CASE REPORT

# Lungs Under Siege

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## Abstract

Sickle cell disease (SCD) is a genetic disorder with unusual shaped haemoglobin that potentially causes red blood cells contorted. Patients with SCD are severely prone to acute chest symptoms, particularly among paediatric patients. The present case study article explores the causes, mechanisms, and management of acute chest syndrome (ACS) through a case study of a boy (19 age) with SCD presenting ACS. Multidisciplinary collaboration and adherence to established guidelines are crucial in optimising patient outcomes. The case highlights the importance of early recognition, comprehensive evaluation, and prompt intervention in ACS management.

**Keywords:** Acute chest syndrome, Case report, Haemoglobin abnormalities, Haemoglobin SS, Lung complications, Pulmonary complications, Red blood cell deformity, Sickle cell disease.

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## Introduction

Sickle cell disease (SCD) is also known as haemoglobin SS, which is a rare blood-borne genetic disorder distinguished by the abnormal synthesis of haemoglobin, Which results in the formation of sickle-type RBC cells.[1] Various research study states that primary genetic material behind formation of SCD is mutated beta-globin (HBB), that is accountable for the haemoglobin synthesis.[2]

Acute chest syndrome (ACS) is a common pulmonary issue in people with SCD, occurring frequently. About 40% of these cases can be traced to specific causes in children. Among these cases, around 40% are associated with infections, including viral illnesses, mycoplasma-related pneumonia, and chlamydia-related.[3]

According to the study done by Takahashi T et al. in 2018 and Jain S et al. in 2017, stated ACS commonly affects paediatric patients aged between 2 and 4 years. In elders, over 70% of ACS cases arise due to vessel obstruction, if this condition prevails it will be one of the leading cause of death in individuals with SCD, i.e., in about 25% of all deaths. The death associated with ACS is estimated to be 4.3% in adults and more than 1.1% in children.[4,5]

The prognosis of ACS in SCD recipients is rapid, underscoring its status as the causative factor behind the demise of SCD patients. The present case study report examines the causes, mechanisms, assessment, and medication of ACS, emphasising more collaborative role of diverse healthcare professionals in managing patients affected by this condition.

## Case Study

A 19-year and 5-month-old boy with a confirmed diagnosis of sickle cell anaemia exhibited with a primary complaint of retrosternal chest discomfort and back pain persisting for 48 hours. Notably, the patient had undergone splenectomy three years earlier in 2021.

### Presenting Complaints

Two days ago, he developed chest pain and retrosternal discomfort, prompting him to seek initial treatment at a nearby hospital before being referred to another facility. Upon examination, he reported additional symptoms including bilateral lower limb pain, multiple joint pain, and back discomfort.

### Primary Survey

#### *Airway*

The airway is clear with no signs of C-spine tenderness.

#### *Breathing*

The patient presents with 28 respiration rates/minute and at room temperature his $SpO_2$ were 82%. Bilateral chest auscultation reveals equal air entry. Non-invasive ventilation has been initiated.

#### *Circulation*

PR 82 beats/minute, BP were 110/70 mm Hg, and intravenous access has been established.

#### *Neurological Status*

Glasgow coma scale (GLS) score is E4v5m6, indicating intact neurological function with movement in all four limbs.

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*Secondary Survey (Head to Toe Examination)*

No facial tenderness or abnormal jugular venous pressure. Increased respiratory effort and use of accessory muscles noted in the chest, with tenderness over the costo-chondral junction.

*Abdomen*

Soft with no tenderness, and a scar from a previous splenectomy is observed.

*Back*

Tenderness is present in the dorsal spine, with no neurological deficits.

*Upper Extremity*

Tenderness noted in one of the interphalangeal joints of the hand.

*Lower Extremity*

Tenderness observed in the knee and metatarsophalangeal joint, with no signs of gangrene.

*Medical History*

Documented history of SCD.

## Diagnostic Assessment

- Patient's CBC revealed an increased mean corpuscular volume (MCV), NT-ProBNP levels were <1000 (725), indicating a cardiac biomarker within normal range, while hyponatraemia was noted with a serum sodium level of 123 mmol/L.
- Chest X-ray exhibited perihilar opacity without effusion, and bedside ultrasound confirmed mild pleural effusion (Figs 1 and 2).
- The inferior vena cava (IVC) demonstrated no phasic variation.
- Peripheral blood smear occasionally showed sickle cells, and haemoglobin electrophoresis revealed an elevated fetal haemoglobin (HbF) level of 4%.
- Genetic diagnostic test like HLA typing was done for patient and his sister, and it showed perfect 10/10 allele match. To access the function of heart, ECG was done, and it showed normal left ventricular ejection fraction and no evidence of pulmonary hypertension (Table 1).

