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Acute Chest Syndrome: A Rare Complication of COVID 19 in a Patient with Sickle Cell Disease

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Abstract **Case Report**

Acute chest syndrome should be suspected in a sickle cell patient presenting with chest pain, fever and/or hypoxemia. Chest imaging is one of the key tests, especially in making the diagnosis. Assessment of hemolysis, hypoxemia and germ testing is also important to direct management. ***Observation:*** Through our work, we report the case of a 10 years old female patient with an acute chest syndrome following an infection with COVID 19 treated with good clinical evolution. ***Conclusion*:** Acute chest syndrome is primarily due to infections, with COVID 19 being a new etiology to look for during the COVID pandemic.

Keywords: Sickle cell disease, acute chest syndrome, COVID 19, chest CT, broad spectrum antibiotic therapy.

INTRODUCTION

Acute chest syndrome is a serious complication and the leading cause of mortality in sickle cell disease. It results from pulmonary capillary occlusion, followed by complex pathophysiological phenomena. The diagnosis is made in the presence of a bilateral radiological pulmonary infiltrate, accompanied by clinical symptoms such as a febrile state or respiratory symptoms. Treatment includes hydration, oxygen therapy, analgesia, broad-spectrum antibiotic therapy including coverage of atypical germs, and transfusion therapies in case of unfavorable evolution (transfusions or exchange transfusions).

OBJECTIVE

We report the medical observation of a patient with sickle cell disease who presented with an acute chest syndrome secondary to COVID 19 infection.

OBSERVATION

A 10-year-old patient, known to be a sickle cell disease carrier since the age of 2 years, followed in the pediatric hematology department, was admitted to the pediatric department at CHU MED VI MARRAKECH for febrile respiratory distress evolving for 4 days associated with intense chest pain with anosmia.

The clinical examination showed a conscious patient, polypneic at 35 cycles per minute, SaO2 at 90% on room air, febrile at 38°C.

A thoracic CT scan was performed, showing diffuse pulmonary infiltrates suggestive of an acute chest syndrome.

The COVID19 PCR was positive, the haemostasis balance was disturbed with a D-DIMERE level of 10,090, a prothrombin level of 66.3% and a drop in the haemoglobin level to 4.9 g/dl.

The patient was put on hyperhydration, antibiotic therapy with ceftriaxone in association with azithromycin, anticoagulant treatment, and corticotherapy. A blood transfusion was necessary due to the poorly tolerated anemia.

The evolution was marked by the disappearance of the respiratory symptomatology, the negativation of the COVID PCR one week later, and the normalization of the D-dimer level after 3 weeks of treatment.

DISCUSSION

Sickle cell disease is a group of genetic diseases of autosomal recessive transmission. It is characterized by a structural abnormality of hemoglobin

linked to a point mutation in the β-chain gene of globin; the most frequent abnormality is a mutation of codon 6, resulting in a replacement of glutamic acid by a valine (HbS). However, the disease phenotype can vary depending on the mutation inherited on the second allele.

Classically, clinical expression is more severe in patients homozygous for HbS, than in double composite Hb S/C or Hb S/β-thalassemia heterozygosity (or possibly other hemoglobin β-chain defects). In contrast, so-called AS heterozygous subjects are asymptomatic and do not have the complications of the disease.

Acute chest syndrome (ACS) is the second most common reason for hospitalization in adults with sickle cell disease, after vaso-occlusive crisis (VOC), and is associated with high morbidity and mortality. On average, 50% of patients with ATS are admitted for another reason for hospitalization (mainly CVO); however, ATS can also occur immediately without a preliminary CVO.

ATS is the leading cause of mortality in sickle cell patients. This pathology is more frequent in the pediatric population with a frequency that decreases with age, the peak incidence being between the ages of 2 and 4 years. The recurrence rate is around 80% in patients who have already had a first episode.

The pathophysiological mechanisms of ATS are complex and interrelated: alveolar hypoventilation, fatty or cruoric embolism, vaso-occlusion, in situ thrombosis and infection.

Red blood cell deformation occurs through polymerization of deoxyhemoglobin S during hypoxemia. The next step is the occlusion of the microcirculation by the deformed red blood cells, especially in the pulmonary capillaries in the case of thoracic syndrome. Recent studies demonstrate that falciformation of erythrocytes increases the expression of α 4 β 1 integrins and their adhesion to the VCAM-1 receptor of endothelial cells. This activation is responsible for a prothrombotic state with also their adhesion to leukocytes, inflammation, and deregulation of vascular tone with vasoconstriction, aggravating ischemia, and thus creating a vicious cycle. Elevation of leukocytes would be correlated with a risk of occurrence of ATS.

Clinically, ATS has been defined by the American National Acute Chest Study group, by the appearance of a new radiological pulmonary infiltrate concerning at least one pulmonary segment and symptoms suggestive of pneumonia, i.e.: fever greater than 38.5°C, chest pain, respiratory signs or symptoms (tachypnea, sibilance, cough or the appearance of respiratory draught) and/or hypoxemia. Note that pulmonary infiltrates are frequently bilateral and predominate at the bases. Pleural effusions may also be present in half of the cases.

Known risk factors for the development of ATS are young age (higher incidence rate in the pediatric population), lowered fetal hemoglobin (HbF), higher chronic elevation of HbS and leukocytes.

The most common possible etiologies are infections, pulmonary infarction, pulmonary embolism, fat embolism following a vaso-occlusive crisis, and steroid treatment.

As for infections, which are involved in about 30% of cases, the majority are related to atypical germs (Mycoplasma pneumoniae and Chlamydia pneumoniae), although viral infections may also be involved.

Recently COVID 19 infection was the cause of decompensation in sickle cell patients during the pandemic period, which was the case in our patient.

In order to make the diagnosis, lung imaging is essential. Conventional radiography can usually suffice, but a frequent radiological delay in relation to the clinic sometimes justifies a CT scan without contrast injection, which also allows the extent of pulmonary involvement to be assessed. However, angio-CT is not indicated unless pulmonary embolism is strongly suspected clinically.

A blood gas analysis is used to assess the presence of hypoxemia, the LDH level assesses the degree of hemolysis and the hemoglobin level guides the choice of therapies. Active search for germs is also essential by performing sputum cultures, nasopharyngeal swabs and blood cultures.

In case of unfavorable evolution, a bronchoalveolar lavage (BAL) can be discussed, in order to search for a germ and optimize antibiotic therapy. The presence of foamy cells in the BAL fluid is in favor of fat embolisms.

The therapeutic management of ATS consists mainly of symptomatic treatment, including oxygen therapy, hydration, analgesia, thromboprophylaxis, physiotherapy in the form of incentive spirometry (to reduce the occurrence of atelectasis favored by chest pain and pulmonary superinfection) and possibly bronchodilators.

In view of the high frequency of infections and the difficulty in differentiating between infectious and non-infectious origins, broad-spectrum antibiotic therapy is recommended, mainly with coverage of typical and atypical germs with a betalactam and a macrolide or a quinolone.

Transfusion therapies (simple transfusions, manual exchanges or machine erythrocytapheresis) have a central place in the management of ATS and should be considered early.

Among long-term treatments, hydroxyurea reduces recurrence rates by decreasing HbS levels and increasing HbF levels, with or without iterative transfusions or erythrocyte exchanges. L-glutamine has recently shown good efficacy in the treatment of sickle cell disease, reducing the risk of ATS.

CONCLUSION

Acute chest syndrome is an extreme therapeutic emergency requiring early management to avoid fatal complications in sickle cell patients.

Infection with COVID 19 is a new infectious etiology of this syndrome that should not be ignored.

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