Pituitary Abnormalities in Short Adolescents and Young Adults with Sickle-Cell Disease (SCD) and Recurrent Vaso-occlusive Crisis

Ashraf Soliman, Mohamed Yassin ^ , ElSaid M Bedair *

Department of Pediatrics  Alexandria University  Egypt Hematology ^ and Radiology *Hamad Medical Center, PO Box: 3050, Doha, Qatar

Introduction

Growth failure is the most frequent endocrine abnormality observed in patients with SCD. Decreased synthesis of IGF-I might be secondary to a disturbed GH-IGF-I axis and defective GH secretion has been reported in some patients. Infarction, atrophy, and hemorrhage may occur in the pituitary gland in SCD during or following the vaso-occlusive crisis.

Objectives

To define the possible abnormalities of pituitary gland in sickle-cell disease (SCD) we measured the circulating concentrations of insulin-like Growth factor –I (IGF-I) and studied the Magnetic Resonance Imaging (MRI) of the pituitary gland in 7 adolescents and young adults with SCD with short stature (HtSDS < -2) and history of recurrent painful crisis.

Methods and Results

Seven patients with SCD (age : 24.2 +/- 4.5 years) and short stature (HtSDS = 2.5 +/- 0.4) and history of severe and recurrent vaso-occlusive crisis ( at least 3 in the past 3 years) were studied. All were transfusion – dependent, with full pubertal development (Tanner’s stage 5) (euogonadal). They were regularly transfused since early childhood and underwent chelation therapy using desferrioxamine which was replaced by deferasirox for the last 4 -5 years.

Results

In the 7 patients with SCD circulating IGF-I were decreased (IGF-I SDS = -2.1+/-.5) compared to adults standards. Pituitary MR imaging showed abnormalities in 4/7 of these patients in the form of heterogeneous appearance of the anterior pituitary, presence of single or multiple hypointense foci due to hemosiderin deposition in the pituitary (4/7) and significantly decreased (2/7) or increased volume (1/7). These lesions can be explained by hemosiderosis of the gland and/or ischemia during the vaso-occlusive crisis.

Conclusion

Pituitary MR imaging showed significant abnormalities of the anterior pituitary gland in SCD patients with short stature and significant history of vaso-occlusive crisis.

This study demonstrated the value of MRI imaging of the pituitary to support investigating of the GH-IGF-I axis in these patients.