A Case of Acute Chest Syndrome

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Abstract :

Of all the organs in the human body, the lungs are placed in the most vulnerable position. Almost all systems of the human body are carefully packed and isolated, such as central nervous system, cardiovascular system, urinary system etc, but the lungs are exposed to the outside enviroment at all times. It has a direct connection with the heart and conditions like acute left ventricular failure, cor-pulmonale etc; directly affect lungs and vice a versa. The lungs are also subject to many systemic disorders like connective tissue diseases, spreading infections, drug induced diseases, and primary or secondary malignancies. A unique and less frequent case of acute lung injury caused by a hematological disorder is presented.

Keywords : lungs, infection, opacities, sickle cell disease.

Case Report :

A male aged 18 yrs, presented to hospital with chief complains, of fever, cough and breathlesness of 6 days duration. He did not give history of chest pain, wheezing, haemoptysis, orthopnea, paroxysmal nocturnal dyspnea, palpitations, pedal edema, pain in abdomen, vomiting, loss of weight/ appetite, altered sensorium or blood transfusion.He had past history of pain in the long bones and easy fatigability since the age of 7-8 years, because of which he was not able to work for long hours. There was no history of consanguineous marriage of parents,or any disease in any of his siblings.

On general examination the patient was thinly built and poorly nourished. He was 160 cm tall, weighed 46 kg and his body mass index was 17.96kg/sq.m. He was febrile (101°F) and dyspneic. His pulse rate was 106/ min, blood pressure was 130/84 mmHg, respiratory rate was 38/min (abdominothoracic with use of accessory muscles of respiration). He had severe pallor, mild icterus, dehydration, prominent zygomatic bones. There was no pedal edema, clubbing or lymphadenopathy . Jugular venous pressure was not raised.

Examination of the respiratory system revealed bilateral rhonchi over all the chest fields,bilateral crepitations were heard in both mammary and both infra axillary regions. Laboratory investigations: Comlete blood count: Hb:5.9 gm/dl,TLC:48,000/cmm, Differential Count: Neutrophils:64%,Lymphocytes:11%, Monocytes:0%, Eosinophils:2%, Promyelocytes:1%,Metamyelocytes:8%,andMetamyelocytes and band forms: 14%. Platelet Count: 7,20,000/ cumm. The peripheral blood smear examination revealed Microcytic Hypochromic anemia with Sickle cells. Serum biochemistry: Total bilirubin 2.7 mg/dl, with conjugated fraction: 1 mg/dl, SGOT: 31 IU/dl, SGPT: 24 IU/dl, alkaline phosphatase: 148 IU/dl. Sickling Test was positive. HIV and HBsAg : Non Reactive. Arterial blood gas values: pH:7.36 , pCO_2:38, pO_2:90, SpO_2: 92% ,HCO_3:19 [with oxygen] , blood Hb Electrophoresis: Suggestive of sickle - beta° thalassaemia trait because of the following features:

- Fetal Hb : 5.60%
- Hb A_2 : 7.30%
- Hb A : 6.70%
- Hb S :79.70%

Sputum for AFB was negative.ECG showed Sinus Tachycardia. USG Abdomen and Pelvis showed hepatomegaly with a liver span of 18 cm. Chest X-ray Showed bilateral diffuse multiple opacities in mid and lower zones (Fig1) and CT scan (Thorax) showed a cavitatory lesion with debris in the left lower zone (Fig2).

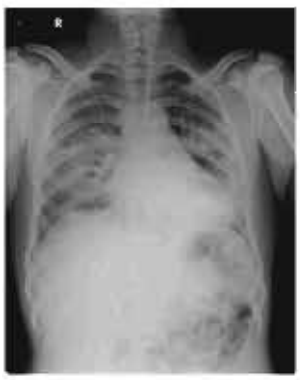

Fig 1: Chest X-ray Showing bilateral opacities in mid and lower zone.

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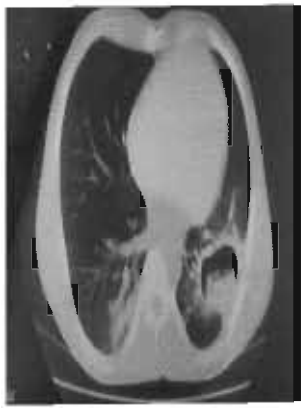

Fig 2: CT scan (Thorax): Showing cavitatory lesion with debris in the left lower zone.

With a positive sickling test, Hb electrophoresis suggestive of beta-thalassemia trait and CT scan (thorax) showing cavitatory lesion with debris in the left lower zone, the diagnosis of acute chest syndrome was made.

Patient was treated with I.V. fluids, blood transfusion (Packed cells), broad spectrum antibiotics and bronchodilators. Patient improved over a period of 7 to 8 days and was discharged after 15 days of hospitalization. Patient was counseled regarding the nature of the disease and was advised follow up but unfortunately did not heed this advice.

Discussion :

Sickle hemoglobin (HbS) occurs when the normal 6th amino acid the glutamic acid residue is replaced by valine in the β chain. The polymerization of deoxygenated HbS is the primary event in the molecular pathogenesis of Sickle cell disease. Sickle cell disease is also characterized by increased adhesion of RBC and other cellular elements to endothelium [1]. Acute chest syndrome is the second most common clinical presentation after painful crises in cases of sickle cell syndrome.

Acute chest syndrome is defined clinically as a new infiltrate on chest radiograph that is accompanied by respiratory symptoms [2]. It is most common in children upto 9 yrs, most prevalent in homozygous SS patients and patients with S/beta^0 thalassemia [2]. Risk factors include young age, winter months, fever, high hemoglobin levels, high steady-state leukocyte count [1, 2].

Pathogenesis: Pulmonary infiltrates may result from either a single process or a combination of several interacting processes, which may include atelectasis, infection, fat embolism, thromboembolism and most commonly, in situ microvascular occlusion within the pulmonary vasculature by sickled erythrocytes. An early rise in the levels of secretory phospholipase A2 (sPLA2) precedes the development of acute chest syndrome and thus may be a useful marker in predicting its occurrence. Furthermore, sPLA2 levels correlate with disease severity. Free heme, released from hemolyzed sickle cells, causes consumption of nitrous oxide as well as generation of free radicals, which further depress nitrous oxide levels leading to vasoconstriction and hypoxia in lungs [2].

Patients can presnt with fever, cough, dyspnea, chest pain, arm and leg pain, abdominal pain, rib or sternal pain [2]. Classical presentation is severe pain in long bones followed several days later by fever, leucocytosis and pulmonary infiltrates.

Treatment consists of oxygen therapy use of bronchodialators, careful fluid management, blood transfusions and broad spectrum antibiotics. Other therapies used in treatment of acute chest syndrome are glucocorticoids, hydroxyurea , stem cell transplantation, nitrous oxide (NO) and varespladib sodium[2,3].

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