

Complete Heart Block in β - Thalassemia Minor with Pregnancy: A Case Report

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Abstract: This is a case report of a 22-year-old Indian woman with 37 week gestation (primi-gravidae), a known case of congenital β -thalassemia trait came for routine check-up without any complaints. She has a fixed bradycardia (heart rate 45 beats/minute-BPM) and her electrocardiogram showed complete heart block (CHB). The relation between β -Thalassemia trait and third degree heart block is not reported in the medical literature. The correct diagnosis of the patient was based on appropriate clinical observations, ECG findings and deliberation of the differential diagnosis.

Keywords: β -Thalassemia Trait, Complete Heart Block and Pregnancy

Introduction

β -Thalassemia trait is an autosomal recessive disorder characterized by a point mutation at IVS1-5GC location on the β -globin gene on chromosome 11 (heterozygous condition), resulting in the defective synthesis of the β -globin subunit of hemoglobin (Kumar et al. 2004, Lekawanvijit & Chattipakorn 2009). β -thalassemia trait affects the hemoglobin (Hb), in the red blood cells (RBCs). People with β -thalassemia do not make enough Hb. People with β -thalassemia trait have both normal hemoglobin-A and the abnormal β -thalassemia Hb in their RBCs. Normally, β -thalassemia trait does not cause any health problems. If one (1) parent has β -thalassemia trait and the other parent has normal hemoglobin A, there is a 50 percent (1 in 2) chance with each pregnancy of having a child with β -thalassemia trait. β -thalassemia is common in people of African, Mediterranean, Asian and Middle Eastern descent. In a population-based study comparing all pregnancies of women with and without β -thalassemia minor conducted during the years 1988-2002 at Soroka University Medical Center, Negev, Israel revealed that of the 1,59, 195 deliveries, 261 (0.2%) occurred in patients with β -thalassemia minor. Furthermore, this study also indicated that β -thalassemia minor was not found as an independent risk factor for cesarean delivery (Sheiner et al. 2004).

First and second-degree heart blocks are a common rhythm disorders in thalassemia patients but the simultaneous presence of complete heart block (CHB) and β -thalassemia trait is a very rare combination. We present here the case of a 22 year old woman with 37 week gestation presented with 3rd degree heart block. To the best of our knowledge, the incidence of complete heart block with β -thalassemia trait has not previously been reported in the literature.

To the best of our knowledge, this is the first case report of 3rd degree or CHB in α and β -thalassemia trait (minor).

Case Report

A 22 year old woman with 37 week gestation came to our MGM hospital, Navi Mumbai for routine checkup. She had no major illness since childhood, and had never received blood transfusion. However, her recent medical history revealed that she had been diagnosed with congenital β -thalassemia minor, which was identified when she was in her first trimester of pregnancy. She has two younger sisters aged 20 and 18 yrs. It is not known whether her parents and younger siblings are also carrier of this trait. Relevant history about her family sibling was not immediately available.

Routine general, clinical and laboratory examination revealed following findings. Her pulse rate (44/min), blood pressure (106/73 mmHg), Hemoglobin (10.9 mg/dL), mean corpuscular volume (MCV) (96.02 fL), hemoglobin-A2 (5.15%), total white cells ($11.26 \times 10^9/L$), platelets ($171 \times 10^6/L$), serum creatine (0.67 mg/dl), blood glucose (70 mg/dl), serum iron (222 μ g/dl), ferritin (7691 ng/ml), total iron binding capacity (TIBC) (233mg/dl), serum alanine aminotransferase (AlAT) (78.12 IU/L), serum aspartate aminotransferase (AAT) 52.18 IU/L. The electrocardiogram (ECG) showed complete heart block. The 2 D echo was normal. She was admitted in the intensive care unit (ICU) and temporary pacemaker was inserted and paced at 60beats/minute (bpm). Since it is a congenital heart block permanent pacemaker is not essential. She underwent a successful caesarian delivery 5 days later. She made an uneventful recovery. Pacemaker sheath was removed on post-operative day 3 and she was discharged on day 5.



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Discussion

We presented here a case of pregnant women with a congenital β -thalassemia minor. Written informed consent was obtained from the patient for publication of this case report. Based on the clinical history, examination, ECG studies and laboratory investigations, the patient was diagnosed with congenital Complete Heart Block (CHB).

We could find one study on CHB with β -thalassemia major (Küçükosmanoğlu et al. 2002). However, we did not find any reports published in English on congenital CHB in β -thalassemia minor. However, Cardiac complications of β -thalassemia trait were first described in 1964 by Engle et al. (Engle et al. 1964). In the year 2004 Cohen and colleagues have demonstrated an increase in heart size in one third of thalassemic patients with first degree of heart block after 10 years of age (Cohen et al. 2004). Bradycardia, ST segment and T wave changes, infrequent premature atrial or ventricular contractions and first-degree atrio-ventricular block are most common findings in early stages (Veglio et al. 1998) that may progress to tachycardia and second-degree or third-degree (complete) heart block in the late stages (Engle et al. 1964).

β -thalassemia trait is a genetic disorder and not a life-threatening health condition, but can affect the quality of life due to mild or moderate anemia and should only be treated if the patient becomes symptomatic. It cannot become worse or turn into a serious disease, but it affects the health of children (1 in 2) born to the couples who are carriers of this trait. β -thalassemia trait, if not treated effectively, erythropoiesis is ineffective thereby iron absorption would be affected. Hence, the use of oral iron supplementation should be monitored by measuring the serum ferritin levels at regular intervals. The co-existence of other disease with β -thalassemia trait has been shown in various other studies including asthma (Palma-Carlos et al. 2005) and mood disorders (Bocchetta 2005). Genetic counseling is an essential part of managing such patients, where β -thalassemia trait testing is not done routinely.

In conclusion, the simultaneous occurrence of CHB and β -thalassemia trait is a rare incidence which has not previously been reported. Therefore it is important that such a case be presented in the international literature as a reference point. Clinical suspicion, ECG and the consideration of genetic diagnosis are important for the correct diagnosis and management of patients with CHB and β -thalassemia trait.

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