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

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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

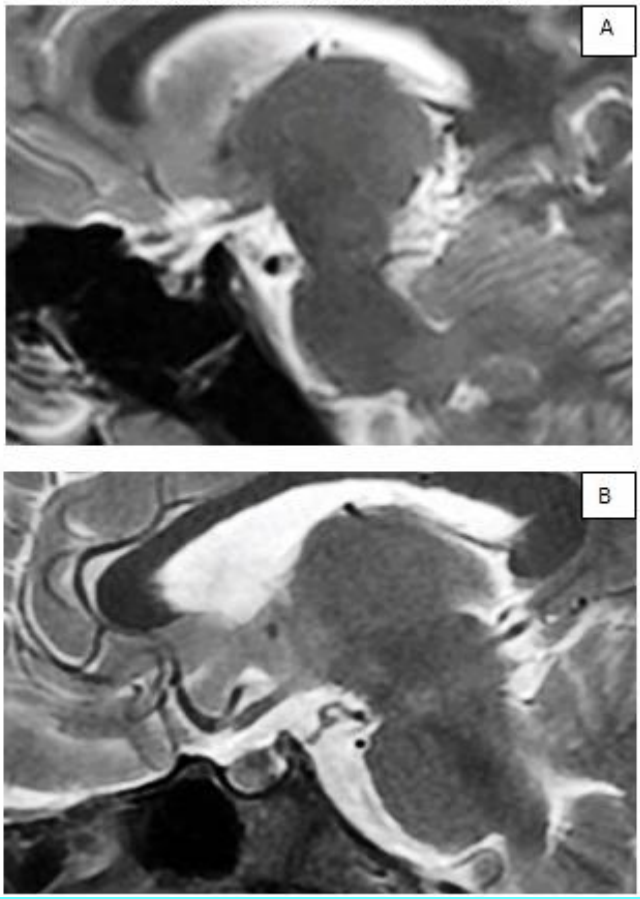
Seven patients with SCD (age : 24.2 +/- 4.5 years) and short stature (HtSDS = 2.5 +/- 0.4) and history of severe and recurrent vaso-occlusive crisis (at least 3 in the past 3 years) were studied. All were transfusion – dependent, with full pubertal development (Tanner's stage 5) (euogonadal). They were 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+/-0.5) 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

Results

Two pituitary MR images showing deposits of hemosidrin : A- one hypointense lesion, B- many spots



MRI in this study and CT imaging of the pituitary gland in patients with SCD (1) showed significant abnormalities of the gland in the form of deposits of hemosiderin as well as volume changes.

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.

Soliman AT, Darwish A, et al . J Trop Pediatr. 1995 Oct;41(5):285-9.