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*RESEARCH ARTICLE*

# BILATERAL PULMONARY EMBOLISM MIMICKING ACUTE CHEST SYNDROME IN A PATIENT WITH SICKLE CELL DISEASE: ACASE REPORT AND REVIEW OF THE LITERATURE

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## *Abstract*

This case report discusses a 24-year-old female with sickle cell disease (SCD) who developed bilateral pulmonary embolism (PE), mimicking acute chest syndrome (ACS) following a cholecystectomy. Despite initial treatment for ACS, the patient's condition deteriorated, leading to a diagnosis of massive bilateral PE via chest CT angiography. The report underscores the importance of considering PE in SCD patients with respiratory symptoms unresponsive to standard ACS treatment. It highlights the hypercoagulable state in SCD, which increases the risk of venous thromboembolism, including PE. The case emphasizes the necessity for heightened clinical suspicion and appropriate diagnostic measures to improve patient outcomes. Unfortunately, the patient succumbed to septic shock despite aggressive management. This report advocates for comprehensive evaluation strategies in similar clinical scenarios to prevent misdiagnosis and improve survival rates.



## Introduction:

Sickle cell disease (SCD), a group of inherited hemoglobinopathies characterized by mutations that affect the β-globin chain, affects more than 3 million people worldwide [1,2]. These mutations result in abnormal hemoglobin polymerization, leading to a cascade of physiological consequences, including erythrocyte rigidity, vaso-occlusion, chronic anemia, hemolysis, and maculopathy [3], leading to a state of hypercoagulability that can be a risk factor for pulmonaryembolism (PE).PE is a potentially fatal disease that requires prompt diagnosis and early treatment, especially among patients with SCD,and symptoms can incorrectly point to an acute chest syndrome (ACS).

As this case demonstrates, pulmonary embolism should be considered in the differential diagnosis of any sickle cell patient who presents with respiratory symptoms,especially those with other predisposing factors.

**Case report:**

A 24-year-old woman with a history of SCD since age eightwas admitted to the medical ICUfor acute respiratory distress.She complained of stage V Sadouldyspneafive days before admission associated with feverand left chest painwithoutsyncope,fainting, or other signs. She denied any trauma or recent respiratory infection.The examination at admission showed a conscious patient with symmetric and reactive pupils.The pulmonary examination showed a symmetrical thorax, arespiratory rate of 30 breaths per minute, oxygensaturation measured by pulse oximetry 92% with15 liters of oxygen with left basal thoracic crackles, and no fluid effusion syndrome.Cardiovascularexamination showed a blood pressure of 130/80 mmHg, aheart rate 125 beats per minuteandno symptom of heart failure. The abdominal examination showed a soft and slightly distended abdomen.She had no swallowed extremities and had a

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negative Homanssign.Thepatientwas placed on non-invasive ventilation (PEEP 6 $cmH_2O$, Inspiratory support 12 $cmH_2O$,fraction of inspired oxygen ($FiO_2$) 100% initially then decreasedto 60% after stabilization);Her hemoglobin was 8.4 g/dL; hematocrit 22% and platelets 266000 /$mm^3$, white blood cells 29800 /$mm^3$, prothrombin time 12 secondsand international normalized ratio 1.1.Primary infectious assessment was negativeincluding a C-reactive protein of 1.7 g/l, procalcitonin 0.11ng/ml, negative respiratory RT-PCR, negative urine culture, and blood culture.The chest radiographshowed segmental atelectasis and consolidation (Fig. 1).

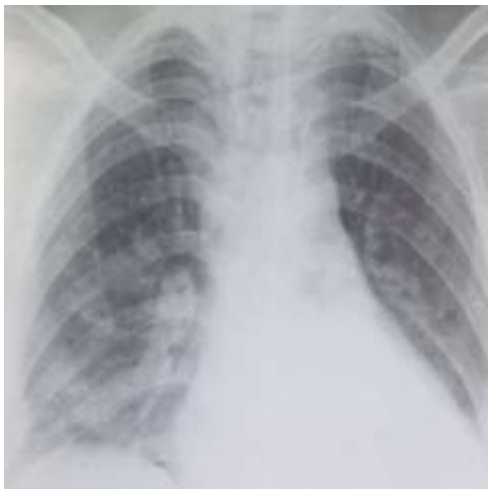

**Figure 1:-** Admission chest X-ray.

She was treated for an acute chest syndrome, received four red blood cell transfusions, phenotyped, and leucoreduced without incident. Despite the initial improvement in symptoms, twodays later, dyspnea progressed with tachypnea of 35 breaths per minute, signs of respiratory distress, andfever 39 °C.The arterial blood gas measurement showed a pH of 7.55, hypocapnia 22 mmHg, hypoxemia 45 mmHg. The patient was intubated. Chest CT angiography confirmed a massive bilateral pulmonary embolism with signs of pulmonary arterial hypertension and probable foci of ventral pulmonary infarction of the right upper lobe (Fig. 2).

