



A Delta Beta Thalessemia Female with Autoimmune Hemolytic Anemia due to Epstein Barr Virus: A Case Report

Authors

Tasha Vasaya, Bageshree Seth, Jeetendra Gavhane

Abstract

A number of extrinsic agents and disorders may lead to premature destruction of red blood cells. Among the most clearly defined are antibodies associated with immune hemolytic anemias.⁽¹⁾ The hallmark of this group of diseases is a positive coombs test (direct antiglobulin).⁽⁴⁾ Various immune hemolytic anemias are autoimmune and may be idiopathic or related to various infections, immunologic diseases, immunodeficiency diseases, neoplasms or drugs. In this case report, we discuss a case having autoimmune hemolytic anemia due to Epstein-barr virus.

Introduction

Autoimmune hemolytic anemia (AIHA) is a rare disorder characterized by the premature destruction (hemolysis) of red blood cells at a rate faster than they can be replaced. They can be further divided into AIHA caused by warm reactive antibodies or cold reactive antibodies. Those RBCs with warm antibodies undergo molecular mimicry and are usually without an underlying cause (primary). If an underlying cause is present it becomes a secondary condition. IgG antibodies that generally react with protein antigens on the RBC surface at body temperature. For this reason, they are called "warm agglutinins" even though they seldom directly agglutinate the RBCs. IgM antibodies that generally react with polysaccharide antigens on the RBC surface only at temperatures below that of the core temperature of the body. They are therefore called "cold agglutinins."⁽¹⁾

Case Report

A 12year old female came with features of CCF, jaundice, cola colored urine since 3 days prior to admission. She was a k/c/o delta beta thalassemia diagnosed 4 months back and required transfusion once in the past. This time she had anemia, thrombocytopenia, indirect hyperbilirubinemia, hepatomegaly and cervical lymphadenopathy. Urine routine was suggestive of hemoglobinuria. Corrected retic count was 3%. HLH was ruled out by doing a bone marrow which was normal. Her direct coombs test came positive. Work up for autoimmune hemolytic anemia was done in which ANA was negative, hepatitis B, HIV and Mycoplasma igM were negative. Her EBV viral capsid antigen igM was positive (118U/ml). She went into secondary renal failure and succumbed the next day.

Discussion

In children hemolytic anemia is usually caused by intrinsic red cell defect leading to hemolysis and anemia is seen typically in most of the inherited conditions.⁽³⁾ At times hemolytic anemia can result due to factors extrinsic to red cell and most of these causes are acquired in nature. The few common causes of extrinsic factors could be any autoimmune disorders (systemic lupus erythematosus, evan's syndrome), specific infections (infectious mononucleosis, mycoplasma), drug induced or malignancies. These secondary or extrinsic causes occurs more common in adolescents and young adults and is more common in females.

Epstein Barr Virus (EBV) is a gamma herpesvirus that has a double-stranded DNA genome of 184-kb pairs in length, encoding nearly 100 proteins. Two distinct types, type 1 and type 2 (also called type A and type B), share 70% to 85% sequence homology. EBV-1 is more prevalent worldwide than EBV-2. EBV is known to cause infectious mononucleosis, various lymphoproliferative diseases and epithelial malignancies like virus associated hemophagocytic syndrome, Burkitt's lymphoma.

Our case presented with fever, hematuria and severe anemia. The child was also a known case of delta beta thalassemia and hence the condition was exaggerated due to the underlying infection.⁽²⁾

Conclusion

The relation of EBV with AIHA has been described as rare in the literature. However if a patient presents with such symptoms EBV should be suspected since that is the most common trigger in children for autoimmune hemolytic anemia.

References

1. Textbook of Hematology And Oncology of Infancy: Nathan and Oski: Chapter 13: Autoimmune Hemolytic Anemia: Stella T. Chou and Alan D. Schreiber: Pg411-429

2. Textbook of Hematology And Oncology of Infancy: Nathan and Oski: Chapter 37: Hematological Manifestations of Systemic Diseases: Rachael F. Grace: Pg 1175-1179.
3. Transfusion Medicine and Hemotherapy: Article- The Clinical Pictures of Autoimmune Hemolytic Anemia: Charles H. Packman. www.karger.com
4. Journal OF Infectious Diseases and Immunity. Vol 3(1). Pp14-16, January 2011: Direct antiglobulin test(Coomb's) positive autoimmune hemolytic anemia induced by Epstein-Barr virus infectious mononucleosis in two children: Mehmet Akin, Ozlem Sahin, Kazim Kucuktasci, Burcin Kaya, Ersin Gozkeser, Murat Caglar et al: Department of Paediatrics Hematology, Faculty of Medicine: Pamukhale University: Denizli, Turkey.