Hypogammaglobulinemia in sub-Saharan Africa: a case report and review of the literature

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Abstract: Patients with hypogammaglobulinemia are susceptible to recurrent bacterial, viral, fungal, and parasitic infections. The most common clinical manifestation includes recurrent severe infections caused by encapsulated bacteria, in which antibody opsonization is the primary defense mechanism. To our knowledge, this is the first case report of hypogammaglobulinemia in a Ugandan child in Sub-Saharan Africa. The case emphasizes the importance of including hypogammaglobulinemia in the differential diagnosis for children presenting with a history of recurrent infections.

Aim: To raise the index of clinical suspicion of hypogammaglobulinemia in an African child and allow for prompt recognition and management of hypogammaglobulinemia.

Keywords: hypogammaglobulinemia, recurrent infections, Uganda

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Introduction

The most common worldwide causes of immunodeficiency include HIV, malnutrition, and immunosuppressive agents. However, when HIV infection, malnutrition, and other secondary causes of hypogammaglobulinemia have been excluded, it is important to consider a primary cause of hypogammaglobulinemia. Primary humoral immunodeficiencies are characterized by recurrent, severe upper and lower respiratory tract infections due to encapsulated bacteria. Although the incidence of hereditary immunodeficiency is low when compared to acquired immunodeficiency, the prevalence of people affected by a primary immunodeficiency is as high as 1 in 500 persons¹. We report a case of a 9-year-old Ugandan boy with the diagnosis of hypogammaglobulinemia.

Case Report

A 9-year-old African boy first presented to Mulago hospital in Kampala, Uganda at the age of 5 years with increasingly frequent and severe recurrent respiratory tract infections since infancy. The infections started with a measles-like illness at 3 months of age, which had responded to broad spectrum antibiotics. He developed bacterial meningitis at the age of two, which was successfully treated. He has had a history of recurrent severe pneumonia's approximately two per year since age 2.

At age 4, chest radiograph and computer tomography of the chest revealed multi-segmental consolidation and collapse of the right middle lobe. He was given several courses of antibiotics with no improvement in symptoms. The recurrent episodes of pneumonia persisted. Six months later the patient underwent a right middle lobectomy for persistent pneumonia unresponsive to medical treatment. Pathology revealed localized bronchiectasis with atelectasis and negative cultures. Despite the lobectomy, the recurrent respiratory tract infections did not subside.

After the lobectomy, he required several hospitalizations, including episodes of septic arthritis of the left knee joint and of pyogenic meningitis. Repeated HIV testing on the child and parents were negative. Immunoglobulin studies were sent to the USA to aid in di-
lymphocytes will be present in patients with X-linked immune activation, hepatosplenomegaly may be present.

Discussion

On physical exam, he was small for his age, but otherwise in fair condition. He had somewhat labored breathing and coarse crepitations, more marked on the right upon posterior chest auscultation. Heart sounds were normal with no murmurs. There was no cervical, supraclavicular, or inguinal lymphadenopathy. Abdomen showed no splenomegaly or masses. Skin was free of neurocutaneous stigmata, rashes or bruises. Neurologic exam was intact.

This child's extensive history of chronic infections, physical exam findings, imaging studies, and lab tests suggest a diagnosis of hypogammaglobulinemia. The child was given Septra for prophylaxis, since IVIG was too costly and not available. The child is alive to date. Diagnosis is based on a careful history, family history, and physical exam. Laboratory tests that should be obtained if a primary humoral immunodeficiency is suspected include a complete blood count with differential, quantitative serum immunoglobulin levels (IgG, IgA, and IgM), and measurement of antibody titers to vaccines. Treatment options include immune globulin replacement therapy which is very effective in reducing bacterial infections, though of limited availability in sub-Saharan Africa. Two forms of immune globulin therapy available include intravenous (IVIG) and subcutaneous (SCIG). The standard initial dose of IVIG is between 300 to 500 mg/kg every three to four weeks. Dosing is determined based on the patient's weight, trough levels of IgG after treatment has started, and the clinical response of the patient after treatment. Not only is an accurate diagnosis important for the patient, but it is also important for the patient's family members who may be female carriers of the disease.

Review of Literature

A literature review shows no other reported cases of X-linked hypogammaglobulinemia reported in Sub-Saharan Africa. This is a case of a 9-year-old boy in sub-Saharan Africa, who presented with frequent and recurrent respiratory tract infections since infancy. Based on immunoglobulin level studies, it was determined that he had X-linked hypogammaglobulinemia. Limitations in making the diagnosis in sub-Saharan Africa include lack of resources available for immunoglobulin testing and potential lack of awareness of the disease. However, with this case report, we hope to make the diagnosis of X-linked hypogammaglobulinemia more known in sub-Saharan Africa.

Author contribution

Each author contributed equally to this case report.

Conflict of interest

None of the authors have a conflict of interest.

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References