembolism. As the symptoms and signs are non-specific, a delay in the diagnosis is usual. Any delay worsens the prognosis and with a strong suspicion and early diagnosis, we can have better outcomes in such cases.

In our third case, we had pulmonary thromboembolism due to protein S deficiency. Protein S is a vitamin K dependent anticoagulant synthesized in the liver. As it was first discovered in Seattle - Washington, the name protein S was coined. It is encoded by the gene PROS I in long arm of chromosome 3. Protein S circulates in bound form (60%) which is non - functional and 40% in free form which is functional. Major function of the protein S is to act as a co factor, it facilitates the action of APC. It functions independently also to directly inhibit factor X activating complex and prothrombin activating complex. Protein S deficiency is a rare blood disorder with a two to eleven times increased risk of thromboembolic events. Men and women are equally affected. The incidence is more in the Japanese[6]. In our case, he was a diabetic and hypertensive with shortness of breath which prompts our mind towards coronary events rather than pulmonary thromboembolism. Hence, as in our case, which showed S1 q3t3 pattern on ECG, we ended up with a suspicion of acute pulmonary thromboembolism with a routine check up of all the possible etiologies, we were able to spot the protein S deficiency. Early diagnosis and prompt management led to a successful recovery in our case. Isolated segmental abnormalities in CT Pulmonary angiograms may be due to pulmonary embolisms even though false positives may occur. We had other clear evidences of acute pulmonary thromboembolism in addition to angiogram findings[7]. D dimer testing may be of limited value because about half of the patients may have positive test due to pulmonary embolisms, but a normal D dimer level may rule more in favour of ruling out pulmonary embolism[8]. Anticoagulant therapy along with symptomatic treatment is adequate in most of the cases as our patients were cardio stable, we did not attempt systemic thrombolysis or embolectomy.

III. Conclusion:

We present 3 rare cases of acute pulmonary thromboembolism with a strong index of suspicion.we diagnosed each of them very early to intervene in a proper manner to prevent serious complications.

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