Sicklemia Complicating Chest Surgery

A Case Report

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The problems confronting the patient with sickle cell anemia have become well known. Only recently, however, has it been accepted that sickle cell trait per se (sicklemia) is capable of producing clinical symptoms and pathological findings.7, 10, 12 The development of conditions favorable to sickling, such as factors producing anoxemia and stasis, may precipitate symptoms in the otherwise asymptomatic patient. Sicklemia may pass unrecognized in the routine preoperative evaluation of the patient, because only one in 15 cases show any anemia.12

The purpose of this paper is to point up this problem as it applies to the preoperative evaluation for chest surgery. The following is a report of sickle cell crisis during surgery in a patient with sicklemia in whom additional features of an unusual nature were encountered.

A 17 year old Negro was admitted with a chief complaint of hemoptysis. His subjective symptoms had started six months prior to admission with the onset of recurrent cough productive of mucoid sputum. One week prior to admission he experienced hemoptysis of bright red blood, resulting in hospitalization.

Systemic review was essentially negative except for intermittent left chest wheeze for the past six to seven years. There was no history of dyspnea or chest pain.

Past history revealed pertussis at the age of two years; frequent upper respiratory infections; and pneumonia at the ages of six, nine, and 11 years. Each occurrence of pneumonia required hospitalization, the longest of which was two and a half months at the age of nine years. No previous history of sickle cell crisis was elicited.

Physical examination revealed an asthenic 17 year old Negro who complained of nonproductive cough. His blood pressure both arms was 108/72, pulse 84 and regular, respirations 16 and weight 148 pounds. Positive physical findings consisted of prolonged expiratory phase of the left hemithorax with resulting left chest lag; hyperresonant left hemithorax to percussion; and crepitant inspiratory post-tussic rales over the left lower lobe area.

Laboratory data consisted of 5900 white blood cells with 51 per cent polys, 47 per cent lymphocytes, one monocyte, and one eosinophil and 5.3 million red blood cells. Hemoglobin was 15.0 grams per cent. Erythrocyte sedimentation rate was 2 mm./hr. Hematocrit was 50 volumes per cent. Sickie cell preparation revealed 30 per cent sickling in six hours. Hemoglobin electrophoresis showed 80 per cent A hemoglobin and 20 per cent S hemoglobin, indicating sicklemia. Urinalysis was negative. Blood urea nitrogen was 14.9 mgs. per cent. Fasting blood sugar was 81 mgs. per cent. Serologic test for syphilis was negative. Purified protein derivative, histoplasmin, and coccidiodin first and second strength skin tests were negative.

Electrocardiogram was within normal limits with an intermediate axis. Postero-anterior inspiratory roentgenogram of the chest (Figure 1) demonstrated marked increase in radiiability on the left and unduly prominent lung vascular markings in the right hilar and paracardiac regions.

Postero-anterior expiratory roentgenogram of the chest (Figure 2) revealed the maintenance of the inspiratory volume of the left lung with resulting marked shift of the mediastinum to the right. Fluoroscopy showed the expiratory lag of the left diaphragm to be in phase with the mediastinal shift.

Bronchogram (Figure 3) revealed a normal right pulmonary tree. The left lower

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lobe showed cylindrical and saccular bronchiectatic changes probably resulting from previous infections superimposed upon the mal-developed left lung.

Angiocardiography (Figure 4) showed the hypoplastic left pulmonary artery with small but normally distributed segmental divisions. The right pulmonary vascular

**FIGURE 1**

*Figure 1:* Postero-anterior inspiratory roentgenogram of the chest demonstrates the increased radiability and scarcity of lung vascular markings in the left hemithorax.—

**FIGURE 2**

*Figure 2:* Postero-anterior expiratory roentgenograms of the chest. Note the marked shift of the mediastinum to the right.

**FIGURE 3**

*Figure 3:* Bronchogram in the postero-anterior projection illustrates the normal right bronchial tree with bronchiectatic changes in the widely spaced left lower lobe bronchi.

**FIGURE 4**

*Figure 4:* Angiocardiogram demonstrates the marked hypoplasia of the left pulmonary artery with the usual segmental divisions. The vessels in the right lung are widened suggesting increased blood flow.
tree was enlarged but normally segmented. No definite aberrant, systemic, blood supply to the left lung was demonstrated.

