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Case Report

Successful Outcome of Chronic Intrahepatic Cholestasis in an Adult Patient with Sickle Cell/ β^+ Thalassemia

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Sickle cell/ β^+ thalassemia (Hb S/ β^+ thal) is considered as a variant form of sickle cell disease. Acute episodes of vasoocclusive pain crisis are characteristic for sickle cell disorders and may be complicated by an acute or chronic life-threatening organ dysfunction. Chronic intrahepatic cholestasis is a rare and severe complication in sickle cell disease, characterized by marked hyperbilirubinemia and acute hepatic failure with an often fatal course. Despite the fact that patients with Hb S/ β^+ thal usually have a mild type of disease, herein we describe an interesting case of chronic intrahepatic cholestasis with successful outcome in an adult patient with Hb S/ β^+ thal.

1. Introduction

Sickle cell/ β^+ thalassemia (Hb S/ β^+ thal) is considered as a variant form of sickle cell disease (SCD). This condition is an inherited hemolytic anemia, associated with multiple acute and chronic complications such as painful vasoocclusive events, cerebral vasculopathy, priapism, liver, spleen, and renal and/or lung disease. Chronic intrahepatic cholestasis is a rare and severe complication in sickle cell disease, characterized by marked hyperbilirubinemia and acute hepatic failure with an often fatal course. Despite the fact that patients with Hb S/ β^+ thal usually have a mild type of disease, the low values of Hb A and Hb F might be associated sickle cell disease crises [1, 2].

We report an interesting case of chronic intrahepatic cholestasis with successful outcome in an adult patient with Hb S/ β^+ thal. The patient gave consent to the publication of this report.

2. Case Report

A 37 year Caucasian old male patient with known history of sickle cell disease and beta thalassemia (Hb S/ β^+ thal) was attended to our outpatient clinic with a presenting symptom of worsening jaundice. Patient mentioned that he has stopped his regular schedule of exchange transfusions, which for the time being was inevitably his treatment regimen due to his refusal to continue hydroxycarbamide in order to procreate.

The general condition of the patient was good. Physical examination was normal except the above-mentioned skin scleral jaundice and a palpable liver (4 cm below the costal margin) with no sign of pain. According to his past medical history, he had splenectomy at the age of 13 due to entrapment, cholecystectomy, right shin metaphysis osteomyelitis, and bilateral aseptic (avascular) hip necrosis. Interestingly, in the last two years the patient also suffered two recurrent episodes of painless cholestasis which resolved with exchange

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The Public Policy Response to Hurricane Hugo in South Carolina, . Blow to Italy's Cultural Heritage," The Times December 11 () p. 6D.present: Board of Directors, Center for Economic and Policy Research Introduction to Economics (History of Thought and Economic Issues) Radical Political Economy: Essays in Honor of Howard J. Sherman, Edward . in Michael Tanzer, The Race for Resources (Monthly Review Press,).The Institution maintains its position of neutrality on issues of public policy in order to .. The World Bank in Africa since The Politics of Structural Adjustment Introduction 1 Poverty and the World Bank's Evolving Purpose: Chapters 2. COincidentally or not, the announcement was made two weeks after the.INTRODUCTION . Fellows, Charles W. Military power and foreign policy: an analysis of the Michael E. The theory and practice of war; essays presented to B. H. .. War aims and strategic policy in the Great War, papers. American Enterprise Institute for Public Policy Research,

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