Acute chest syndrome in children with sickle cell disease: Saudi Arabian experience

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Vol. 9, No. 1 (2005-10 - 2005-12)

Curr Pediatr Res 2005; 9 (1 & 2): 23-26

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Key words: Sickle cell disease, children, acute chest syndrome

Accepted June 14, 2005

Abstract

Acute chest syndrome (ACS) is a known complication of sickle cell disease (SCD). It carries high morbidity and mortality. We conducted a retrospective study to evaluate the frequency, clinical, laboratory, and radiological features of this complication among children with sickle cell disease in the Southern Province of Saudi Arabia. Our results were generally comparable to international published data among similar population. Our results revealed that the frequency of ACS episodes are age dependent, which occurred more frequently in young children i.e. < 4 years of age (57%). Fever and cough were the most frequent symptoms; 93% and 86% respectively. Most of the cases experienced respiratory distress such as tachypnea (86%), chest retraction (64%), and decreased breath sounds (57%). On the other hand only small number of patients (11%) had complete normal chest examination. ACS, as other sickle cell crises, usually develops in association with other complication. In our study, painful crisis was the most commonly associated complication along with ACS (79%). All of the chest X-rays were positive at different anatomical sites; bilateral involvement was observed frequently (36%). This study is unique being the first description of such problem in children in the Southern Province of Saudi Arabia. It will serve as a base s for subsequent studies.

Introduction

Acute chest syndrome is an acute pulmonary illness in patients with sickle cell disease. It is defined as an acute episode associated with clinical and radiological evidence of new pulmonary abnormalities in patients with sickle cell disease (SCD) and often accompanied by fever, bone pain, pleuritic chest pain, cough, dyspnea, hypoxia, leukocytosis and decline in hemoglobin below the usual steady-state level [1-3]. It is a common problem, causing significant morbidity and mortality. Many factors may cause this syndrome. It is responsible for up to 25% of deaths, and for more than 90% of hospital admission [4-7].

The term “acute chest syndrome” was first suggested by Charache et al in 1979 [8], acknowledging the difficulties in determining its pathogenesis. Treatment is primarily supportive. Therapy includes hydration, analgesia, supplemental oxygen, antibiotics, blood transfusion and mechanical ventilation. Early detection and aggressive management may limit its severity and prevent its complications. It is estimated that ACS will occur in nearly one third of patients at risk [9-11]

Materials and Methods

This is a retrospective study performed at Kamiss Military Hospital and Aseer Central Hospital, Southern province, Saudi Arabia. Medical records of children with SCD who were diagnosed with acute chest syndrome between 1995-2001 were carefully reviewed. The following information were extracted: Sex, age, presenting symptoms, and signs, laboratory values, radiological findings as well as associated complications. Patients were divided into three groups on the basis of age (< 4 years, 4-8 years, and 9-12 years) with a view to assessing clinical presentation. The incidence of symptoms, signs, laboratory and radiological findings were studied. Treatment including Blood transfusion, exchange, antibiotics, course of the patients, intensive care admission, intubations and duration of hospital stay were noted.

Data was entered into IBM compatible computer at the Department of Family and Community Medicine, College of Medicine, King Khalid University. The Statistical Package for Social Science software (SPSSversion 10) was used for analysis of the data.

Results

Patients: Most of the episodes occurred in young children < 4 years of age (57%) and least common in older children > 9 years (11%). ACS noted to occur in both sexes with slight male predilection i.e. 54% compared to 46% in females. Most of the patients in the study had single episode of ACS(71%), while repeated episodes (second or more) have occurred in 28% (Table 1).

Table 1: Patient Characteristics

Age Group	No. of Patient	%
< 4 year	16	57
4 – 8 year	9	32
9 – 12 year	3	11
Male	15	54
Female	13	46
Single episode	20	71
Repeated episodes	8	28

Table 3: Presenting Signs

Signs	Number	%
Temperature	22	79
>39° C	8	21
< 39° C	24	86
Tachypnea	18	64
Retraction	26	93
Tachycardia	15	54

Dullness to percussion	16	57
Decreased breath sound	20	71
Wheezing	7	29
Bronchial breathing	9	32
Normal exam	3	11

Table 4: Baseline and ACS laboratory values

Test	Mean	CI
Hb (g/dl)	7.6	6.41-8.90
WBC (X 10^9/L)	18.9	16.70-20.81
Platelet (X 10^9/L)	345.7	321.91-393.50
Reticulocytes (%)	9.8	7.40-10.60
Serum bilirubin	3.8	3.10-4.21

Table 5: Associated problems with ACS

Problems	Number	%
Painful Crisis	22	79
Infection		
Bacteria	15	50
URTS	10	33
UTI	2	7
Acute Cholecystitis	2	7
Postoperative	4	14
Sequestration crisis	2	7
Bronchial asthma	4	14

