

Corneal Leucomas in a Child with Sickle Cell Disease

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Introduction

Occlusion of blood vessels in various parts of the body occurs continually in sickle cell disease (SCD). Most important acute features of SCD are: recurrent painful vaso-occlusive crisis, acute chest syndrome, acute splenic sequestration, priapism and anemia [1]. Chronic organ dysfunctions are: cerebral vasculopathy leading to stroke, leg ulcers, osteonecrosis and retinopathy [2]. Corneal leucomas can be detected only with a formal eyes exam performed by an ophthalmologist, due to the fact that in the early stages of SCD eye's involvement is asymptomatic. All clinical manifestations of sickle hemoglobinopathies are classified according to the presence or absence of neo-vascularization in the eye: non proliferative and proliferative disease [3,4]. Ocular signs can affect the entire vascular bed in the eye and involve even other structures, such as the retina.

Case Report

We present herein a case of a 9 years old black male, with both parents hailing from Ghana, affected by SCD SS phenotype. The diagnosis was made when he was 13 months old, during a febrile infectious episode with severe anemia. The electrophoresis revealed: HbA 0%, HbF 17.2%, HbA2 4.5%,

HbS 78.3% and the genetic test showed homozygosity for Hb S (Hb beta Glu-6-Val). According to the International and National Guidelines for the management of sickle cell disease [5,6] the child was yearly followed up by an ophthalmologist. When he was 9 years old, multiple asymptomatic sub-epithelial corneal leucomas have been diagnosed. He was treated with eyes drop of betamethasone for 15 days with complete remission.

Discussion

Most studies of ocular involvement in SCD are focused on retinopathy, however we do suggest the importance of clearly differentiating ocular manifestations that require a therapy, from others that can be physiologic.

To our knowledge, there are little published data about non-retinal manifestation of SCD in children. Since childhood, all patients with sickle cell disease should be followed-up by an ophthalmologist.

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