Bronchoscopy revealed diffuse inflammation throughout the left main stem bronchus. Pulmonary function tests showed uneven alveolar mixing and an air velocity index of .84; otherwise all other values were within normal limits.

In order to further differentiate pulmonary function, right and left lung bronchospirometry was done (Table I) revealing a gross ventilatory defect of the left lung. Arterial blood study showed no evidence of oxygen desaturation or carbon dioxide accumulation with exercise.

Thoracotomy was elected to determine if surgical benefit could be obtained. On entry into the left chest a pale left lung with little pigment was found. The left lower lobe was fixed by what appeared to be inflammatory adhesions to the diaphragm and posterolateral chest wall. Pulmonary function studies at the time of operation revealed poor aeration of the left lung which was able to empty only 150 cc. of air by its own compliance. The pulmonary artery was hypoplastic and less than 50 per cent of its normal caliber. An anomalous pulmonary artery, about 6 mm. in diameter, was found in the inferior pulmonary ligament coming from below the diaphragm. Emphysema was diffuse and not localized. He tolerated anesthesia poorly with early appearance of tachypnea, pink facies and precipitous rise of blood pressure to 188/120. Lung biopsy was obtained and thoracotomy judiciously terminated.

Post-operative blood studies showed sickling and drop of hemoglobin to 9.5 grams. By exposing an unsealed, wet film of the blood to an atmosphere of 18 per cent oxygen and 10 per cent carbon dioxide 100 per cent sickling in two minutes was produced postoperatively.

Discussion

In vitro sickling has been shown to depend on the dissociation of oxyhemoglobin in the red blood cell. When hemoglobin is in the reduced state, the cells sickle. When the cells combine with oxygen, they resume their normal discoid shape. An oxygen tension of 45 mm. Hg. has been reported to produce sickling in cells from a patient with sickle cell anemia, while it was necessary to reduce the tension to 18 mm. Hg. in the case of cells from a person with sicklema. A lowering of blood Ph likewise has been found to favor sickling. The danger of airplane flight and lowered oxygen tension to persons with sicklema has previously been reported. During surgery the flushed, pink facies, and precipitous rise in blood pressure are best explained by hypercapnia caused by obstruction to respiration or narcotic depression of it. The patient most likely sickled at this point further hindering pulmonary blood flow and producing a vicious cycle of hypoxemia and sickle cell crisis. This sequence of rapidly occurring events endangered the patient and the operative procedure had to be terminated.

The presence of sicklema adds an additional risk to chest surgery. In the susceptible patient sickle cell tests and hemoglobin electrophoresis should be done in order to better evaluate operative risk.

| TABLE I |
|----------|---------|---------|
| Bronchospirometry | Right Lung | Left Lung |
| Per Cent | Per Cent | Per Cent |
| Min. Ventilation (52-58) | 83 | 17 |
| Min. Oxygen Uptake (52-58) | 83 | 14 |
| Ventilatory Equivalent (1.1 to 1.3) | 1.90 | 2.39 |
| Vital Capacity (52-58) | 68 | 32 |
| Residual Volume (52-58) | 68 | 32 |
The association of anomalous pulmonary arteries with bronchiectasis, pulmonary cysts and intra-lobar sequestration has been well documented\(^1\)\(^\text{-}^4\) and embryology has been described.\(^1\)\(^\text{-}^1\) The role that anomalous pulmonary arteries may play as a cause of dyspnea, recurrent hemorrhage or recurrent infection has been suggested by Mair.\(^13\) This condition is not sufficiently appreciated by the internist, and the diagnosis is rarely made preoperatively. Accidental transection of an anomalous pulmonary artery during surgery has resulted in three deaths from hemorrhage.\(^2\)\(^\text{-}^4\)\(^\text{-}^6\) Angiocardiography\(^5\)\(^\text{-}^8\) has, on occasion, demonstrated anomalous pulmonary arteries and has become of practical importance in preoperative evaluation.

The role that the anomalous pulmonary artery may have played as a cause of hemorrhage in this patient is open to speculation.

REFERENCES