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Case Report

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Hemoglobin Osu-Christiansborg: A Rare Variant

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Abstract

Hemoglobin Osu-Christiansborg is a rare beta globin chain variant reported mostly in the families of Ghana, though a few cases have been reported in other parts of the world as well. This variant is mostly reported in combination with hemoglobin S but has not been identified to cause an overt sickle cell disease. Here, we report a case of a 3-year-old boy from Ghana who was found to have hemoglobin S+ Osu-Christiansborg on screening hemoglobin electrophoresis.

Keywords: Osu-Christiansborg hemoglobin; Sickle cell; Rare variant

Introduction

Hemoglobin Osu-Christiansborg is a rare variant resulting from mutations in beta globin chain of hemoglobin [1]. The variant was originally described in an individual from Ghana, West Africa, but over the years, a few cases have been reported from different parts of the world including Iran and Brazil [2]. Here, we present a case of a 3-year-old boy, admitted with acute bronchiolitis episode and incidentally found to have hemoglobin S+ Osu-Christiansborg on screening hemoglobin electrophoresis.

Case Report

This was a 3-year-old boy who presented to the emergency department with 4 days of intermittent fever, non-productive cough and decreased oral intake. In the ED, physical examination was remarkable for nasal congestion and wheezing on lung auscultation. Chest X-ray revealed bilateral haziness. The patient was treated with albuterol/ipratropium X3, prednisolone (2 mg/kg/dose) X1, ceftriaxone and referred for admission to pediatric floor for further management. On the floor, the patient was started on frequent bronchodilator therapy which was spaced as tolerated. The IV antibiotic and IV fluids were also continued. On detailed history from the mother, it was revealed that the patient had some sort of sickle cell variant/disease and that both the parents were sickle cell trait. Mother denied any episodes of dactylitis or pain crisis. The family came from Ghana, and mother did report that some family members on father's side have a rare type of sickle cell variant. Since the patient's primary care was at a different institution and no records were available, a screening hemoglobin electrophoresis was sent, among other labs for infection panel. The acid and alkaline hemoglobin electrophoresis revealed Hb pattern: S+ Osu-Christiansborg with HbS 48.2% and 43.8% of Osu-Christiansborg variant (beta variant). Hematologist was consulted and recommendation was made to refer the patient for outpatient follow-up. During initial hospitalization, the patient had persistent respiratory distress requiring addition of PO antibiotic (azithromycin). During the course of hospitalization, the respiratory distress gradually improved and the bronchodilator was spaced as tolerated. The patient was discharged after 3 days of hospitalization to continue treatment at home (albuterol, prednisolone, and azithromycin). A follow-up appointment was scheduled with hematologist for further evaluation of the sickle cell variant and to plan for intervention, if needed.

Discussion

Hemoglobin Osu-Christiansborg is a rare, non-pathological hemoglobin variant mostly seen clustered in families from Ghana, though few scattered cases from different parts of the world have also been recognized. This variant is thought to result from substitution of aspartic acid (at beta 52) by aspargine at an external position in the D helix of beta chain of hemoglobin. Hemoglobin Osu-Christiansborg migrates like hemoglobin S at alkaline pH and like hemoglobin A at acidic pH, making it possible for a compound heterozygous state to be mistaken for sickle cell disease. On agar gel electrophoresis at pH of 6.0, the two hemoglobins separate, one in position of hemoglobin S and one in the position of hemoglobin A. From the

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review of all the cases detected until now, this variant seems to be clinically benign and not associated with any major apparent clinical manifestations. Even when present in combination with hemoglobin S, this variant has not been reported to cause an overt sickle cell disease [3]. However, a case of a 63-year-old man from Ghana with hypertension and hemoglobin S+ Osu-Christiansborg did report occurrence of bone pains in his twenties with no recurrence later on [3].

Conclusion

Hemoglobin Osu-Christiansborg is a rare non-pathologic hemoglobin variant that is generally only recognized as a part of routine screening. Even in compound heterozygosity with sickle hemoglobin, it does not cause clinical complications although can be mistaken for sickle cell anemia on screening laboratory assays. Its higher occurrence in a cluster of families from Ghana highlights the need to screen these individuals using both acid and alkaline hemoglobin electrophoresis to avoid misinterpretation as sickle cell trait or disease and to identify the true hemoglobin variant. Through this case, we want to bring to light the importance of studying these variants both biochemically and in relation to clinical course so that clinical guidelines can be developed for this special group of hemoglobin variants.

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