Table 6: Radiological Manifestations

Chest radiograph	Number	%
Upper zone involvement	2	7

Middle zone involvement	8	27
Lower zone involvement	4	14
More than one zone in one side	6	20
Bilateral involvement	10	33
Pleural effusion	7	31

Presenting symptoms: The most common presenting symptoms were fever, cough, and chest pain. Some patients experienced shortness of breath, wheezing, chills and productive cough. Fever was the most common presentation, (93%), cough and chest pain were found in 86% and 71% respectively while dyspnea was recognized in 61%. The frequency of presenting symptoms was agedependent with fever and cough being more common in young children (age 2-4 years) and the incidence of chest pain, shortness of breath, productive cough occurred in less than quarter of patients. (Table 2).

Table 7: Presenting symptoms

	Number	%
Fever	26	93
Cough	24	86
Chest pain	20	71
Productive cough	16	57
Shortness of breath	17	61

Physical findings: Vital signs at the time of hospitalization were age dependent with young children experiencing higher temperature, pulse rate, and respiratory rate than older children. The most frequent physical exam findings were an abnormal chest examination in the form of signs of respiratory distress, decreased air entry, rales and dullness to percussion, whereas normal chest examination was observed in only (11%) of the cases (Table 3). Painful vasoocclusive crisis was associated with (79%) of patients. It has been considered the most common associated event with ACS followed by an underlying infectious process (55%).

Laboratory findings: Blood count documented during acute chest syndrome was compared with steady state value. Hemoglobin and white blood counts showed significant changes with the severity of the disease. (Table 4)

Radiographic findings

Radiographic findings vary by age. The predominant radiological findings were bilateral lungs involvement (36%). Young children had isolated upper and middle lobe disease significantly more often and lower disease less often than older children (Table 6).

Discussion

Episodes of acute chest pain in patients with SCD associated with a new infiltrate on chest film are called “acute chest syndrome” (ACS). These episodes are second only to acute painful episodes in term of incidence and need for hospitalization. These episodes are more common in children than adults and more common in patients with low levels of fetal hemoglobin and high level of total hemoglobin. [12].

Acute chest syndrome consists of combination of signs and symptoms. It is a form of lung injury that can lead to adult respiratory distress syndrome (ARDS). Pulmonary disease is the leading cause of death in sickle cell disease [13]. There are both acute and chronic pulmonary manifestations of sickle cell disease. The acute syndrome consists of dyspnea, chest pain, fever, tachypnea, pulmonary infiltrate on radiography and leukocytosis. It affects approximately 30% of patients with sickle cell disease and may be lifethreatening. [14]. There is scanty data about the acute chest syndrome among Saudi children with SCD. Al-Dabbous et al reported the frequency of ACS in Qatif (a medium size city in the Eastern Province of the Saudi Arabia) in the order of 5- 7.7% [11, 15]. It affects both boys and girls, with predominance of males [11,16]. The process is usually due to infection or vasoocclusion but may also be the result of non cardiogenic pulmonary embolization from a distant thrombus or infracted bone marrow. [14,17]. Approximately 50% of patients experience at least one episode of ACS and they ultimately have a higher chance of dying at an early age, and the mortality rate after such an event can be as high as 10-12% [18-20]. It is important to recognize and treat these events aggressively since it is the leading cause of death among sickle cell patients [21].

This retrospective study of 28 patients with sickle cell disease who developed ACS is a representative sample of such patients. Our data revealed that the incidence of ACS was strongly influenced by patient age, being most common in younger children and least frequent in older children. The association of ACS with young children could also be explained by reasoning that the increased susceptibility to viral respiratory infection in young children could precipitate ACS in children who are also more likely to have significant abnormal respiratory finding on chest exam at presentation [22].

Other factors associated with a high ACS incidence were a higher steadystate leukocyte counts. The reason for the association between high leukocyte counts and ACS incidence is not clear. It could be explained by the increasing susceptibility to viral respiratory infection in young children as a precipitating factor for ACS [23].

The predominant radiological findings in our study revealed diffuse lung involvement, unlike lower lobes being involved in other studies [24,25]. This difference in the site of the lung involvement could be attributed the fact that most of the cases in this study are very young children and the possibility of an associated infectious process rendering both lung to be affected.

This is the first study in Aseer Central Hospital to record the experience of ACS in children with SCD, and to the best of our knowledge, it has not been reported from the Southern Province of the Kingdom of Saudi Arabia. This retrospective study demonstrates the clinical presentation of ACS in children with SCD in this part of Saudi Arabia. It is of great value as baseline study. Nevertheless, further studies of such condition are required to clearly understand this important complication of SCD.

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