Consequently, the patient was admitted under the care of the pulmonology department and as per the protocols issued by National Heart, Lung, and Blood Institute (NHLBI), all patients diagnosed with ACS must be hospitalised, and oxygen levels closely monitored.[6]

Early and aggressive intervention is particularly beneficial, especially when patients exhibit risk factors such as involvement of multiple lung lobes, presence of concurrent respiratory ailments, or limited access to blood transfusions. A schematic representation of affected individual with paediatric ACS combined with SCD, due to severity of his condition, recipient was admitted to ICU.

Therapeutic interventions included intravenous Meropenem, intravenous Doxycycline 100 mg, Tablet Myelostat 500 mg, and Tablet Pentids 400 mg. Peripheral smear analysis demonstrated occasional sickle cells. Progressive hypoxemia necessitated urgent intubation and initiation of mechanical ventilation.

Following multidisciplinary consultation with haematologists and pathologists, Hb electrophoresis was performed, confirming

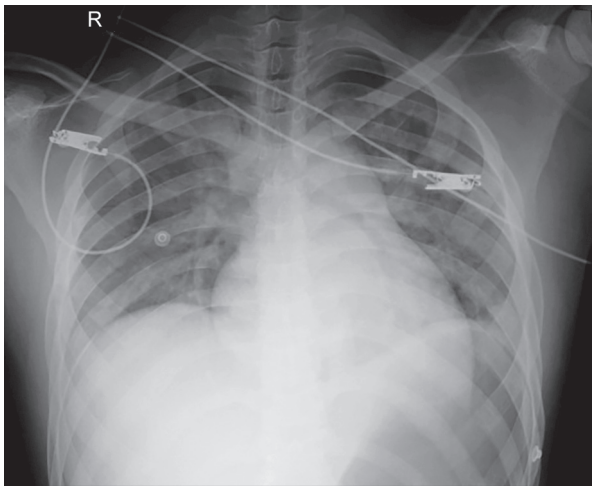


**Fig. 1:** Thorax radiograph on the day of admission

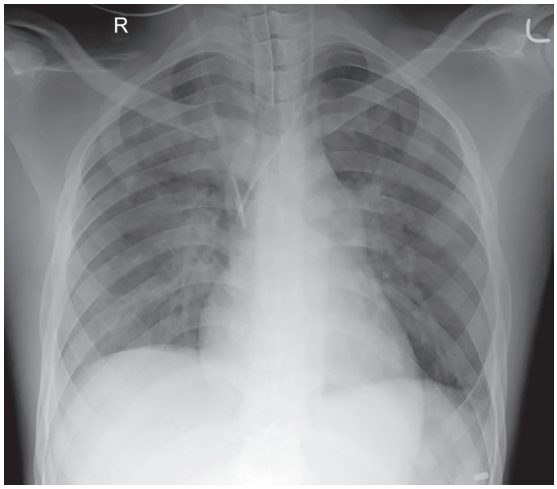


**Fig. 2:** Thorax radiograph after intubation and placement of right and left central venous catheter

**Table 1:** Diagnosis of HLA typing of the patient and his sister

| *LOCUS* | *HLA-A** | *HLA-B** | *HLA-C** | *HLA-DRB1* | *HLA-DQB1** |
|---|---|---|---|---|---|
| *Patient (19 years)* | | | | | |
| HLA-CLASS I and II | A*11:01:01G | B*35:01:01G | C*04:01:01G | DRB1*01:01G | DQB1*05:01:01G |
| | A*68:01:02:01 | B*40:06:01 | C*07:02:01G | DRBI*15:01:01G | DQBI*06:01:01G |
| *Patient's sister (21 years)* | | | | | |
| HLA-CLASS I and II | A*11:01:01G | B*35:01:01G | C*04:01:01G | DRB1*01:01G | DQB1*05:01:01G |
| | A*68:01:02:01 | B*40:06:01 | C*07:02:01G | DRBI*15:01:01G | DQBI*06:01:01G |

the diagnosis of sickle cell anaemia. An exchange transfusion comprising 8 units of packed red blood cells was implemented.

Despite initial interventions, persistent fever prompted further investigation, including endotracheal aspirate analysis revealing yellowish discolouration suggestive of possible infection. Concurrently, flu panel and COVID RT-PCR tests were conducted alongside blood and endotracheal aspirate cultures (Fig. 3).

