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Asthma Attack Revealing Sickle Cell Disease

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Abstract

Sickle cell disease is a genetic disorder characterized by the presence of hemoglobin S (HbS) and a significant reduction in normal hemoglobin A (HbA) in red blood cells. In deoxygenated conditions, HbS molecules polymerize, causing vascular occlusions and hemolysis. Pulmonary complications associated with this disease result from vascular occlusion, ischemia-reperfusion, and inflammation. Literature reports that asthma is observed in 30% to 70% of patients with sickle cell disease. We present the case of a boy whose sickle cell disease diagnosis was established following an asthma attack.

Keywords

Sickle Cell Disease, Asthma, Morbidity

1. Introduction

Sickle cell disease, also known as sickle cell anemia, is a genetic disorder of hemoglobin with autosomal recessive inheritance [1]. A point mutation causes the substitution of glutamic acid with valine at the sixth codon of chromosome 11. This mutation leads to a structural and functional alteration of hemoglobin, known as hemoglobin S. Under certain conditions, this results in the formation of abnormal red blood cells, called sickle cells [2]-[4]. Pulmonary complications in sickle cell disease are related to vascular occlusion, ischemia-reperfusion, and inflammation [5]. Asthma appears to be more frequently underestimated in this population, with an unknown pathogenesis. We report the case of a boy whose sickle cell disease was diagnosed following an asthma attack.

2. Clinical Case

A 10-year-old patient, the eldest in his family, from a non-consanguineous marriage, with no family history of atopy, and no history of chronic vomiting or gastroesophageal reflux, was seen in a pediatric pulmonology consultation at

Hassan II University Hospital (CHU) in Fes for the management of wheezing dyspnea.

The onset of symptoms began at the age of 8, with repeated episodes of wheezing dyspnea prompting the family to seek emergency care at the provincial hospital (CHP) in their region each time. The child was placed on symptomatic treatment, including nebulization with Ventolin and oxygen therapy.

On clinical examination, the child is alert, with a respiratory rate of 25 cycles per minute and a heart rate of 89 beats per minute, both normal for his age. He weighs 28 kg, which is average for his age, and is 134.5 cm tall, within the normal range for standard deviation (SD), with a slight conjunctival jaundice. The pleuropulmonary examination reveals a thorax of normal morphology, clear vesicular breath sounds, well-transmitted vocal vibrations, and bilateral wheezing without signs of respiratory distress. Abdominal examination shows no hepatosplenomegaly. The cardiovascular examination and the rest of the clinical exam show no abnormalities.

In the biological assessment, the complete blood count shows a regenerative normochromic normocytic anemia with hemoglobin (Hb) at 9.4 g/dL (11.9 - 13.5 g/dl), a mean corpuscular volume (MCV) of 77 fL (77 - 84 fL) , a mean corpuscular hemoglobin concentration (MCHC) of 36.7 g/dL (32 - 38 g/dL), and a reticulocyte count of 153,864/µL. The leukocyte count is 9,180/µL (4500 - 13,500 cells/µL), with neutrophils at 2,607 cells/µL (1500 - 7000/µL), lymphocytes at 2,740 cells/µL (1500 - 7000 cells/µL), and eosinophils at 1120 cells/µL (50 - 500/µL). The platelet count is 354,000 cells/µL (150,000 - 450,000 cells/µL). An additional hemolysis panel showed lactate dehydrogenase (LDH) at 480 U/L (180 - 400 U/L), a severely decreased haptoglobin level of 0.03 g/L (0.52 - 1.38 g/L), and hemoglobin electrophoresis revealed: Hb F at 2.4% (less than 1%), Hb S at 53%, Hb C at 42.9%, and Hb A2 at 1.6% (2.2% - 3.3%), indicating heterozygous sickle cell disease. The chest X-ray was normal, the abdominal ultrasound was normal, and spirometry showed obstructive dysfunction. Based on these findings, the diagnosis of asthma was confirmed using clinical and functional criteria in a child with sickle cell disease. The patient was started on asthma maintenance therapy with inhaled corticosteroids and a short-acting $\beta 2$-agonist as needed, and was scheduled for follow-up in pediatric hematology. Genetic testing and evaluation of the parents and other children in the family were recommended, but could not be performed due to the family's financial limitations.

The outcome included a reduction in the number of exacerbations and hospitalizations.

3. Discussion

The increased prevalence of asthma in children with sickle cell disease compared to the general population is controversial. Retrospective studies have reported asthma rates of up to 50% [6], while recent large-scale prospective studies show that the frequency of asthma diagnosed by a physician ranges from 14 to 28%, comparable to that of the general population of the same origin [7] [8]. Compared

to atopic asthmatic children, children with sickle cell disease who are diagnosed with asthma less frequently present with nasal or skin allergic manifestations and have less frequent family histories of asthma [8]. Their rate of sensitization is also lower [8]. However, elevated IgE levels are more common in children with sickle cell disease than in the general population and are associated with more severe asthma and increased morbidity [8]. There is no significant hyper eosinophilia in sickle cell patients [8] nor a significant increase in FeNO levels [10] [11]. Asthma increases the risk of acute chest syndrome, vaso-occlusive crises, and transfusion needs [12]-[15]. Asthmatic children hospitalized for a painful crisis are four times more likely to develop acute chest syndrome during their stay compared to non-asthmatic children [15].

There are no specific recommendations for the treatment of asthma in patients with sickle cell disease, and no medication is contraindicated for maintenance therapy. Given the high risk of acute complications associated with asthma, maintenance therapy is necessary to ensure optimal symptom control and prevent exacerbations [16]. Systemic corticosteroids, particularly at high doses and with abrupt discontinuation, are associated with an increased risk of readmission for vaso-occlusive crisis and prolonged hospitalization [16] [17]. Their use in asthma exacerbations requires close monitoring and, if necessary, a blood transfusion exchange [3].

4. Conclusion

The management of the child is quite complex, requiring a comprehensive approach, as subtle signs can be crucial for establishing the correct diagnosis. This was demonstrated in our case, where the patient was treated solely for asthma for two years before the accurate diagnosis was made. On the other hand, asthma increases the risk of acute chest syndrome, vaso-occlusive crises, and transfusion needs. This underscores the importance of effective asthma management to reduce the mortality risk associated with sickle cell disease, particularly in developing countries.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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