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Case Report

A rare case of pulmonary thromboembolism with lobar pneumonia in a factor 7 deficiency patient

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ABSTRACT

While thrombotic events are uncommon in individuals with Factor VII deficiency, they have been documented, particularly when other prothrombotic risk factors are present, such as recent surgical interventions, trauma, or factor replacement therapy. This case report discusses a 45-year-old male with a known history of Factor VII deficiency who initially presented with symptoms of fever and productive cough, leading to a diagnosis of lobar pneumonia based on clinical examination and chest X-ray results. However, after four days of hospitalization, the patient exhibited persistent coughing, occasional hemoptysis, and breathlessness, raising clinical suspicion of pulmonary thromboembolism (PTE), which was later confirmed via computed tomography pulmonary angiography. Treatment with anticoagulation led to significant clinical improvement. This case illustrates that despite having Factor VII deficiency, the patient developed PTE.

Keywords: Factor 7 deficiency, Pneumonia, Pulmonary embolism

INTRODUCTION

Factor VII deficiency was first described in the medical literature by Dr. Alexander, et al. in 1951 and was referred to as prothrombin conversion accelerator deficiency. The disorder has also been known as Alexander's disease.[1] It is a rare genetic bleeding disorder characterized by a deficiency or reduced activity of clotting factor VII.[2] Factor VII (FVII) deficiency is the most frequent among rare congenital bleeding disorders, accounting for one symptomatic individual per 500,000 population, apparently without any racial/ethnic predilection. FVII deficiency prevalence in the general population is probably higher because of the presence of asymptomatic and poorly symptomatic individuals.[3] FVII deficiency, the most common among the rare congenital coagulation disorders, is transmitted with autosomal recessive inheritance.[4] Clotting factors are specialized proteins that are essential for the blood to clot normally. Individuals with factor VII deficiency can experience prolonged, uncontrolled bleeding episodes. The severity of factor VII deficiency can vary greatly from one person to another. Some individuals may have no symptoms (asymptomatic); others may develop mild, moderate or potentially severe, life-threatening complications as early as in infancy.[1]

Thrombotic events in bleeding disorders such as hemophilia A or B, Von Willebrand disease, afibrinogenemia, factor VII deficiency, and factor XI deficiency are rare but have been reported. These events usually occur in the presence of prothrombotic risk factors such as recent surgery, trauma, or factor replacement therapy and infections.[5] Paradoxically, FVIID does not protect affected patients from venous thromboembolism (VTE).[6] We report a case of thrombosis in a patient with FVIID in the setting of pneumonia.

CASE REPORT

A 45-year-old male presented with a four-day history of fever, breathlessness, palpitations, and productive cough. He had been in his usual state of health until four days prior when he experienced an episode of fever accompanied by chills and rigors, which subsided temporarily with medication.

The following day, he developed exertional breathlessness, which progressively worsened to include breathlessness at rest over the next three days. As his condition deteriorated, he sought care at the hospital. Upon evaluation in the emergency room, his room air saturation was 80%, respiratory rate was 26 breaths per minute, and blood pressure was 80/50 mmHg. Initial management included oxygen support, fluid resuscitation, and initiation of inotropes due to persistent hypotension.

Based on clinical presentation and chest X-ray findings (Figure 1), he was diagnosed with lobar pneumonia and commenced on empirical antibiotics while awaiting results from sputum and other routine investigations (Table 1).

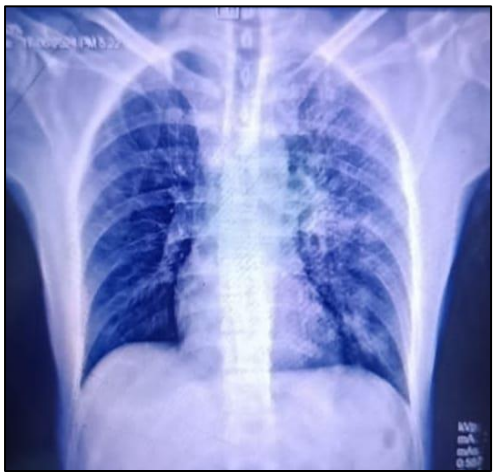

Figure 1: Chest X-ray of patient showing left sided upper and middle zone haziness.

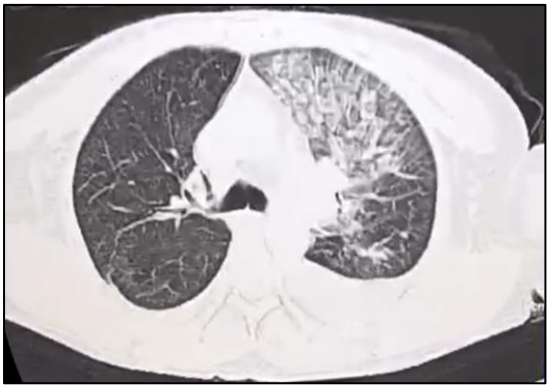

Figure2: CT Image showing left side upper lobe ground glass opacities.