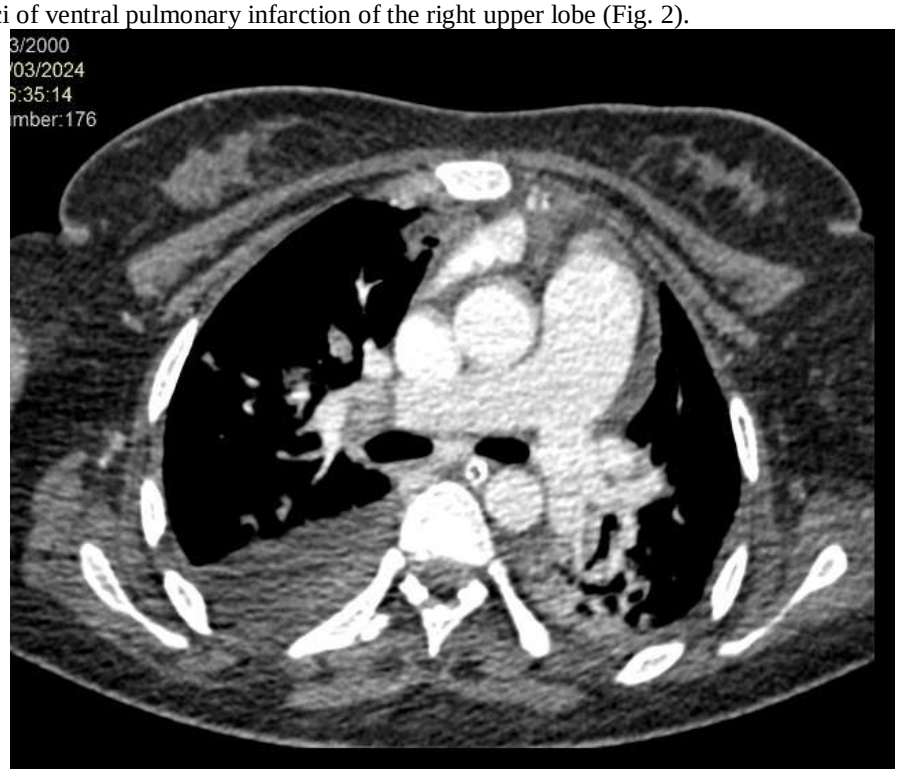


**Figure 2**:-Chest CT angiography showing an emblism ofboth left and right pulmonary arteries.

Anticoagulation with low molecular weight heparin (LMWH) (enoxaparin) (100 anti-Xa IU/kg/12 hours) was started immediately. One week later, she presented signs of hypoperfusion,increased heart rate (142 beats per minute),hypotension(64/42 mmHg), cold extremities,and reduced urine output (<0.5mL/kg/hour)and fever (40 °C).Antibiotics, norepinephrine, and hydrocortisone were administered.Blood and urine cultures were positive for E.coli. However, the patient died two days laterdue to a refractory septic shock.

## Discussion:-

Acute chest syndrome (ACS) is one of the most severe complications in adult patients with SCD. Itis the most common cause of death and the second most common cause of hospitalization in patients with SCD [4].

Diagnostic criteria include cough, wheezing, hypoxemia (PaO2 < 60 mm Hg), tachypnea, chest pain or fever > 38.5°C (101.3°F) associated with a new infiltrate on pulmonary imaging other than atelectasis, in the absence of an alternative cause such as volume overload [5, 6]. The chest radiograph is considered the gold standard for imaging modality [6].

Risk factors are varied, and it is generally difficult to specify the exact factor that generates the crisis. Infectionisamong the most risk factors [7-8], asthma and hyperresponsiveness (especially among children)[9], genotype (HbSS, Hb-SC), young age (2-4 years),hypoxia,surgery and anesthesia,smoking,andpulmonarythromboembolism [4].

Venousthromboembolism (VTE) in SCD appears to be caused by a hypercoagulable state due to multiple proposed mechanisms, including enhanced platelet function [10], activation of the coagulation cascade, and impaired fibrinolysis [10]. Although this multifactorial hypercoagulability is a risk factor for thrombosis formation, PE is rarely cited as a possible complication of SCD [11]and is likely not diagnosed [12].Graham et al. observed PE in 38.1% from autopsies of 21 patients with SCD after sudden death [13]. Darbari et al. reportedPE in 14.9% of autopsiesfrompatientswithSCD [14].Symptoms of PDare often unspecific and can be wrongly referred to as ACS, and treated as such,although it is not thought to be a cause of PD, some studies show that 17% of patients with ACS develop PE [11].

VTE is common in patients with SCD and is primarily due to frequent hospitalization, central venous catheters, and surgeries, especially orthopedic surgeries for avascular necrosis [4], but SCD has not been identified as a risk factor in any series of pediatric or adult PE[15].

Treatment for ACS and PE differs;ACS management of ACS is based on Hydroxyurea or L-glutamine, red blood cell (RBC) transfusion [16],antibioticsalongwith adequate pain control, supplemental oxygen, and fluid management. Anticoagulation is not used therapeutically in ACS, and since the treatment of a PE can greatly reduce mortality [17], a high index of suspicion should be maintained in sickle cell patients with chest pain.

## Conclusion:-

Patients with sickle cell disease have a hypercoagulable state and are at risk for venous thrombosis and pulmonary embolism. This association is rarely listed in the literature.Our case demonstrates that patients with sickle cell disease who do not improve with standard ACS treatment modalities should be evaluated for PE.

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