Acute chest syndrome in adult sickle cell disease in eastern Saudi Arabia

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Pulmonary complications with sickle cell disease are serious and carry significant morbidity and mortality. Acute chest syndrome is a spectrum of pulmonary pathology, which can include common chest pain, fever, cough, and dyspnea with abnormal clinical and radiological chest signs.[1-3] The aim of this study was to report the clinical features and outcome of acute chest syndrome episodes in adult sickle cell disease in patients admitted to King Fahad Hospital, Hofuf, in eastern Saudi Arabia.

Methods

In this retrospective study, we reviewed the records of 135 adult patients with sickle cell disease (age 12 years and above) with acute chest syndrome (ACS). During the study period, total admissions to the medical ward at King Fahad Hospital were 46 878, of which 7345 (15.7%) were for sickle cell disease. Of the latter, 135 patients (1.8%) had 190 admissions with a diagnosis of acute chest syndrome, including development of a new lung infiltration on chest x-ray during hospitalization between 1998 and 2003. There were 79 males and 56 females (1.3:1 ratio). Ages ranged from 12 years to 50 years, with a mean age of 20.4±8 years. We recorded age, sex, clinical presentation, chest x- ray results, CBC, cultures, and blood gases, and if available, hemoglobin electrophoresis course during hospitalization, blood transfusion and blood exchange, blood group, ICU admission, and bronchoscopy results, it done duration of admission, and outcome.

Results

ACS developed on admission in 79% of the 135 patients and during hospitalization in 21%. One hundred and three patients (76%) had a single episode of ACS and 32 (24%) patients had recurrent episodes of ACS. One hundred and eighteen patients (87%) had sickle cell anemia, 13 patients (10%) HB Sb0 sickle cell syndrome, and 4 patients (3%) had HB Sb+ sickle cell syndrome. Fever, cough, and chest pain were the most common symptoms. Raised temperature and tachypnea were the most common findings. ACS was associated with painful crisis of varying intensity in more than 90% of the patients. There was a significant increase in leukocytes and a significant decrease in hemoglobin. Radiological studies revealed unilateral infiltration in 75%, bilateral infiltration in 25%, and pleural effusion in 22% of patients.

Bacteremia was found only in 4.2%. Simple blood transfusion was required in 44% and exchange transfusion in 5%. Nineteen percent needed admission to ICU and 12% required a mechanical ventilator. Thirteen patients died during an attack of ACS (9.6%). The mean duration of admission was 8.5±5 days.

Discussion

Sickle cell disease and its complications are a major heath problem in the Al-Hassa area,[1,2] accounting for more than 15% of total admissions to the medical ward. Although the frequency of ACS is in decline in adults, it is associated with significant morbidity and mortality despite our knowledge about SCD and improvements in health services.[3,4] In general, our patients had clinical features and hematological changes (leukocytosis, decreases in hemoglobin, and increases in platelets) similar to that re-

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Table 1. Distribution of patients with sickle cell disease/acute chest sydrome by age and sex.

Age group (years)	No. males	No. females	No. male deaths	No. female deaths
12-19	23	30	2	1
20-29	41	22	5	2
30-39	10	3	0	1
40-49	4	0	1	0
> 50	1	1	1	0
Total	79	56	9	4

No statistically significant differences between males and females for numbers or deaths ($P \geq 0.05$).

Table 2. Frequncy of episodes of acute chest syndrome (ACS).

No. of episodes	Male	Female	Total ACS	No. of deaths
1X	58	45	103	9
2X	10	7	34	3
3X	4	4	24	1
4X	5	1	24	0
5X	0	1	5	0
total	79	56	190	13

No statistically significant differences between the frequency of ACS and death (χ^2=0.39 and $P \geq 0.05$).

ported in other studies, accept for rhonchi which was not observed in our patients.[5-7] It is not known specifically why the incidence of ACS is in decline after adolescence and why there is a predilection in males. Radiologically, unilateral lower lobe lung infiltrations (75%) predominated in our study. Bacterial infection was only documented in 4.2%, which supports the importance of other etiologies that may contribute to the pathogenesis, and account for the clinical picture in ACS.[7,8-11]

Despite the belief that SCD in the Eastern Province of Saudi Arabia is a milder form and associated with a high level of hemoglobin F, that does not prevent the occurrence of SCD-related complications. During the period of this study, 13 of 135 (9.6%) patients died, which may be the highest reported.[5,6] This reflects less the nature of the disease than the lack of close observation, and

Table 3. Hematological values during acute chest syndrome compared to baseline.

Test	Baseline (mean±SD)	ACS (mean±SD)
WBC (x10^9/L)	11.3±4.8	18.6±7*
Hb (gm\dL)	9.3±1.6	7.1±1.5
Platelets (x10^{12}/L)	0.364±0.153	0.366±0.175

*P<0.001 vs. baseline

Table 4. Site of lung infiltration.

Site	No. episodes	%	Effusion	%	Deaths
Right	84	44	16	19	3
Left	58	31	17	29	2
Bilateral	48	2	9	19	8
Total	190	100	42	22	13

*Death with bilateral lung infiltration was significantly higher than unilateral lung infiltration (χ^2= 9.7 and P value<0.01); no significant difference was noted between right and left (χ^2=.0005 $P \leq 0.05$)

Table 5. Mortality during acute chest syndrome in relation to hematological values.

Hematological parameter	Number of patients	Deaths
White blood cell count (x10^9/L)		
<12	46	2
12-20	72	4
>20	72	7
Hb (g/dL)		
< 8	47	4
> 8	143	9
Platelets*(x10^{12}/L)		
<0.15	32	7
0.15-0.199	12	0
≥0.20	146	6

*P<0.001 for mortality in comparison to platelet counts

suboptimal therapeutic intervention. Bilateral lung infiltrations and thrombocytopenia associated with a severe course of ACS in adult sickle cell disease patients increases the relative risk of mortality.

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