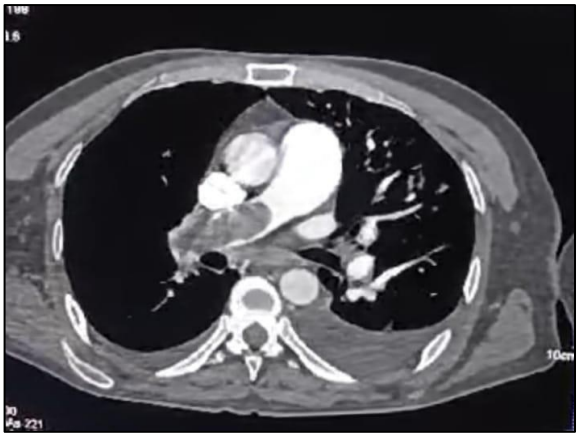

Figure 3: CT Image showing large thrombus in right pulmonary artery.

The patient's past medical history revealed a diagnosis of Factor VII deficiency made ten years ago during bilateral hip replacement surgery for osteoarthritis. Despite this, he had no history of bleeding tendencies or hospital admissions related to his condition, and there was no family history of similar disorders.

After two days of admission, patient recovered from shock with decreased episodes of fever and recovered from acute kidney injury. However, he continued to exhibit persistent cough and could not maintain oxygen saturation without support, desaturating whenever oxygen was reduced, accompanied by increased respiratory effort. Clinical examination revealed clear lung fields on the right, with minimal fine crepitations on the left.

Table 1: Lab parameters.

Parameters	On admission	On day 4
Haemoglobin (gm/dl)	10.2	10
Hematocrit (%)	37	38
WBC (per µl)	20000	12000
Differential count		
Neutrophils	90 %	68 %
Lymphocytes	6 %	29 %
Monocytes	3%	2 %
Eosinophils	1%	1 %
basophils	0 %	0%
Platelet count (per µl}	3,50,000	3,00,000
Urea	68	24
Creatinine	2.04	1.2
Lft	WNL	WNL
Serum electrolytes	WNL	WNL
Sputum aFB	Negative	
Sputum cbnaat	Negative	
Sputum culture	*Klebsiella pneumonia*	
Blood culture	No growth	
HIV	Negative	

On the third day, he developed a severe cough, producing sputum occasionally streaked with blood. A bedside 2D echocardiogram indicated right atrium and right ventricle dilation along with severe pulmonary hypertension. His electrocardiogram showed sinus tachycardia. A Wells score of 5.5 suggested a moderate risk of pulmonary embolism (Table 2).

Table 2: Wells criteria.

S. no.	Criteria	Points
1.	Clinical signs and symptoms of DVT	3
2.	PE is most likely the diagnosis	3
3.	tachycardia (HR >100)	1
4.	HEMOPTYSIS	1.5
5.	recent surgery or immobilization	1.5
6.	prior DVT or PE	1.5
7.	Active malignancy (last 6 months)	1

Subsequently, CT pulmonary angiography confirmed a nearly complete thrombus occluding the right pulmonary artery, extending into the upper lobar, interlobar, and subsegmental branches. He was treated initially with heparin for five days, followed by ongoing therapy with dabigatran. Despite his underlying factor VII deficiency, the patient developed pulmonary thromboembolism."

DISCUSSION

Inherited deficiencies of coagulation factors typically lead to a lifelong bleeding tendency. However, paradoxically, even in disorders with a pronounced bleeding risk, such as hemophilia and Factor VII deficiency, thrombotic events-both arterial and venous-can still occur. Thrombosis in hemophilia patients, for instance, often has a multifactorial origin, linked to the use of long-term central venous catheters, intensive replacement therapy during surgical procedures, bypassing agents, or the presence of additional prothrombotic factors, whether acquired or inherited. For patients with a history of thrombotic events, conducting a thrombophilia screen to identify any coexisting prothrombotic conditions is advisable, and managing cardiovascular risk factors remains crucial.[7]

Various recognized risk factors for venous thromboembolism (VTE) include recent surgery, prolonged immobility, malignancy, advanced age, obesity, hormonal influences (e.g., oral contraceptives), a prior history of thromboembolism, and inherited thrombophilic conditions.[8,9] Infections can also enhance the risk of VTE by triggering systemic inflammation, which may lead to one or more of Virchow's triad of thrombosis mechanisms: venous stasis, hypercoagulability, and endothelial damage.[10-13] Research has identified an increased risk of VTE in patients with infectious conditions like cytomegalovirus, Chlamydia pneumoniae, HIV, hepatitis C, and respiratory tract infections.[14-21]

The occurrence of pulmonary thromboembolism (PTE) in this case challenges the conventional understanding of Factor VII deficiency, which is more commonly linked with hemorrhagic manifestations than with thrombosis. This case emphasizes that Factor VII deficiency does not inherently provide protection against thrombotic events, especially when other risk factors such as infections or systemic inflammation are present.

Our findings are consistent with earlier studies, such as those by Cohoon et al, who found a significant association between lower respiratory and genitourinary infections and thromboembolism.[22] This case further contributes to the growing evidence that bleeding disorders do not universally safeguard against thrombosis. It underscores the importance of maintaining a high index of suspicion for thrombotic events, even in patients with known bleeding disorders, particularly when other predisposing factors are present.

Prompt diagnostic imaging should be considered when there are clinical signs suggestive of thromboembolism, irrespective of the underlying bleeding condition.

CONCLUSION

This case serves as a critical reminder that clinical vigilance is necessary when managing patients with bleeding disorders who present with atypical symptoms. The occurrence of PTE in a patient with Factor VII deficiency is rare but possible, especially in the context of additional prothrombotic conditions like infection. Effective management requires a nuanced understanding of both the bleeding risks associated with Factor VII deficiency and the potential for thrombotic complications, ensuring that both are appropriately addressed to optimize patient outcomes.

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