

## Feasibility of Early Diagnosis and Treatment of Acute Chest Syndrome in Sickle Cell Anaemia- A Case Report

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### ABSTRACT

Acute chest syndrome is a serious complication and one of the causes of mortality in sickle cell disease. Twenty eight year old male was admitted in our hospital with fever, severe chest pain and haemolytic crisis. He was treated with intravenous antibiotics, fluids, parenteral analgesics and blood transfusion. Severe hypoxemia developed after 72 hours of hospitalization. The patient was transferred to the intensive care unit of our hospital. Oxygen therapy and inotropic support were initiated. Vital parameters and organ functions returned to normal after treatment.

**KEYWORDS:** Acute chest syndrome, Sickle cell anaemia

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### CASE REPORT

A 28 year old customer service officer presented at our peripheral hospital, with a 2 day history of fever, headache and passage of dark coloured urine. He also vomited occasionally. He is known to have sickle cell disease.

Physical examination revealed a young man with a temperature of 38.3 ° C, pale and icteric. Respiratory examination showed a respiratory rate of 28 cycles per minute with vesicular breath sounds on auscultation. Abdominal examination revealed epigastric tenderness. Other systems were normal.

An initial diagnosis of Malaria in Sickle cell disease was made and hemogram showed a packed cell volume of 18%, a white cell count of 16, 200/mm<sup>3</sup> with normal differential. He was admitted and commenced on intravenous fluid, anti-malarials, analgesics, antibiotics and antacids.

By the 2<sup>nd</sup> day he complained of severe back and chest pain with high grade fever. His chest was still clear but repeat blood test showed a further fall in packed cell volume (PCV 16%) with a rise in the white cell count (19, 000/mm<sup>3</sup>). He was then transferred to our main hospital for further management.

On arrival at the hospital, physical examination was essentially as before. Oxygen saturation at room air was 98%. Epigastric tenderness was present on abdominal examination.

A diagnosis of Sepsis and Acute Chest Syndrome in sickle cell disease patient was made. Management was commenced with transfusion of 2 pints of blood, intravenous antibiotics, parenteral analgesics and fluids. A full sepsis work up including blood culture, chest x-ray and chest CT scan was ordered. By the 2<sup>nd</sup> day on admission physical examination revealed crepitations on the lower lung zones and renal angle tenderness.



Fig 1: Chest x-ray showing a homogeneous opacity in the left lower lung zone with pulmonary congestion.

Chest X-ray (Fig 1) showed a mild left sided pleural effusion that was judged not to require drainage by our Cardiothoracic surgeon. By the 3<sup>rd</sup> day his oxygen saturation at room air dropped to 71%, his pulse was 103 beats per minute and his blood pressure 130/90 mmHg. An urgent chest computerized tomography was done which showed features of acute respiratory distress syndrome (Fig 2).

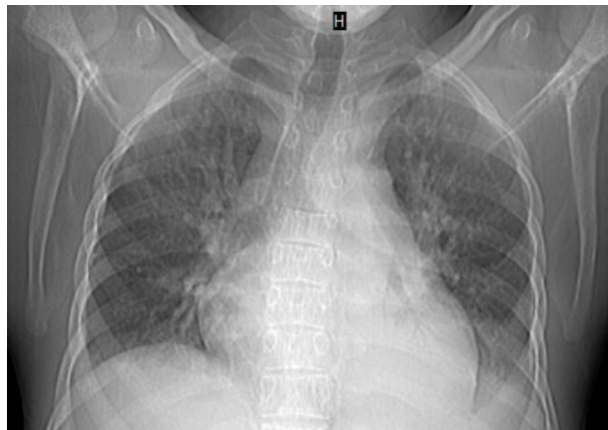


Fig 2: Chest CT scan showing bilateral lower lobe consolidation with bilateral pleural effusion.

He was then transferred to the intensive care unit for closer monitoring with multi-parameter cardiac monitor, oxygen therapy via non-rebreathable face mask and inotropic support with upward titration to improve tissue perfusion. His blood pressure was 130/80 mmHg and pulse 68 beats per minute on admission into the intensive care unit.

Arterial blood gas values while he breathed 10 L/min were pH= 7.45; PaCO<sub>2</sub> = 39.5 mmHg; PaO<sub>2</sub> = 147.4 mmHg; Oxygen saturation = 82%. Antibiotics were tailored according to urine and blood culture results. Pain control was maintained with both oral and parenteral analgesics. Chest physiotherapy was commenced 4 days later.

The patient eventually improved significantly and was weaned off oxygen after 6 days and was discharged home in a clinically stable state 2 days later.

## DISCUSSION

Acute chest syndrome(ACS) is defined as an acute episode associated with clinical and radiologic evidence of new pulmonary abnormalities in patients with SCD and often accompanied by fever, bone pain, chest pain, cough, dyspnea, hypoxia, leukocytosis and decline in hemoglobin below the usual steady-state level.<sup>1-4</sup> Our patient, known to be having sickle cell disease, presented in relatively stable clinical state but with a rapid clinical deterioration and features of acute chest syndrome, underscoring the importance of close monitoring and early institution of aggressive management in sickle cell disease patients presenting with acute complications especially when respiratory compromise is suspected.

