
Brief Reports

Acute Chest Syndrome in Sickle Cell Disease

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Acute chest syndrome (ACS) is a term used to describe fever, clinical findings of pulmonic process and roentgenographic evidence of a new pulmonic infiltrate in a patient with sickle cell hemoglobinopathy (1). ACS is considered to be an alarming symptom necessitating hospitalization. As prevalence of sickle cell disease (SCD) is high (6.42%) in hospitalized children of Western Orissa (2) and very often children are admitted in different crises(3), this study was conducted to evaluate the frequency, clinical presentations and outcomes of ACS in SCD in this hospitalized population.

Subjects and Methods

All patients admitted with SCD to the Pediatric ward of V.S.S. Medical College Hospital, Burla from January, 1993 to December, 1995 were examined for presence of ACS. Sickle cell diagnosis was made on the basis of standard electrophoresis. Baseline hematological tests were conducted and X-ray chest and blood cultures were taken in all cases. Those included in the study had at least two of the

following: Temperature $\geq 38^{\circ}\text{C}$, physical findings consistent with pulmonic inflammation and chest radiograph findings consistent with an acute pulmonic process(4). The patients were managed with maintenance of proper hydration, analgesics and antibiotics, wherever indicated. Blood transfusions were used for severe respiratory distress with extensive pulmonic involvement or in severe anemia with marked fall in hemoglobin concentration.

Results

Out of the total 228 cases of SCD admitted to the Pediatric Ward, 40 children had ACS (13.2%). There were 24 boys and 16 girls. The age ranged from 6 months to 12 years (mean 7.2 ± 2.4 yr). All the patients were anemic, 25 (62.5%) were febrile, 15 (37.5%) had chest pain and 23 (57.5%) had poor air entry with diffuse rales in lung fields. Radiologically 24 (60%) had right sided, 13 (32%) left sided and 3 (7.5%) bilateral pulmonic infiltration. On the right side, 15 (37.5%) had non-homogeneous opacities, 7 (17.5%) collapse with consolidation and 2 (5%) minimal pleural effusion. Similar figures for left side were 8 (20%), 3 (7.5%) and 2 (5%) cases, respectively. In 3 (7.5%) cases, both lungs were involved with collapse and consolidation.

Laboratory examination revealed mean hemoglobin (g/dl), TLC (thousand per cu mm) and ESR (mm fall in first hour) to be 5.6 ± 1.24 , 12.8 ± 3.8 and 27 ± 8.5 , respectively. Positive bacterial growths were obtained in 4 (10%) blood cultures only, of which 2 had *Streptococcus pneumoniae* and one each had *Staphylococcus coagulase positive* and *H. influenzae*.

The patients were managed with fluid,

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Manuscript received: October 17, 1996;

Initial review completed: December 31, 1996;

Revision accepted: February 27, 1997

analgesics and antibiotics wherever needed. Thirty patients (75%) required packed cell transfusions and all were cured and discharged.

Discussion

ACS is a frequent cause of morbidity and mortality in SCD (4,5). Previously ACS was believed to be due mostly to bacterial involvement of lungs (1,4). However, recent reports suggest that pulmonary infarction due to local sickling and emboli formation play a major role (6,7).

In the present study, blood culture yielded bacteria in only 4 (10%) cases. The low isolation rate may be due to widespread indiscriminate use of antimicrobials even for minor illnesses or penicillin prophylaxis in some cases of SCD. Further, no studies were done for virus or mycoplasma identification which might also cause ACS. Fifteen (37.5%) patients had pain chest which may favor pulmonary infarcts, either due to vaso-occlusive process or multiple emboli, though pulmonary infection due to pleural invasion cannot be dogmatically ruled out. Similarly poor air entry with diffuse rales observed in 23 (57.5%) cases might suggest either pulmonary infection or infarction. Radiological evidence of pulmonary invasion in form of non-homogeneous opacities and collapse with consolidation were seen in 90% cases whereas minor pleural effusions were documented in 10% cases. Similar pulmonic involvement may either occur in bacterial pneumonia or pulmonic infarcts. A single lobe infiltrate was previously considered to indicate bacterial pneumonia (4); however no such differences were detected in other studies (5). Pyrexia, leucocytosis and raised ESR were observed in 25 (62.5%), 20(50%) and 21(52.5%) cases, respectively. These factors are not very sensitive indicators of infection as patients with vaso-occlusive crisis

may also have high temperature, leucocytosis and raised ESR(7,8).

Thirty patients received packed cell transfusions. Blood transfusion hastened clinical recovery and shortened duration of hospitalization. It has been well documented that with extensive pulmonic involvement, blood transfusion, more particularly exchange transfusion, hastens clinical improvement by lowering fetal hemoglobin and thereby improving oxygenation (6,7,9).

This study indicates that no single factor can be assigned for causation of acute chest syndrome. ACS in SCD is probably of multifactorial origin where both pulmonic infection and infarction play vital roles. So while managing such cases, in addition to appropriate antibiotics and proper hydration, blood transfusion is to be considered with extensive pulmonic involvement.

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