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Gyrate Atrophy of the Choroid and Retina in a Young Patient: A Case Report

Romaissae Benkirane*

Department of Ophthalmology, Ibn Sina University Hospital Centre, Mohamed V Souissi University, Rabat, Morocco

***Corresponding author:**

Romaissae Benkirane,
Department of Ophthalmology, Ibn Sina University Hospital Centre, Mohamed V Souissi University, Rabat, Morocco

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1. Abstract

Gyrate atrophy (GA) of the fundus is a rare autosomal recessive disease characterized by deficiency of orni-thine-δ-amino-transferase (OAT). OAT deficiency causes hyperornithinaemia, which results in progressive Chori retinal atrophy. We report the case of a young male with Gyrate atrophy

2. Introduction

Gyrate atrophy (GA) of the choroid and retina is a rare, autosomal recessive inherited disease causing progressive Chori retinal degeneration resulting in blindness. The primary defect is due to a deficiency of the enzyme ornithine-aminotransferase, which is responsible for markedly elevated levels of ornithine in plasma and other body fluids [1].

3. Presentation

A 24-year-old male patient presented with complaints of gradual visual loss and progressive night vision deterioration occurring over the past several years. The best corrected visual acuity was 4/10 in both eyes. The refractive error was-2;00 D in the right eye and -1.00 D in the left eye. On examination, his fundus exhibited bilateral severe Chori retinal atrophy involving the mid-periphery. Fundus fluorescein angiography revealed leakage at the margin of Chori retinal atrophy. Dynamic Goldmann perimetry demonstrated visual field constriction in both eyes, and a full-field electroretinogram showed markedly impaired photopic and scotopic responses.

On plasma amino acid analysis, he had markedly elevated plasma levels of ornithine. The clinical diagnosis of both patients was consistent with OAT deficiency and GA of the choroid and retina.

The patient was treated with vitamin B6 (pyridoxine) 300 mg daily and an arginine-restricted diet.

4. Discussion

Gyrate atrophy of the choroid and retina is a rare autosomal recessive dystrophy; their incidence is less than one in 1,000,000 in the world [2]. Patients with GA initially complain of decreasing visual acuity and loss of night vision. Eventually, loss of central vision occurs in the fourth to fifth decades. The fundus in patients with GA exhibits circular, well demarcated chorioretinal atrophy with hyperpigmented margins in the midperiphery. Patients with GA may also have myopia and posterior subcapsular cataracts. Usually, the fundus finding of scalloped Chori retinal atrophy in the midperiphery is sufficiently characteristic to determine GA [3].

Plasma ornithine levels help to confirm GA. GA is a genetic disorder caused by OAT deficiency that results in markedly elevated levels of ornithine in plasma and other body fluids. The exact mechanism of Chori retinal atrophy due to hyperornithinaemia is not known, although a low-arginine diet and vitamin B6 supplementation may decrease plasma ornithine levels and reduce the progression of GA [4].

5. Conclusion

Gyrate atrophy (GA) of the fundus is a rare autosomal recessive disease characterized by deficiency of ornithine-δ-amino-transferase (OAT). OAT deficiency causes hyperornithinaemia, which results in progressive Chori retinal atrophy [4].

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