According to Shilpa Jain and colleagues in 2017, they mentioned that pulmonary infarction is a noteworthy factor in the onset of ACS, accounting for 16% of ACS cases in the National Acute Chest Syndrome Study (NACSS).

This occurrence occurs because RBCs adhere more strongly to the squamous epithelium of a blood vessel. Thus leading to blockages in blood vessels, which in turn causes a disparity in ventilation-perfusion and exacerbates hypoxemia.[5]

In consultation with infectious disease specialists, antibiotic therapy was escalated to address suspected gram-negative infection. The patient was transitioned from the ICU to the general ward following stabilization of vital signs and gradual weaning off oxygen support. Subsequent culture and sensitivity testing of endotracheal aspirate (Fig. 4) and blood samples confirmed the presence of *Acinetobacter baumannii*. Upon infectious disease recommendations, the patient was referred to a haematologist-oncologist for further management.

The patient responded favourably to the instituted treatment regimen, with a resolution of fever and stabilization of vital signs. Upon discharge, he was prescribed a regimen consisting of T. Myelostat 500 mg, T. Folvite 5 mg, and Cap. Calmitone, and T. Glevo 500 mg.

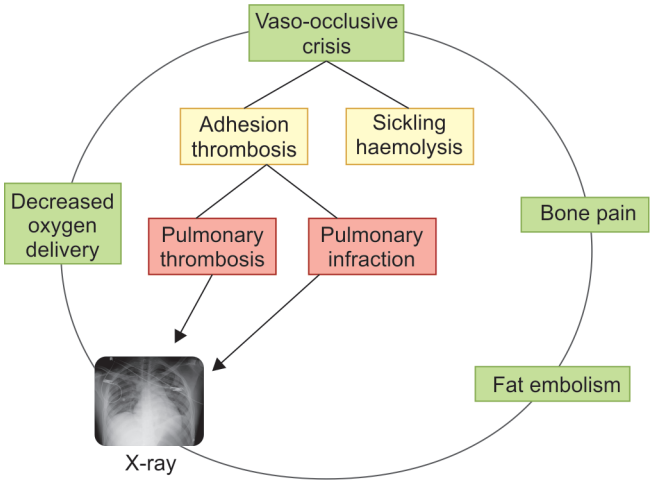


**Fig. 3:** Schematic representation showing the prognosis of ACS patient with sickle cell RBC showing vascular obstruction, lung infection

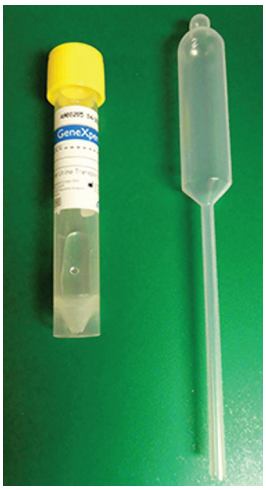

**Fig. 4:** Endotracheal aspirate collection

## Discussion

This case study illustrates the multifaceted nature of ACS in SCD patients, emphasising the importance of prompt recognition and comprehensive management. For the paediatric as well as adult populations of concern, ACS emphasises a significant causative factor for death, especially individuals having SCD. The patient presented with typical symptoms of ACS, including retrosternal chest pain and back discomfort, necessitating hospitalization and aggressive intervention.

Prompt admission to the hospital and vigilant assessment of oxygen levels and overall health are crucial in handling ACS. Early identification of risk elements, such as involvement of multiple lung lobes and the presence of concurrent respiratory ailments, allows medical professionals to promptly initiate suitable treatment approaches. Diagnostic assessments, encompassing laboratory analyses and imaging examinations, assist in confirming the diagnosis and directing treatment plans.

In this case, a multidisciplinary collaboration among pulmonologists, haematologists, infectious disease specialists, and critical care teams facilitated optimal patient care. Empiric antibiotic therapy, mechanical ventilation, and exchange transfusion were implemented based on clinical findings and consultation with relevant specialists.

## Conclusion

In conclusion, this case highlights the significance of a coordinated and multidisciplinary approach in managing ACS in patients with SCD. Adherence to established guidelines and protocols, alongside early recognition of risk factors and prompt initiation of treatment, are essential for improving patient outcomes and reducing morbidity and mortality associated with ACS. Continued research efforts and enhanced clinical protocols are warranted to further optimise the care of individuals with SCD and ACS.

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