In addition to the aforementioned features in ACS, there are new pulmonary infiltrates (involving at least one complete lung segment-not atelectasis), Fever is usually high grade (Temperature more than 38.5°C) with tachypnoea, wheezing or cough<sup>4</sup>. ACS is the commonest cause of mortality in sickle cell disease and second only to vaso-occlusive crisis as the most frequent complication. ACS is known to frequently present insidiously and non specifically, often complicating other conditions like fat embolism, infection, hypoventilation from any cause and infarction. It may occur in patient hospitalised for other indications like surgery<sup>5</sup>. Risk factors indentified for ACS include younger age, homozygous haemoglobin SS, low haemoglobin F levels, High steady state leukocyte count, previous history of acute chest syndrome and avascular necrosis of bone and a high steady-state hemoglobin level<sup>6, 10</sup>. Both children and adults frequently have a painful event preceding the development of ACS<sup>7</sup>. Deterioration in pulmonary function was preceded by severe back and chest pain in the patient under study. This represents a predictable pathophysiologic course of ACS which essentially is a vaso-occlusive phenomenon affecting the pulmonary vasculature. However, it is recognised that a

patient may have normal findings or only slight abnormalities at presentation, yet be at risk for significant disease<sup>5</sup>. Notably, up to 50% of patients diagnosed with ACS are initially admitted to the hospital for other reasons and subsequently develop the disease. The reason for admission in these cases is most often vaso-occlusive crisis. The average time to development of ACS after hospitalization is 2.5 days<sup>10</sup>.

Polymerisation which stiffens and sickles the HbSS containing erythrocyte making it a hard and unyielding obstacle with consequent hypoxia and tissue damage downstream is known to be key to the sickling disorder. The lung is the depolymerising organ protecting the arterial side of the circulation from cells with polymerized haemoglobin. The lungs become injured in this process of inflammation with adherence of sickle red cells to activated endothelium leading to hypo ventilation. These prevent re oxygenation causing more inflammation and infarction. Local damage to the lungs and poor oxygenation makes the lung prone to severe manifestations of pneumonia<sup>4</sup>. Hypoxaemia and tachypnoea are constant features of acute chest syndrome which expectedly also occurred in our patient. He was noted to have suddenly become tachypnoeic with a precipitous dip in oxygen saturation. Arterial blood gas analysis confirmed severe hypoxaemia.

The aim of treatment of ACS is to reverse the pulmonary pathology, to correct significant hypoxemia, to relieve the chest pain and to prevent recurrent attacks<sup>1</sup>. This includes the well established supportive care like hydration, pain relief and psychosocial support<sup>7</sup>. The goal of hydration is to give 3-4L of fluid over 24 hours. Liberal oral fluid intake is encouraged, supplemented by intravenous fluid. In patients requiring opiate analgesic however, who may not be able to drink, fluid may be almost entirely intravenous. Supplemental oxygen would be useful in most patients since it reduces sickling in the poorly ventilated portions of the lungs. However, in subjects with normal oxygen saturation (SPO<sub>2</sub> >97%) and normal respiratory rates, supplemental oxygen may not offer any benefits<sup>1</sup>. High dose of opiates is usually required at the initial stage. This has a double benefit of relieving patient's painful distress and helping with pain free lung expansion and cough.

Red cell transfusion in selected patients, which has been shown to yield similar result with the more cumbersome exchange blood transfusion, is one of the major interventions in acute chest syndrome. This is especially true in circumstances like the presentation of our patient in which the haemoglobin concentration is low in addition to a serious complication of sickle cell disease. Generally simple blood transfusion is preferred if the haemoglobin concentration is less than 8-9g/dl. Simple transfusion above this haemoglobin level or increasing haemoglobin concentration to more than 11g/dl by

whatever means would result in increased viscosity and consequent complications. Exchange blood transfusion would be indicated if the haemoglobin level is more than 8-9g/dl in the setting of serious complication of sickle cell disease or rapid deterioration in clinical condition<sup>4,7,8</sup>. Ideally, blood units should be matched also for C, E and Kell antigens. We do not do this routinely. Blood transfusion increases the oxygen carrying capacity of the blood by increased haemoglobin AA and decreasing the level of the sickle cells.

Use of hydroxyurea (an antimetabolite/cytotoxic drug) on the long term has been established as a major preventive measure in reducing the incidence of acute chest syndrome. Benefit derived from hydroxyurea is related to its ability to increase fetal haemoglobin, reduction of baseline white blood cell levels, increased deformability of red blood cell, altered adhesive receptors on sickle reticulocytes and production of nitric oxide from the metabolism of hydroxyurea (hydroxycarbamide)<sup>8,9</sup>. Cultures carried out in our patient were negative. This is not surprising since previous workers have shown that despite documentation of infections in subjects with acute chest syndrome, bacteraemia was demonstrable in negligible fraction of the subjects<sup>6,10</sup>. It has been postulated that viral infection, Chlamydia and mycoplasma are the major pathogens causing lower respiratory tract infection. Hence, macrolide and Quinolone antibiotics are considered routine in subjects with acute chest syndrome<sup>4</sup>.

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